CASE REPORT POSTER VIEWING PRESENTATIONS

PV280 / #741

Case Report Poster Topic: *AS03 - Antiphospholipid Syndrome*
Late-Breaking Abstract

THE HEART OF IT ALL: DILATED CARDIOMYOPATHY AS THE INITIAL PRESENTATION OF ANTIPHOSPHOLIPID SYNDROME

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Introduction: Cardiac manifestations in primary antiphospholipid syndrome (APS) range from mild valvular disease, to more devastating disorders associated with morbidity and mortality. Cardiomyopathy has seldom been reported as the initial manifestation of the disease. Therefore, we present the case of a 47 year-old lady with dilated cardiomyopathy (DCM) as the initial presentation of APS.

Case Presentation With Investigation: A 47 year-old lady, previously healthy with no known comorbidities, presented with acute dyspnoea. This started hours prior to presentation. She described orthopnoea as well as gradual worsening of lower limb oedema in the preceeding days. The patient had no cardiac family history, and was a non-smoker and teetotal. Chest auscultation revealed crackles up till the lung apices. Further examination confirmed an elevated jugular venous pulse, as well as pitting lower limb oedema extending to the thighs bilaterally. Oxygen saturations were 89% on room air, correcting with oxygen. Inspection revealed livedo reticularis of the thighs and arms. A bedside echocardiogram revealed severe diastolic dysfunction. An NT-proBNP assay was elevated at 35,000 pg/mL. The patient was admitted under the cardiologists for diuresis and investigation. The patient underqent a coronary angiogram, which showed fully patent coronary arteries. A Cardiac MRI confirmed the presence of an advanced DCM, with thickening of the mitral and tricuspid valvular apparatus. Rheumatology was consulted in view of these findings. The patient tested positive for lupus anticoagulant, anti-β2 glycoprotein antibodies (IgG and IgM > 200 IU/mL) and anti-cardiolipin antibodies (IgG and IgM > 120 IU/mL). A double stranded DNA antibody assay was negative, with normal complement protein levels. She denied a history of thrombotic episodes and miscarriages. A diagnosis of primary APS causing DCM was made, and warfarin, corticosteroids, mycophenolate mofetil and heart failure optimisation were initiated. The patient subsequently improved and was discharged home.

Literature Review: The etiological basis of DCM in APS is hypothesised to be microvascular thrombotic insults, [1.], as evidenced by autopsy studies. Recurrent microthrombotic inflammatory activity has been associated with a risk of progression to DCM, with contributions from myofibroblasts and fibromuscular remodelling of the myoendocardium. Tumour necrosis factor alpha and transforming growth factor beta

are the main cytokines implicated in this process. APS patients, particularly primary APS patients and those with strongly positive serology, have been shown to exhibit asymptomatic diastolic dysfunction in up to 20% of cases [1.]. This suggests that the remodeling changes in the myocardium are asymptomatic and subclinical, yet capable of having catastrophic consequences. Immunosuppression and anticoagulation have shown success in the sparse case reports of APS-DCM in the literature, particularly with mycophenolate and IVIG [1]. Reference: [1.] Coletto L. Autoimmun Rev 2022; 21:102990

Discussion: The key learning points from this case are: 1. All women of middle-age and younger, presenting or having been diagnosed with DCM, should be screened for APS. 2. Cardiac MRI was, in this case, a reliable surrogate to endomyocardial biopsy. Further studies are needed to validate this. 3. Female sex, younger age and strongly positive serology are associated with an increased risk of DCM in APS patients. 4. Up to 20% of APS patients can have subclinical diastolic dysfunction. The authors recommend echocardiographic screening in view of the events of this case. 5. The etiology of APS-DCM is related to microvascular thrombosis, and therefore coronary angiography and macrovascular imaging will be normal in such cases. 6. Management involves warfarinisation and immunosuppression with steroids and DMARDs.

PV281 / #595

Case Report Poster Topic: *AS03 - Antiphospholipid Syndrome*

LUNG IS A BATTLEFIELD: DIFFUSE ALVEOLAR HEMORRHAGE IN ANTIPHOSPHOLIPID SYNDROME

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Introduction: This case highlights a rare and life-threatening complication of catastrophic antiphospholipid syndrome (CAPS) – diffuse alveolar hemorrhage (DAH). We emphasize that early recognition and treatment of DAH is vital due to significant morbidity and mortality.

Case Presentation With Investigation: A 41-year-old woman with a history of hypertension, stage 4 chronic kidney disease, and triple positive anti-phospholipid syndrome (APS) complicated by ischemic stroke, cerebral venous sinuous thrombosis on warfarin, spontaneous abortion, and multiple deep vein thromboses presented with increasing confusion. Her creatinine and blood urea nitrogen were rising (baseline creatinine 2.65 mg/dL to 3.63 mg/dL; BUN 68 mg/dL to 84 mg/dL), she developed oliguria, wasadmitted for progressive renal failure requiring intermittent hemodialysis, and started on IV heparin She became febrile and developed acute hypoxic respiratory failure with worsening encephalopathy, prompting transfer to the medical ICU for intubation and continuous renal replacement therapy. She underwent imaging which was negative for pulmonary embolism but demonstrated diffuse patchy airspaces, pulmonary vascular congestion, airspace infiltration, and a left lower lobe consolidation [Figure 1A-B]. She developed hemoptysis, was intubated due to decompensationand underwent bronchoscopy with gross bloody return. Hemoglobin and platelets trended down, raising concern for CAPS complicated by DAH. She met criteria for probable CAPS for triple positive antiphospholipid antibodies (aPL), and involvement of renal, respiratory, and neurological systems, though histological evidence of intravascular thrombosis was not obtained. Hematology was consulted, and high-dose glucocorticoids and plasma exchange therapy (PLEX) were initiated. She received daratumumab for immunosuppression. The patient was extubated on day five of PLEX and two days after starting daratumumab.

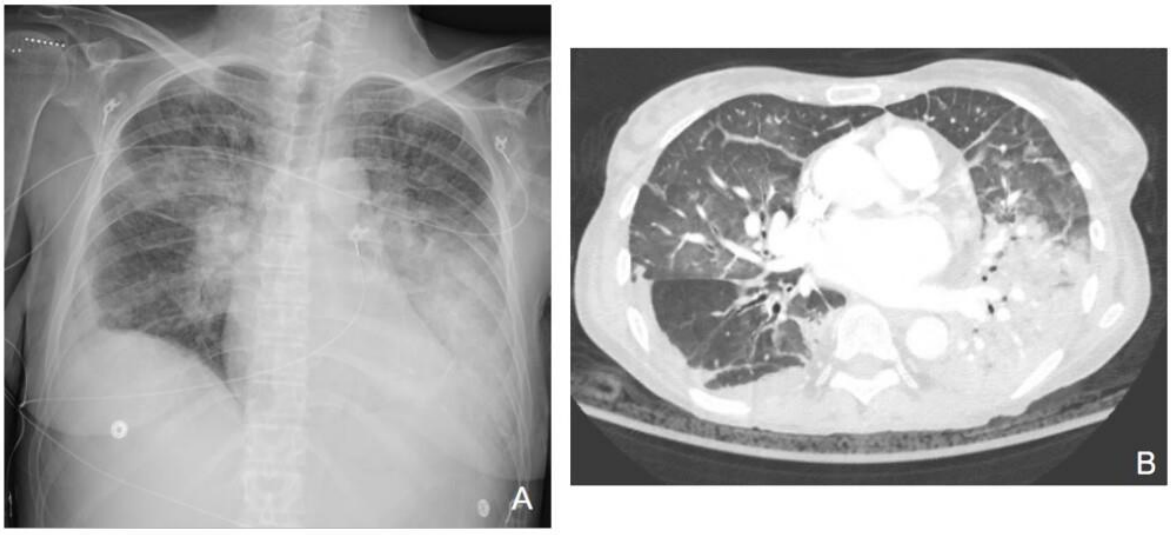

Figure 1. (A) Chest X-ray, AP view, diffuse airspace infiltrates, pulmonary vascular congestion. (B) CT angiogram, coronal view, scattered groundglass and consolidative opacities, extensive airspace infiltration.

Literature Review: Antiphospholipid syndrome (APS) is characterized by recurrent thrombotic events, and 30-40% of those with systemic lupus erythematous are positive for aPL [1-2]. A rare complication of APS is CAPS, where microvascular thrombi result in multi-organ failure, under 10% with CAPS experience DAH [2]. DAH in APS is attributed to an inflammatory process of up-regulated endothelial cell adhesion with neutrophil recruitment causing tissue destruction, and complement 5 activation mediating capillaritis [1]. Both processes result in hemorrhage precipitated by aPL [1]. Anti-beta-2-glycoprotein I antibodies, specifically, bind to platelets leading thrombosis [1-2]. Management includes anticoagulation and ventilatory support with high positive end expiratory pressure to reduce active lung bleeding [3]. PLEX has been shown to be beneficial in removal of antibodies and other cytokine mediators [1-3]. High-dose glucocorticoids remain a mainstay of treatment [1-3]. Immunotherapy has expanded to include the use of daratumumab to target long-lived plasma cells to induce antibody-dependent cellular cytotoxicity and complement-dependent cytotoxicity creating a degree of synergism [3].

Discussion: This case underscores that DAH, although rare, is life-threatening and necessitates prompt identification and intervention. Clinicians should have a high index of suspicion for DAH when patients with APS develop respiratory compromise and hemoptysis. This case also reinforces the value of a multi-disciplinary approach in patients with APS, combining critical care, hematology, nephrology, and rheumatology expertise to manage the complex interplay of thrombotic, inflammatory, and bleeding risks. The literature supports the use of high-dose glucocorticoids and PLEX, but use of daratumumab in severe CAPS is rare, and clinical trials are still ongoing. [1.] Cartin-

Ceba R. Arthritis Care Res (Hoboken) 2014;66:301-310. [2.] Loza C. Case Rep Rheumatol 2019:3284258. [3.] Yun Z. Front Immunol 2023;14:1144145.

PV282 / #36

Case Report Poster Topic: *AS03 - Antiphospholipid Syndrome*

ADRENAL FAILURE: WHEN ANTIPHOSPHOLIPID SYNDROME LEAVES SCARS

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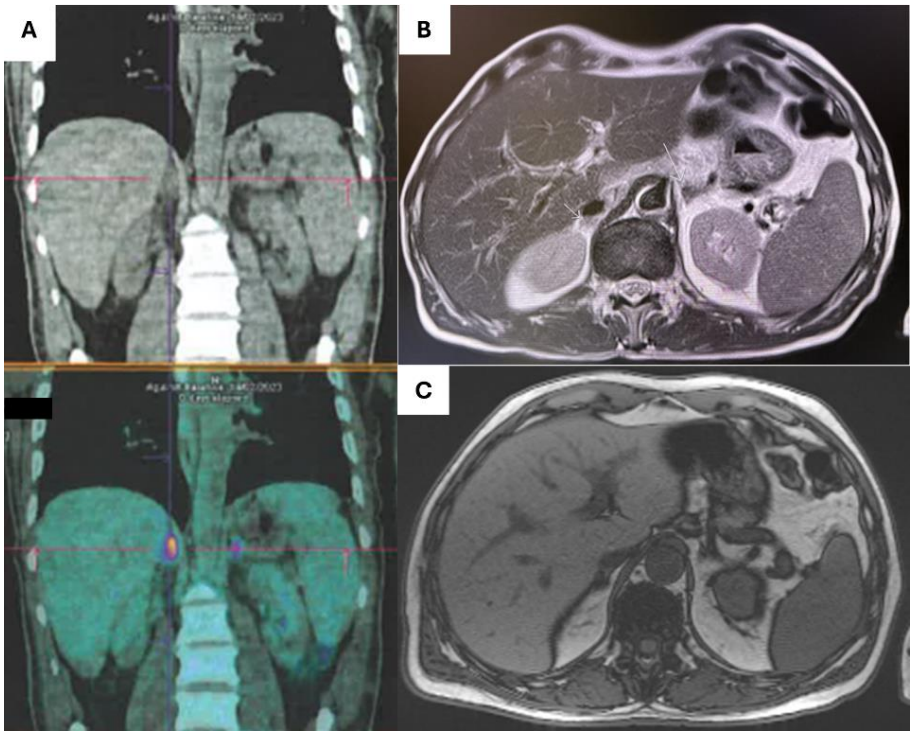
Introduction: We report a case of a 56-year-old male with a previous history of chronic mild thrombocytopenia, assumed to be a consequence of alcohol consumption (despite the absence of other alcoholic stigmas). He was admitted to the Internal Medicine Department with a 3-month history of extreme fatigue, anorexia, and weight loss (20%). Upon admission, he was hypotensive (100/54mmHg).

Case Presentation With Investigation: Blood tests revealed normocytic normochromic anaemia (Hb 9.9g/dL), thrombocytopenia (94 000x10^6/L), elevated activated partial thromboplastin time (76.4s, N 28-40), hyperkaliaemia (7.11mmol/L, N 3.5-5.2) (but normal sodium) and elevated inflammatory parameters (CRP 2.25 mg/dL, ESR 118 mm/h). An extensive workup study was conducted to exclude malignancy and infection. A PET-FDG showed intense uptake in both adrenal glands, with heterogeneity and areas of necrosis, especially in the right adrenal gland (Figure 1A). The endocrinology department was consulted, and hormonal assessments revealed a low serum cortisol (1.9 µg/dL; N 6.2-19.4) and a high adrenocorticotropic hormone (626.0pg/ml; N 7.2-63.3). PAI was assumed and intravenous hydrocortisone (200 mg/day) was started, with subsequent clinical (blood pressure, constitutional symptoms) and laboratory (blood cells count and inflammatory markers) improvement. The main causes for PAI, namely autoimmune Addison's disease, tuberculosis and human immunodeficiency virus infection, were excluded. At this moment, the Rheumatology department was consulted. Further workup revealed a positive lupus anticoagulant antibody (2 times in 12 weeks apart), ANAs 1/1280 (homogeneous nuclear pattern), anti-dsDNA antibodies elevation (517 UI/mL) and a weekly positive anti-nucleosome antibody. MRI scans showed atrophy of the adrenal glands (Figure 1B and 1C). The patient was diagnosed with SLE and APS, and after PAI control, he was discharged under glucocorticoid tapering (prednisolone 15 mg/day and fludrocortisone 0.05 mg/day), warfarin and hydroxychloroquine 400mg/day. Later on, azathioprine was also started (100mg/day) and the patient remained asymptomatic and with normal

laboratory parameters. Figure 1 – A: PET-FDG at diagnosis moment, showing intense uptake in both adrenal glands; B: abdominal MRI (T2) 3 months after the diagnosis, showing atrophy of both adrenal glands; C: abdominal MRI (T2) 6 months after the diagnosis, showing almost complete disappearance of both adrenal glands, comparing to the previous MRI.



Literature Review: Antiphospholipid syndrome (APS) is a multisystemic autoimmune disorder characterised by recurrent arterial, venous and/or microvascular thrombotic events. The disease rarely affects the endocrine system, especially at presentation. The involvement of the adrenal gland, although rare, can be severe. Possible mechanisms behind adrenal manifestations include multiple microthrombosis of the suprarenal vein leading to infarction and adrenal haemorrhage, atrophy and finally failure (primary adrenal insufficiency [PAI]). (1)(2)

Discussion: This case illustrates one of the rarest and still most severe consequences of APS. Patients with APS and adrenal haemorrhage, typically have bilateral involvement and develop adrenal insufficiency, just like our patient. (3) The disease can be fatal, thus early diagnosis and treatment as well as a close follow-up and multidisciplinary approach is needed to improve the prognosis of this rare disease.

References:

1. Hochberg MC, et al. Rheumatology 8th Ed. Chapter 135: Clinical features of systemic lupus erythematosus. P1113. Elsevier, 2022.
2. Bouki K. Hormones (Athens). 2023;22(3):521-531
3. Meade-Aguilar JA, et al. Clin Immunol. 2024; 260:109906

PV283 / #106
Case Report Poster Topic: *AS05 - CNS Lupus*

IMAGING INSIGHTS INTO JC VIRUS-ASSOCIATED PROGRESSIVE MULTIFOCAL LEUKOENCEPHALOPATHY IN NEUROPSYCHIATRIC LUPUS: A CASE REPORT

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Introduction: Progressive multifocal leukoencephalopathy (PML) is a rare, often fatal demyelinating disease of the central nervous system caused by JC virus reactivation in immunocompromised patients. [1.] PML is particularly challenging in patients with systemic lupus erythematosus (SLE), where the differential diagnosis between PML and neuropsychiatric lupus (NPSLE) can be difficult, especially when complicated by the use of immunosuppressive therapies like rituximab.

Case Presentation With Investigation: We present a 40-year-old female diagnosed with SLE and class IV lupus nephritis, managed with hydroxychloroquine 200 mg/day, mycophenolate mofetil 500 mg/day, and prednisolone 10 mg/week, presented with right hemiparesis. Neurological examination revealed muscle power scores of 3 in the right limbs and 5 on the left side. She was diagnosed with an SLE flare-up and left-predominant brain vasculitis, resulting in neuropsychiatric SLE with right hemiplegia. Pulse steroid therapy combined with Cyclosporine 100mg/day was initiated. However, her right limb muscle power declined to 0 after one month. Subsequently, pulse steroid therapy, plasma exchange, IVIG, and rituximab (total accumulated dose of 1000mg) were administered in the following month. However, one month following rituximab infusion, she exhibited cognitive dysfunction, including dyscalculia, apraxia, and dysphagia. Over the subsequent two months, her conscious level gradually declined to E2V1M2, accompanied with quadriparesis. Contrast-enhanced MRI revealed larger T2-hyperintense areas involving bilateral cerebral white matter, basal ganglia, thalami, left midbrain, and left cerebellum (Figure 1). Electroencephalography demonstrated continuously diffuse theta to delta slow waves over bilateral hemispheres without epileptiform discharges or spike waves, indicative of mild generalized encephalopathy. A lumbar puncture was performed, and cerebrospinal fluid examination identified JC virus by PCR method. Now she is under IVIG/3 weeks, and immunosuppressive medications is tapered gradually. Additionally, mefloquine with mirtazapine was initiated after family consultation.

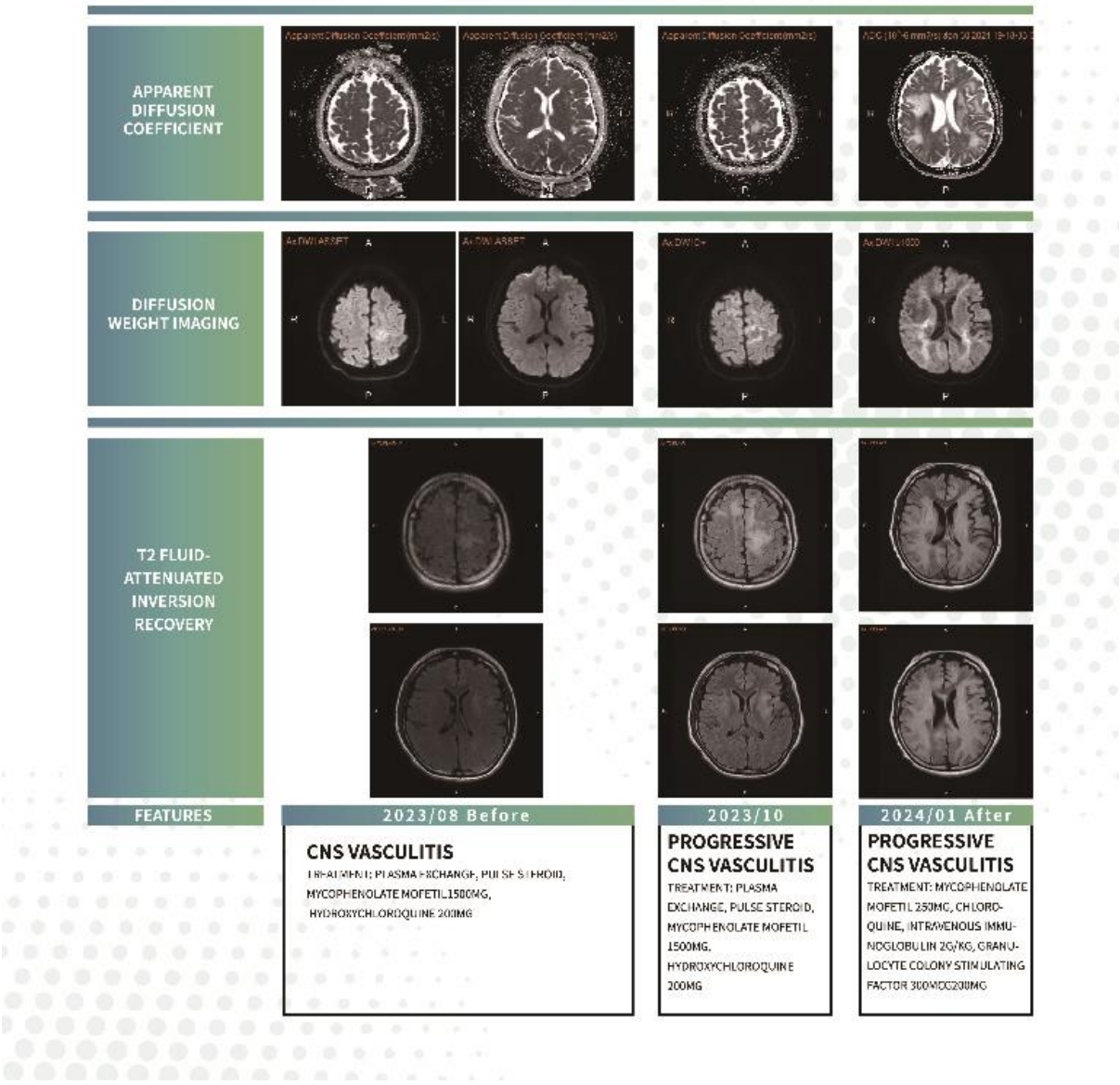

Figure 1|MRI progression in a 40-year-old female with SLE and CNS vasculitis evolving into JC virus-related progressive multifocal leukoencephalopathy (PML). Serial axial MRI images from 2023/08 to 2024/01 demonstrate progressive white matter changes.

Literature Review: JC virus-related PML is a rare and often fatal disease primarily seen in immunocompromised patients. [1.] Iatrogenic PML has been associated with rituximab use in lymphoproliferative disorders and occasionally in SLE. [2-3.] MRI typically shows hyperintense lesions on T2-weighted FLAIR involving subcortical and juxtacortical white matter. [1.] Immune reconstitution is the main treatment strategy due to limited efficacy of direct antiviral agents. [1-3.] **References** 1.Cortese I, Reich DS, Nath A. Progressive multifocal leukoencephalopathy and the spectrum of JC virus-related disease. Nat Rev Neurol. 2021;17:37-51. 2.Carson KR, Evens AM, Richey EA, et al. Progressive multifocal leukoencephalopathy after rituximab therapy in HIV-negative patients: A report of 57 cases. Blood. 2009;113:4834-4840. 3.Raisch DW, Rafi JA, Chen C, Bennett CL. Detection of cases of progressive multifocal leukoencephalopathy

associated with new biologicals and targeted cancer therapies. Expert Opin Drug Saf. 2016;15:1003-1011.

Discussion: This case highlights the difficulty in differentiating between NPSLE and rituximab-associated PML, both of which can present with similar neurological symptoms and imaging findings. While rituximab has proven effective in treating various autoimmune disorders, its association with PML raises concerns, especially in long-term immunosuppressed SLE patients. [3.] This case underlines the need for vigilance in monitoring such patients, as early recognition and intervention are key to managing PML. Clinicians should consider a careful balance between treating NPSLE and minimizing the risk of JC virus reactivation, especially in high-risk individuals undergoing rituximab therapy.

PV284 / #590

Case Report Poster Topic: *AS05 - CNS Lupus*

LOSING SIGHT OF THE DIAGNOSIS: A CASE REPORT OF NEUROMYELITIS OPTICA SPECTRUM DISEASE AND SYSTEMIC LUPUS ERYTHEMATOSUS-NEUROAUTOIMMUNITY IN FOCUS, CORRELATION OR COINCIDENCE?

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Introduction: Optic Neuritis is a rare, albeit, severe manifestation of Systemic Lupus Erythematosus (SLE). It causes immune-mediated inflammation in the central nervous system (CNS), leading to demyelination and vision loss. It is also a prominent feature of Neuromyelitis Optica Spectrum Disease (NMOSD), which can affect both the optic nerves and the spinal cord. These two conditions share overlapping traits, causing neurologic manifestations that complicate the diagnosis and may require distinct therapeutic approach, especially in refractory cases.[1]

Case Presentation With Investigation: A 64-year-old female with a medical history of arterial hypertension and mild thrombocytopenia, first noted four years ago, presented with progressively deteriorating vision loss that began two months before her visit. Ophthalmological examination revealed severe loss of visual acuity: only light perception in the right eye and finger counting at 2 meters in the left, with no signs of inflammation or macular edema on fundoscopy. Brain CT and CT-A revealed no significant abnormalities other than a mild narrowing of the right internal carotid artery (22%). Clinical examination, aside from vision loss, was unremarkable. Laboratory findings revealed mild thrombocytopenia (PLT: 80×10^3), normal inflammatory markers (ESR:18 mm/1st h; CRP:1.8 mg/L), urinalysis and biochemical panel. T2-weighted brain MRI demonstrated mild enhancement of the right optic nerve near the optic canal, consistent with optic neuritis (Image 1). Cerebrospinal fluid analysis showed normal cell count. Serologic testing revealed elevated anti-aquaporin-4 antibodies (32× ULN), hypocomplementemia (C3:73.7 mg/dL; C4:10 mg/dL) and high anti-ds-DNA binding (2× ULN). A diagnosis of coexistent NMOSD and SLE was established. Treatment with pulses of glucocorticoids and Rituximab led to mild visual improvement (left eye: 8/10; right eye: 2/10) and resolution of thrombocytopenia (PLT: 326×10^3). The patient received maintenance therapy with Rituximab plus Azathioprine and was tapered of steroids succesfully. No relapses were observed over a two-year follow-up period. Repeat brain MRI showed atrophy of the right optic nerve without active inflammation or new lesions (Image 2). Image 1: T2-weighted axial orbital brain MRI (fat suppression)

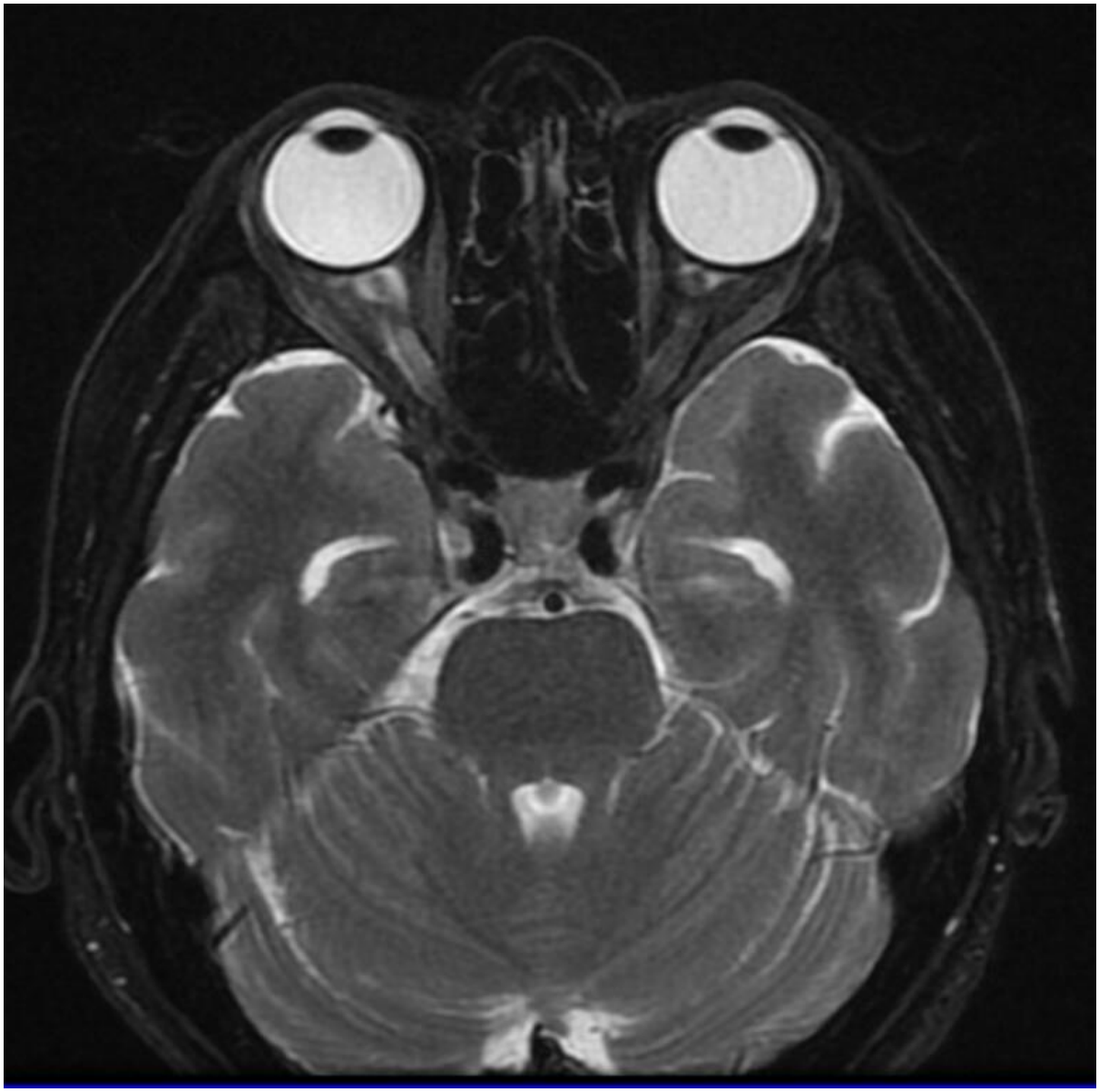
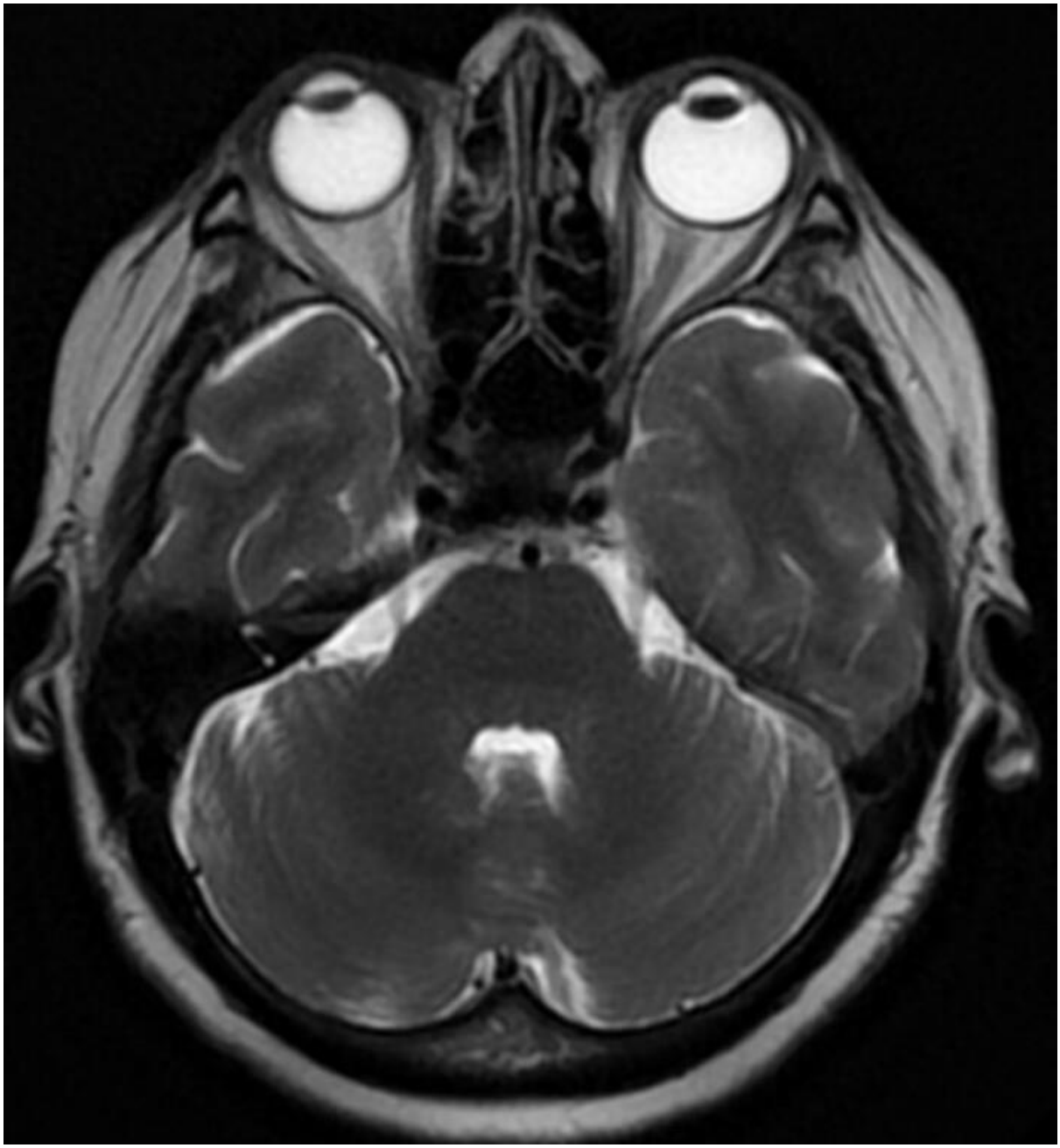


Image 2: T2-weighted axial brain MRI



Consent for publication obtained directly from patient.

Literature Review: Optic neuritis, as a manifestation of NMOSD, is associated with systemic autoimmune disorders such as SLE, and may also present alongside transverse myelitis. It is often linked with specific antibodies against Aquaporin-4 (AQP-4) or against Myelin-Oligodendrocyte-Glycoprotein (MOG).[1] NMOSD presents with longitudinally extensive myelitis lesions and lacks systemic involvement, which is commonly seen in SLE. Both conditions require treatment with high doses of glucocorticoids and immunosuppressive therapy, but first-line treatments differ. First-line treatment for NMOSD includes biologic therapies such as Eculizumab (anti-C-5a), Inebilizumab (anti-CD19), Satralizumab (anti-Interleukin-6 receptor inhibitor) and

Plasma Exchange. In contrast, CNS SLE therapy includes cyclophosphamide and Rituximab in refractory cases.[1,2]

Discussion: This case highlights the complex coexistence of NMOSD and SLE, two distinct autoimmune diseases that may overlap. While optic neuritis is a common manifestation of NMOSD, its occurrence in SLE is rare, leading to diagnostic confusion. Early recognition and appropriate management are crucial for preventing irreversible organ damage and improving overall prognosis. Effective treatment hinges on identifying the predominant disease in each clinical setting and prompts further inquiry into the overlapping characteristics of these conditions. **References** 1 Ochi MGS et al.Lupus and NMOSD:The Blending of Humoral Autoimmunity.Case Rep Rheumatol.2020 Oct 15;2020:8820071 2 Adawi M et al.Systemic Lupus Erythematosus (SLE) Complicated by Neuromyelitis Optica (NMO-Devic's Disease):Clinic-Pathological Report and Review of the Literature.Clin Med Insights Case Rep.2014 Jun 2;7:41-7

PV285 / #617

Case Report Poster Topic: *AS07 - Cutaneous Lupus*

THE OCCURRENCE OF DISCOID LUPUS ERYTHROMATOSUS PROGRESSING TO SYSTEMIC LUPUS ERYTHEMATOSUS, WITH OVERLAPPING ANCA-ASSOCIATED VASCULITIS, AND RHEUMATOID ARTHRITIS PRESENTING AS PAPULAR-PURPURIC GLOVE AND SOCK SYNDROME

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Introduction: Chronic cutaneous lupus erythematosus (CCLE), particularly discoid lupus erythematosus (DLE), is usually the most disfiguring of the lupus erythematosus (LE) – specific skin lesions. It manifests with sharply-demarcated skin lesions that are photosensitive. While it physically causes scarring, systemic involvement is rare. In certain cases, however, DLE may be part of, or may progress to systemic lupus erythematosus (SLE). The occurrence of concomitant DLE and Rheumatoid Arthritis (RA), DLE and ANCA-Associated Vasculitis (AAV) have only been reported in a few cases. There are also more cases of overlap syndromes occurring in subacute and acute LE, rather than in the discoid subtype. The paucity of local data in the Philippines and Southeast Asia also adds to the diagnostic dilemma. Finally, the role of a viral infection prompting papular-purpuric glove and sock syndrome (PPGSS) in heralding a CTD remains to be determined.

Case Presentation With Investigation: We present the case of 28-year-old female, with known history of papular-purpuric glove and sock syndrome attributed to systemic viral infection occurring two years prior. She presented to the emergency room with a chief complaint of fever and rash. Accompanying symptoms included unintentional weight loss, joint pain, muscle pain, alopecia, with marked headache upon sun exposure, and photosensitive discoid rashes. PE showed scarring alopecia and generalized discoid rashes with raised, erythematosus plaques and adherent scales on the scalp, face, neck, cheeks, nose, ears, and upper lip. Both hands showed signs of active synovitis. Despite the presence of discoid lesions, and absence of the traditional acute cutaneous lesions, patient progressed to SLE. She presented with the following criteria: fever, hemolytic anemia with direct Coomb's positivity, oral ulcers, arthritis, and hypocomplementemia, satisfying the ACR/EULAR 2019 Criteria for SLE. Morning stiffness and hand joint deformities typical of RA prompted RF testing, which turned out to be seropositive. c-ANCA was likewise seropositive at 1:80. Infection and malignancy were ruled out. Patient was started on corticosteroids and conventional synthetic DMARDs which prompted resolution of fever, rash, and arthritis. While discoid skin lesions remain permanent, her disease activity is presently quiescent.

Literature Review: CCLE encompasses wide range of dermatologic manifestations which may or may not include systemic disease, and some cutaneous manifestations, these includes discoid lupus erythromatosus, lupus erythematosus profundus, chilblain cutaneous lupus, and lupus tumidus, which may incidentally include systemic disease that would provide clinical clues, signs, and symptoms for MCTD or an overlap syndrome. Early signs and symptoms may involve hands, fingers, fingertips. Although dermatologic manifestations are common and may occur at initial presentation, skin lesions vary in morphology like cutaneous nodules, macular eruptions, and ecchymoses. SLE and RA overlap, termed “Rhupus Syndrome” is rare, and is estimated to be prevalent in 1% of patients with RA, the occurrence of DLE and AAV also affects a range of internal organs which are managed with high-dose glucocorticoids, immunosuppressants and targeted biologic medications.

Discussion: While CCLE lesions usually remain limited to cutaneous involvement, vigilance should always be exercised due to its possible progression to SLE. Likewise, confirmation of SLE, if with concomitant symptoms typical of another CTD, should prompt further workup. In this case, our patient presented with an overlap syndrome, comprised of SLE, RA, and AAV. The role of a prior PPGSS remains unknown. Our case highlights the need to have high index of suspicion for overlapping syndromes, especially in The Philippines, which is developing and is resource-limited.

PV286 / #392

Case Report Poster Topic: *AS07 - Cutaneous Lupus*

A RARE PRESENTATION OF AN OVERLAP CONNECTIVITY AS PURPURA FULMINANS: A CASE REPORT

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Introduction: Autoimmunity, by its complexity, involves various lesional mechanisms both structurally and functionally. Its association with hemostasis disorders amplifies this damage and darkens the prognosis

Case Presentation With Investigation: We report a case of purpura fulminans revealing an overlapping syndrome associating dermatomyositis, systemic lupus with secondary antiphospholipid syndrome. Observation: A 56-year-old diabetic woman, hypertensive, followed for Hashimoto's thyroiditis is admitted for the management of a systemic picture made of: an inflammatory myopathy retained on the clinic, morphology and histopathology; a glomerular syndrome with on the kidney membranous-proliferative nephritis.The histopathological study of the skin biopsy is in favor of immune pauci leukocytoclastic vasculitis with multiple microthrombi.All of this is evolving in a febrile context. After excluding infectious causes, the etiological investigation is in favor of an overlapping syndrome: systemic lupus, dermatomyositis with secondary APS. The patient benefited from immunosuppressive treatment combined with plasmapheresis sessions and antithrombotic treatment with a favorable outcome. Conclusion: Purpura fulminans is a manifestation that is not limited to an infectious cause. He may be the witness of a dysimmune and/or hematological disorder.

Literature Review: Atypical presentation of purpura fulminans following sepsis in an adult Paul Lyon, Rabi Nambi, Faisal Faruqi BMJ Case Rep. 2011; 2011: bcr0320113996 - Infectious purpura fulminans J. Harikrishna, Alladi Mohan Indian J Med Res. 2015 Jan; 141(1): 130–131. BMJ Case Rep. 2011; 2011: bcr0320113996. Indian J Med Res. 2015 Jan; 141(1): 130–131.

Discussion: Purpura fulminans is a manifestation that is not limited to an infectious cause. He may be the witness of a dysimmune and/or hematological disorder.

PV287 / #776

Case Report Poster Topic: *AS07 - Cutaneous Lupus*
Late-Breaking Abstract

IMPORTANCE OF SEQUENTIAL FOLLOW-UP OF AUTOANTIBODIES IN ANA-POSITIVE PATIENTS WITH DIFFUSE HAIR LOSS

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Introduction: Diffuse hair loss can be an early manifestation of systemic lupus erythematosus (SLE), particularly when other common causes such as alopecia areata, drug-induced alopecia, iron deficiency, and androgenic alopecia are excluded. The presence of high-titer anti-nuclear antibodies (ANA) without immediate clinical signs of SLE necessitates close monitoring, as the disease may evolve over time. Sequential follow-up of autoantibodies in ANA-positive patients with unexplained hair loss is crucial for early detection and intervention.

Case Presentation With Investigation: A 20-year-old female initially presented to the dermatology department at CHA Bundang Medical Center on June 9, 2021, with diffuse hair loss and folliculitis. She had no prior treatment history and was prescribed topical minoxidil 5% and alfatradiol solution 0.025% for daily application. Laboratory tests, including CBC, U/A, routine chemistry, ESR, CRP, ANA, anti-dsDNA IgG, SS-A/Ro Ab, SS-B/La Ab, syphilis reagin test, serum ferritin, T3, fT4, TSH, free testosterone, DHEA-S, zinc, iron, and SHBG, revealed ANA positivity at 1:1250 (speckled pattern), fT4 of 0.85 ng/dL, and a zinc level of 61.09 µg/dL, with all other results within normal limits. No clinical evidence of polycystic ovary syndrome (PCOS), arthritis, or thyroid disease was noted. The patient was treated with topical minoxidil 5% and alfatradiol solution 0.025% for five months, along with oral polaprezinc twice daily. Due to gastrointestinal side effects, polaprezinc was discontinued, and dietary zinc intake was encouraged. Despite eight months of topical treatment, hair regrowth was inadequate, leading to the addition of low-dose oral minoxidil (5 mg/day) for eight months. Over time, serial autoantibody testing revealed progression to positive anti-dsDNA IgG (>150), ANA 1:2560 (speckled pattern), and anti-Sm Ab positivity. The patient also developed proteinuria (urine protein 323 mg/24 hours), multiple arthralgia, fatigue, and fever. A kidney biopsy confirmed focal lupus nephritis (Class III), leading to a definitive diagnosis of SLE. She was initiated on mycophenolate mofetil (2 g/day), hydroxychloroquine, and low-dose prednisolone (5 mg/day). Following treatment, arthralgia resolved, proteinuria decreased (323 mg to 30 mg/24 hours), and anti-dsDNA IgG levels significantly declined (from 379 to 6).

Literature Review: Non-scarring alopecia is a common and reversible manifestation in SLE, primarily driven by immune-mediated inflammation, hair cycle disruption, vascular dysfunction, oxidative stress, hormonal dysregulation, and drug-induced effects. 1)

Immune-Mediated Inflammation: Autoantibodies and immune complexes in SLE contribute to follicular damage through increased IFN-α signaling, CD4+ T-cell and B-cell infiltration, and microvascular injury. 2) Alterations in the Hair Cycle: Inflammatory stress promotes telogen effluvium by shifting follicles to the resting phase, while anagen arrest leads to premature shedding and impaired regrowth. 3) Vascular Damage & Oxidative Stress: Vasculitis-induced endothelial damage compromises scalp microvasculature, reducing blood flow. Oxidative stress further exacerbates follicular cell apoptosis. 4) Hormonal & Metabolic Dysregulation: Chronic inflammation activates the HPA axis, increasing cortisol levels that inhibit hair cycling. Autoimmune thyroid disorders, such as Hashimoto's thyroiditis, contribute to hair thinning. 5) Drug-Induced Hair Loss: Medications like hydroxychloroquine, methotrexate, mycophenolate mofetil, and corticosteroids can trigger telogen effluvium, disrupting normal follicular cycling.

Discussion: Regular follow-up of autoantibodies in ANA-positive patients with diffuse hair loss is essential for early SLE detection. Monitoring ANA, anti-dsDNA, and lupus-specific antibodies aids in timely intervention, improving patient outcomes. Early recognition and surveillance facilitate prompt diagnosis and management, reducing disease progression risks.

PV288 / #731

Case Report Poster Topic: *AS09 - Emerging Approaches in SLE Management*
Late-Breaking Abstract

CONSULTATIVE, PROACTIVE PHYSICAL THERAPY FOR PEOPLE WITH LUPUS: A CASE REPORT

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Introduction: Physical therapy (PT) can facilitate increased exercise through education, an individualized exercise routine, goal setting, and supportive resources. However, traditional PT focuses on function restoration rather than proactive measures to prevent the advancement of morbidity. We present the application of a proactive physical therapy (PAPT) approach for patients with SLE.

Case Presentation With Investigation: Four patients with SLE were referred to PT by a Rheumatologist shortly after diagnosis. Patients 1) completed a PT evaluation of their current functional level including gait speed measured by the 10-meter walk test (10mwt), functional exercise capacity measured by the 6-minute walk test (6mwt), functional strength measured by the 5x sit-to-stand (5xSTS), balance measured by the MiniBESTest, 2) received education on aerobic and strengthening exercise guidelines, 3) received goal setting support, and 4) established an exercise routine at an appropriate intensity using shared decision-making. Additional assessments included the fatigue severity scale (FSS), pain detect questionnaire, Godin leisure time questionnaire, and self-reported exercise behavior. Patients were seen at baseline, week 2, week 6, week 10, week 16, and week 24. Aerobic exercise was introduced at baseline, and strength exercise was introduced at week 2. Each week the exercise prescription progressed with the goal of meeting recommended exercise guidelines by week 16. All outcomes were reassessed at week 24. At week 24, two patients reported regular aerobic and strengthening participation that met or exceeded exercise guidelines. Both patients demonstrated improvements in gait and functional mobility measured by functional outcome measures. P1 demonstrated a 0.09m/s increase in her 10mwt SSV, no change in her 10mwt FV, a 33 meter increase in her distance walked in the 6mwt, 3.23 second improvement in the 5xSTS, and remained stable in her balance on the MiniBESTest. P2 demonstrated a 0.28m/s increase in her 10mwt SSV, 0.58m/s increase in her 10mwt FV, indicating clinically meaningful change. She also demonstrated a 35 meter increase in her distance walked in the 6mwt, 0.87 second improvement in the 5x STS, and remained stable in her balance on the MiniBESTest. P1 reported improvement on her FSS from 61 to 34, and P2 improved from 51 to 36, indicating significant improvement in fatigue. P1

improved her score on the pain detect from 17/35 to 11/35, and P2 improved from 25/35 to 12/35, indicating clinically meaningful improvements in self-reported pain. P1 reported improved self-reported exercise minutes via the Godin-leisure-time questionnaire from 36 to 37, and P2 improved from 12 to 85, indicating significant improvement in time spent exercising. One patient was unable to schedule her evaluation secondary to personal issues and one patient was lost to follow-up after week 10.

Literature Review: Physical activity is an evidence-based modifiable lifestyle behavior that helps manage symptoms of SLE. Specifically, exercise training may improve cardiovascular capacity and physical function and decrease fatigue in people with systemic lupus erythematosus (SLE) without symptom exacerbation. Unfortunately, 60-72% of people with SLE are not sufficiently physically active (<150 minutes of moderate-to-vigorous physical activity (MVPA)/week per WHO guidelines). Efforts to improve physical activity levels of people with SLE are sorely needed to optimize physical health and functioning.

Discussion: A PAPT model of care is feasible and effective for patients newly diagnosed with lupus. Patients met established exercise guidelines and maintained or improved physical function. Modifications to the number of visits may be considered to improve follow-through.

PV289 / #142

Case Report Poster Topic: *AS14 - Innate Immunity*

ASSOCIATION OF SYSTEMIC LUPUS ERYTHEMATOSUS AND CHRONIC GRANULOMATOUS DISEASE IN ADULTS: REPORT OF TWO CASES

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Introduction: Chronic granulomatous disease (CGD) is a genetic immunodeficiency disorder characterized by recurrent bacterial and fungal infections. CGD can manifest in various ways, ranging from mild infections to potentially life-threatening complications. The association between CGD and systemic lupus erythematosus (SLE) is rare.We report the cases of two female patients with CGD who developed manifestations of systemic lupus erythematosus.

Case Presentation With Investigation: Observation 1:This case involves a 38-year-old female patient from a consanguineous marriage. Her medical history included multiple episodes of urinary infections with catalase-positive organisms since the age of 18, recurrent pulmonary infections, and a bacterial meningitis episode at the age of 32. In the course of the etiological assessment of these infections, a nitroblue tetrazolium (NBT) test showed collapsed oxidative activity in neutrophils. Therefore, the patient was diagnosed with chronic granulomatous disease. Six years later, at the age of 38 years, she presented with a sudden onset of painful, erythematous papules and nodules on her face, forearms, and legs, accompanied by a fever of 39°C. A skin biopsy revealed neutrophilic dermatitis, suggestive of Sweet syndrome. She also exhibited joint involvement in the form of chronic nondeforming bilateral polyarthritis, significant pericardial effusion, and moderate bilateral pleurisy. Laboratory tests revealed positive antinuclear antibodies at 1/1280 with a homogeneous pattern, positive anti-DNA antibodies at 40, positive anti-nucleosome antibodies, and consumption of C3 and C4 complement levels. A diagnosis of SLE, revealed by Sweet's syndrome, was established, with hematological, articular, and serosal involvement. Hydroxychloroquine was not initiated because of visual field impairment, but the patient was administered corticosteroid therapy with good clinical and biological evolution. Observation 2:This case involved a 23-year-old female patient, also from a consanguineous marriage, who had been followed up for CGD since the age of 6. Her medical history included tuberculous adenitis at age 4 years, pulmonary aspergillosis at age 6 years, and osseous aspergillosis at age 8 years. The NBT test showed no oxidative activity of neutrophils, and she was placed on prophylactic treatment with sulfamethoxazole/trimethoprim. At age 20 years, the patient experienced deep vein thrombosis of the right lower limb, for which she was treated with low-molecular-weight heparin, followed by anticoagulation with a vitamin K antagonist. Two years later, the

patient presented with a sudden decrease in visual acuity, revealing occlusion of the central retinal artery and vein. The antiphospholipid syndrome workup revealed positive anti-cardiolipin IgG antibodies on two occasions, along with the presence of circulating lupus anticoagulant. A few months later, she presented with malar rash, photosensitivity, and finger chilblains. Laboratory tests showed positive antinuclear antibodies at 1/640, anti-DNA antibodies, anti-Sm antibodies, and consumption of C3 and C4 complement levels. SLE was diagnosed, and hydroxychloroquine was started with good clinical outcomes.

Literature Review: An association between CGD and autoimmune diseases has been rarely observed, but this relationship is poorly understood. Insufficient bactericidal activity may lead to chronic antigenic stimulation, which could explain the development of autoimmune diseases. Another hypothesis is that this association may result from T cell dysregulation and an inflammatory state characterized by excessive interleukin-7 production, particularly in cases of pulmonary aspergillosis.

Discussion: Although the association between CGD and SLE is rare, it underscores the importance of thorough clinical evaluation of patients with recurrent infections and autoimmune symptoms. Recognizing this association can be crucial for diagnosis and therapeutic management, as it may influence the choice of immunosuppressive treatment and monitoring of infectious complications.

PV290 / #641

Case Report Poster Topic: *AS17 - Miscellaneous*

THE UNMET NEEDS OF LUPUS PATIENTS: CREATING VISIBILITY AND SUPPORTING PATIENTS AND FAMILIES IN THEIR JOURNEY

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Introduction: Right from the first symptom, through diagnosis and coping with the many phases of living with lupus, this chronic autoimmune disease has proven to be a lonely journey that most patients thread. With three rheumatologists to a population of about 31 million, the huge provider to patient gap in the workforce necessitates the need for interdisciplinary and collaborative efforts from both clinical and non-clinical personnel to meet the demand for holistic care of not only patients, but their caregivers, families, loved ones and society at large.

Case Presentation With Investigation: Over the years, the increasing report of autoimmune conditions, particularly lupus, in Ghana, and the continent of Africa at large, suggests their ubiquity and not rareness as was previously perceived. The autoimmune disease landscape, as studies reveal, is complex, almost other-worldly in contexts with dearth of information. Lupus has been a mysterious silent killer in Ghana. It is significantly difficult to get any relevant information required to inform policy as well as garner support for lupus patients.

Literature Review: In Ghana, this situation is further compounded by challenges such as misdiagnosis, delayed referrals, economic challenges and sociocultural beliefs. The labyrinth of accessing the needed healthcare, lengthy process of obtaining clinical diagnoses and its ripple effect on the quality of life, finances and social life in themselves, often overwhelms many patients and pushes them to the edge of despair. Moreover, the lack of awareness among relevant stakeholders and the public further exacerbates the challenges that exist. Acknowledgement of these challenges has been the reason for the work of Oyemam Autoimmune Foundation (OYEMAM) in creating visibility about lupus and raising support for those affected directly and indirectly through its flagship lupus awareness campaign since 2016 in Ghana.

Discussion: The Foundation, which is a patient-led registered non-profit has remained committed to inspiring hope through advocacy with policymakers and other relevant stakeholders; education and raising awareness through the media, engaging with diverse audiences in-person and virtually, as well as providing support for people living with lupus and other autoimmune conditions. OYEMAM has been deliberate about the

counselling needs for those affected, recognizing this often-overlooked burden and addressing it in many little ways has brough much relief and hope to many patients and their families. This paper adopts a walkthrough approach to examine the strategies of OYEMAM's interventions by presenting the lived experiences of those we have engaged with and some of the impact made so far. We acknowledge patients and all who have and continue to contact OYEMAM giving us a reason to strive on and contribute to this misunderstood and underserved aspect of human life and health.

PV291 / #290

Case Report Poster Topic: *AS17 - Miscellaneous*

FUNGUS BALL, AN HEMOPTYSIS CAUSE IN A SLE PATIENT

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Introduction: A fungus ball (mycetoma, aspergilloma) is a conglomeration of fungal hyphae intermingled with mucus and cellular debris, typically found within a pulmonary cavity. The most common cause is the colonization of pre-existing fibrocavitary diseases, resulting from healed tuberculosis. Cavitary tuberculosis is relatively uncommon in individuals with systemic lupus erythematosus (SLE), and only few cases of fungus ball formation in tuberculosis cavities in these patients have been reported.

Case Presentation With Investigation: A 48-year-old man was diagnosed with SLE and antiphospholipid syndrome in 2007 based on the presence of positive antinuclear and anti-double stranded DNA antibodies, immunoglobulin G and M anticardiolipin antibodies, oral ulcers, lymphopenia, non-erosive polyarthritis, class-IV lupus nephritis, and a pulmonary embolism. His treatment included hydroxychloroquine, high-dose oral prednisolone (which was gradually tapered to a lower dose after achieving remission), and monthly intravenous cyclophosphamide up to a total of 7 grams, followed by azathioprine 100 mg/ day. In 2009, he developed disseminated tuberculosis (pulmonary and gastrointestinal compromise), for which he completed a 12-month anti-tuberculosis treatment course. Fifteen years later, he was admitted due to recurring hemoptysis. Laboratory and imaging tests revealed native mitral valve endocarditis caused by Moraxella bovis. Chest imaging showed a mobile mass within a cavity in the left upper lobe (Figure). At that time, he was treated with warfarin, hydroxychloroquine 200 mg/day, and prednisolone 5 mg/day, with no signs of lupus activity (SLEDAI 0). The patient underwent surgery and histopathological studies confirmed the presence of a fungus ball due to Aspergillus flavus.

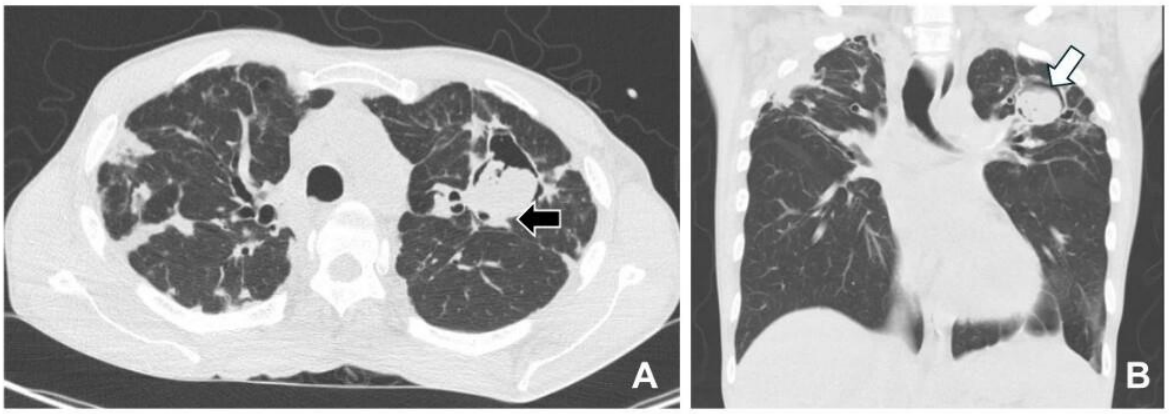

Figure. A. Contrast-enhanced axial cranial tomography (CT) of chest: Black arrow points an oval shape mass at the dependent region of a cavity (Monod sign). B. Contrast-enhanced coronal cranial tomography (CT) of chest: Fungus ball with air crescent sign (white arrow)

Literature Review: The development of an aspergilloma usually occurs silently and is often asymptomatic; however, patients may experience chronic cough and hemoptysis. Radiologically, it typically appears as a mobile mass within a cavity to a gravity-dependent position frequently located in upper lobes (Monod sign). It may also show an "air crescent sign" (Figure) which indicates a separation between the fungus ball and the cavity wall, either partially or completely, and it is also observed in cases of pulmonary necrosis, pulmonary tuberculosis, hydatid cysts, Rasmussen aneurysm, and lung carcinoma. Due to its frequent surroundings of scarred and fibrotic lung tissue, the lesion can be challenging to detect on standard chest X-rays, making CT scans essential for confirmation. Treatment has not been standardized and some aspects remain controversial due to the unpredictable nature of the infection. Surgical resection is considered the gold standard for management, and perioperative or postoperative antifungal therapy with triazoles is recommended in patients with a high risk of surgical spillage.

Discussion: Impaired host immunity is considered a risk factor for postprimary tuberculosis. Patients with SLE often receive high doses of glucocorticoids and immunosuppressives, which can lead to reduced cellular immunity, thus increasing the risk of active tuberculosis. Aspergilloma formation can also occur in SLE patients within the same cavity. CT scans are a reliable diagnostic method for confirming this condition. Air crescent and Monod signs are distinctive features of aspergilloma, the latter helping to differentiate the fungal ball from other conditions and also helps in differentiating between causes of hemoptysis in SLE patients (e.g. disease activity, infection, pulmonary embolism), thereby preventing unnecessary or invasive diagnostic measures and reducing the risk of potentially life-threatening complications.

PV292 / #338

Case Report Poster Topic: *AS17 - Miscellaneous*

UNUSUAL PRESENTATION OF LUPUS ENTERITIS WITH COLORECTAL INVOLVEMENT: A RARE CASE REPORT IN BANGLADESH

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Introduction: Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease of young that can affect any organs of the body. About 40 percent of SLE patients have gastrointestinal issues during their lifetime. Lupus enteritis (LE) may be a serious complication carries a high risk of mortality. It usually involves the mesenteric arteries causing ischemic changes of the small bowels but rarely involves large bowels, especially to the colon and rectum.

Case Presentation With Investigation: Here we presented a 17-year-old girl who is a known case of SLE, diagnosed with colorectal LE after initial presentation with intermittent abdominal pain, vomiting and occasional diarrhea for two months at Rheumatology department of BIRDEM General Hospital, Dhaka, Bangladesh on 29th September 2024. During her hospital course, she had mid abdominal pain for 3 days, which was sudden, severe, colicky, had no aggravating or relieving factors, no-radiation, associated with vomiting & bloody diarrhea. She didn't give any history of hemoptysis, weight loss or fever. One year back, she was diagnosed as SLE on the basis of polyarthritis, oral ulcer, photosensitivity, excessive hair loss and skin rash with positive ANA and Anti dsDNA. She was on hydroxychloroquine, low dose prednisolone and methotrexate. But she had been irregular in taking medications, which may have contributed to the onset of LE and led to her current hospital admission. According to the 2019 European Alliance of Associations for Rheumatology (EULAR)/American College of Rheumatology (ACR) Classification criteria for SLE her score was 22. Additionally, abdominal computed tomography (CT) and CT angiogram of abdominal aorta with other relevant investigations revealed distended bowel loops, significant wall thickening and luminal narrowing with hallow sign at intestinal wall especially proximal rectum including rectosigmoid junction. Increased echogenicity surrounding the bowel loops with engorged mesenteric vessels and mild to moderate ascites was found. All these findings were consistent with colorectal LE. Laboratory tests also showed lower levels of complement C3 and C4, with a very high titer (1:800) of anti dsDNA. Overall, it was clear that this case involved colorectum, representing a rare manifestation of lupus gut. The patient received treatment with hydroxychloroquine, intravenous pulse methylprednisolone followed by high dose oral prednisolone and pulse cyclophosphamide along with nutritional support. After one week of treatment her condition improved significantly.

Literature Review: SLE can affect the entire GI tract, from the oral mucosa to the rectum. The percent of patients of GI tract related symptom is up to 40%-50% but the development of LE is present in only about 0.2% to 5.8% of patients. LE involving the jejunum and ileum (83% and 84%, respectively) are relatively common, but involvement of the colon (19%) and rectum (4%) with or without involvement of the small intestine is extremely rare.

Discussion: Lupus enteritis generally affects the small intestine, but in rare cases, it can involve the colon presenting an unusual and diagnostically challenging manifestation. Colorectal involvement may lead to symptoms like abdominal pain and bloody stools which can mimic inflammatory bowel disease or infectious colitis. Recognizing atypical presentation is crucial, as early treatment with corticosteroids and immunosuppressants can prevent complications including ischemia and perforation, associated with colonic involvement.

PV293 / #569

Case Report Poster Topic: *AS18 - Paediatric SLE*

GASTROINTESTINAL TUBERCULOSIS PRESENTING AS INTESTINAL OBSTRUCTION IN A 16-YEAR-OLD FILIPINO WITH PEDIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS

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Introduction: Systemic lupus erythematosus (SLE) is a systemic disease, which can affect multiple organ systems. Gastrointestinal (GI) involvement is rare in pediatric SLE and is under-recognized. On the other hand, gastrointestinal tuberculosis (TB) is also a rare disease that can be easily missed clinically. Consequently, intestinal pseudo-obstruction in SLE and GI TB are both rare diseases and usually misdiagnosed, which may result in delay in treatment.

Case Presentation With Investigation: A 16-year-old Filipino female diagnosed with SLE one month prior to onset of GI symptoms. She presented with prolonged fever, joint pains, malar rash, weight loss, easy fatigability, increased hair fall, and positive antinuclear antibody (ANA) and anti-double stranded DNA. She was also diagnosed with latent TB infection that time due to more than 10-millimeter (mm) induration on tuberculin skin test. She was subsequently started on isoniazid, rifampicin, hydroxychloroquine, and oral corticosteroids. She had generalized abdominal pain, nausea, and vomiting one month from SLE diagnosis. Whole abdominal computed tomography (CT) scan showed enhancing circumferential wall thickening involving the terminal ileum with subsequent proximal dilatation of the more proximal segments with possible reactive cecal and proximal ascending colon involvement. She underwent emergency exploratory laparotomy, segmental ileocecal resection, and double barrel ileocolostomy. The histopathologic findings showed the ileocecal segment with chronic granulomatous inflammation, ulceration, abscess formation, and acute serositis, consistent with ileocecal TB. She was discharged improved and with complete resolution of GI symptoms.

Literature Review: Gastrointestinal tuberculosis (TB) accounts for 1% to 3 % of all TB cases worldwide. It can occur in the context of active pulmonary disease or as a primary infection without pulmonary involvement. Diagnosis is challenging and is often delayed due to its non-specific presentation. The terminal ileum and ileocecal valve are the commonly involved segments. Differentiating the symptoms of IPO and GI TB can be difficult, as both have similar non-specific symptoms such as nausea, vomiting, abdominal pain, abdominal distention, and anorexia. Acute abdomen in patients with SLE is a challenging diagnostic and therapeutic problem. Medications such as steroids and immunosuppressive drugs, mask the physical findings of obstruction, perforation,

and ischemia. Thus, early recognition and prompt management for both entities are lifesaving.

Discussion: The incidence and prevalence of GI involvement in SLE vary widely – this could be due to less attention being paid to GI manifestations than other organ symptoms. The most prevalent GI symptoms are non-specific and include nausea, vomiting, anorexia, and abdominal pain. Our patient was on prednisone at 1mg/kg/day and hydroxychloroquine which could have masked the features of GI perforation and ischemia. IPO can be diagnosed based on clinical manifestations, physical findings, and radiological examinations such as abdominal radiography and CT.
Both disease entities, gastrointestinal tuberculosis and SLE IPO, is a diagnostic challenge as both typically presents with non-specific clinical and radiologic features. The diagnosis is often delayed due to its vague presentation resulting in various complications. A high index of clinical suspicion and appropriate use of various investigative modalities can aid in early diagnosis, thereby reducing associated morbidity and mortality.

PV294 / #139

Case Report Poster Topic: *AS19 - Patient-Reported Outcome Measures*

2 SIDES OF LUPUS-CASE REPORT

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Introduction: Systemic lupus erythematosus (SLE) is a complex disease, often described as the "disease with 1000 faces". Its impact on the various organs and systems has been thoroughly described, however we believe that it is important to assess the impact it has on patients that are also physicians-on their professional activity, quality of life and relation towards fellow patients.

Case Presentation With Investigation: My first symptoms, consisting of lower limb rash, fatigue and pancytopenia, appeared around the age of 16. Subsequent investigations suggested an autoimmune disease and although there was no clear diagnosis I received intensive immunosuppression (high dose oral steroids, iv cyclophosphamide and mycophenolate) which only partially improved the symptoms and caused significant side effects (nausea, vomiting, diarrhea, frequent upper respiratory and urinary infections, menstrual cycle abnormalities), thus leading to educational deficits and isolation from peers. Over the next few years treatment was reassessed, maintaining only low dose prednisone and hydroxychloroquine (HCQ), which was enough at that point to control the disease and enabling me to start studying medicine. The first five years were mostly uneventful, except for the occasional rash, lower limb oedema and intense itching from HCQ. Afterwards I started experiencing joint pain (hips, knees, Costo-vertebral, fingers) as well as myalgia and muscle spasms, which lead to extensive use of NSAIDS. Finally in 2012 (the year I graduated) I received the definitive diagnosis of SLE due to positive serology. In addition to steroids and HCQ I was started on azathioprine, which seemed to improve symptoms during the first few months but then was less effective as I started working night shifts as an internal medicine resident. In my second year of residency, I quit these shifts and took part in a study on Belimumab (Benlysta)- being effective both on symptoms and laboratory results. After the study ended, I continued on azathioprine, unfortunately leading to severe neutropenia. For the next two years I took only oral vitamin D until a severe flare in 2017 when I was started on sc methotrexate, which lead to anal fissures and a very poor quality of life. Next year (my first as a specialist physician) consisted of extensive joint involvement not controlled by low-dose prednisone. In 2019 Benlysta was finally approved in Romania and I started infusions, finally obtaining remission of symptoms. Unfortunately, my kidney function began deteriorating, biopsy showing class III lupus nephritis. With add-on mycophenolate, HCQ and ACE inhibitor, creatinine and

proteinuria decreased, having stable values to the present time. This also determined me to pursue nephrology as a second specialty.

Literature Review: No relevant information

Discussion: Dealing with SLE from both perspectives (patient and doctor) allowed me to better understand all the implications of this disease-on one's career, family and relationships, self-esteem and overall quality of life, helping me to have a more holistic approach of each case and offer a qualitative care for my patients.

PV295 / #313

Case Report Poster Topic: *AS22 - SLE Heterogeneity*

BULLOUS LUPUS,AN UNCOMMON CAUSE OF ATYPICAL ESOPHAGITIS:A CASE REPORT.

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Introduction: Systemic lupus is a condition that continually reveals its complexities,both systemically and dermatologicaly.Bullous lupus which accounts for less than 5%,poses a true diagnostic and therapeutic challenge.We report a case of bullous lupus with severe esophageal involvement.

Case Presentation With Investigation: Observation: A 42 years old woman with no significant medical history apart from an esophagitis resistant to treatment for two years, presented with dysphagia. An upper digestive endoscopy revealed multiple esophageal strictures with fragile mucous membrane which bleed on contact with the endoscope.This esophagitis was resistant to proton pump inhibitor treatment.Histological examination of the esophageal biopsies suggested a fleshy polyp without evidence of eosinophilic infiltration or tuberculoid granuloma.The immunological assessment(ANA,ANCA,anti-CCP,rheumatoid factor,antiphospholipid antibodies and celia serology)was negative.Additionnally,the patient exhibited fragile skin,oral ulcers and some vesicular and crusty lesions in sunexposed areas.Histological study and direct immunofluorescence of the skin biopsy found an epidermolysis with antiC1q of IgM and IgG type;and C3-C4 complement fraction deposits; which argues for the diagnosis of bullous lupus with lupus esophageal involvement.The patient was placed on immunosuppressive treatment(corticosteroids and azathioprine),with favorable clinical outcomes.

Literature Review: Bullous systemic lupus erythematosus in females Grant Sprow, BAa,b, Mohsen Afarideh, MD, MPHa,b, Joshua Dan, BAa,b, Matthew L. Hedberg, MD, PhDa, Victoria P. Werth, MDa,b, International Journal of Women's Dermatology (2022) **Lupus erythematosus**-specific **bullous** lesions. Smith KN, Maddy AJ, Motaparthi K.Dermatol Online J. 2023 Dec 15;29(6).

Discussion: Bullous lupus is an extremely rare entity,and its esophageal involvement complicates diagnosis.Although its therapeutic management may seem straightforward,it remains a significant challenge for practitioners.

PV296 / #329

Case Report Poster Topic: *AS22 - SLE Heterogeneity*

THE SECRET OF C3 GLOMERULOPATHY IN A LUPUS PATIENT

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Introduction: Objective Nephrological abnormalities in a patient with SLE are not exclusive to lupus nephropathy, but can reveal unexpected causes, as is the case in our patient.

Case Presentation With Investigation: Method Patient followed for an SLE whose histopathological study of the renal biopsy puncture objective an isolated C3 glomerulopathy. Results A 48 years old man followed for articular and haematological systemic lupus since the age of 19 years,presented persistent proteinuria with hematuria and consumption of complement. The histopathology of renal biopsy puncture,was in favour of a C3 glomerulopathy with active lesions.The exploration of this entity found a low third component of complement(C3:0.8g/l)and a monoclonal component of the IgG Lambda type. Glomerular filtration rate was normal(80ml/mn).The diagnosis of monoclonal gammopathy of renal significance is more likely. Conclusion The SLE is a provider of nephropathy but the slightest atypia should alarm the clinician and the anatomical pathologist in order to broaden their diagnostic horizons

Literature Review: -C3 Glomerulopathy: Pathogenesis and Treatment Syeda Behjat Ahmad and Andrew S. Bomback Adv Chronic Kidney Dis. 2020;27(2):104-110 C3 glomerulopathy: Understanding an ultra-rare complement[1]mediated renal disease Amanda K. Heiderscheit, Jill J. Hauer Richard J. H. Smith Am J Med Genet. 2022;190C:344–357.

Discussion: Conclusion The SLE is a provider of nephropathy but the slightest atypia should alarm the clinician and the anatomical pathologist in order to broaden their diagnostic horizons

PV297 / #642

Case Report Poster Topic: *AS22 - SLE Heterogeneity*

MIXED CONNECTIVE TISSUE DISEASE EVOLVING FROM THE SEQUENTIAL OVERLAP OF SYSTEMIC LUPUS ERYTHEMATOSUS, SJOGREN'S SYNDROME, RHEUMATOID ARTHRITIS AND DERMATOMYOSITIS: A FOLLOW-UP

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Introduction: Mixed Connective Tissue Disease (MCTD) is a rare autoimmune disease, generally described as having overlapping features of at least two connective tissue diseases. Anti-U1-RNP, in high titers, is distinctly associated with such. Observational studies have reported sequential evolution of the connective tissue diseases. We aim to present a case of MCTD, with the sequential evolution of Systemic Lupus Erythematosus, Sjogren's Syndrome, Rheumatoid Arthritis, followed by Amyopathic Dermatomyositis.

Case Presentation With Investigation: She presented to the emergency room with fatigue, high-grade fever, cough, and myalgia. Physical examination revealed violaceous rash on both eyelids (heliotrope rash), erythematous rashes on her upper chest (V Sign) and back (Shawl Sign). Serial manual muscle tests were 5/5 on all extremities. Creatine kinase were normal. A diagnosis of Amyopathic Dermatomyositis was made. She concomitantly developed cough, with computed tomography scan showing features of honeycombing, consistent with usual interstitial pneumonia. Treatment armamentarium comprised of corticosteroids, conventional synthetic DMARDs, & nintedanib. We present a case of a 32 year old Filipino female whose initial manifestations occurred seven years prior. Fever, alopecia, arthritis, and hypocomplementemia with high-titre ANA (1:160, Speckled) and Anti-Smith seropositivity (592.5 U/mL), fulfilled the 2019 EULAR/ACR Criteria for SLE. Six years prior, she presented with dry eyes, dry mouth, with otorhinolaryngologic symptoms of lip mucocele and recurrent tonsillitis. Serologies revealed high-titre Anti-SSA 173.4 U/mL and Anti-SSB 24.6 U/mL; symptoms were consistent with Secondary Sjogren's. Few months later, she reported morning stiffness with chronic hand joint pains. Physical examination by a rheumatologist revealed symmetric arthritis involving the hand joints; with concomitant seropositivity of Rheumatoid Factor. A diagnosis of Seropositive Rheumatoid Arthritis was made, in concordance with the 2010 ACR/EULAR Criteria.

Literature Review: Mixed connective tissue disease (MCTD) is a rare systemic autoimmune disease which presents with at least two overlapping connect tissue diseases. Among the disease included - systemic lupus erythematosus (SLE), Sjogren's

syndrome, systemic sclerosis, dermatomyositis, polymyositis and rheumatoid arthritis. Interstitial lung disease may also be involve as a result of a complication of the MCTD and is responsible for significant morbidity. Amyopathic dermatomyositis (ADM) is a clinical subtype of dermatomyositis, presents with dermatologic lesions of the dermatomyositis but lacks the myopathic. In addition to the symptoms of Raynaud syndrome, arthritis, myositis and pulmonary hypertension among others with a high anti-U1 RNP antibody titers. A hallmark of the disease is the presence of Anti-U1 ribonucleoprotein (RNP). This may have a prognostic value, titer levels may be associated with prognosis of MCTD. Since MCTD has no unique clinical features, diagnosis may be challenging. Overall goals of therapy are to control symptoms, reduce risk for future diseases.

Discussion: The sequential overlap of connective tissue diseases is rarely reported. Her constellation of symptoms is consistent with MCTD, SLE being the initial autoimmune disease. Despite conferring a better prognosis, manifestations such as ILD may be more common in overlap syndromes; hence, must be vigilantly monitored.

PV298 / #813

Case Report Poster Topic: *AS23 - SLE-Diagnosis, Manifestations, & Outcomes*
Late-Breaking Abstract

WHEN SILENCE TAKES OVER: A CASE OF CATATONIC SYNDROME REVEALING SYSTEMIC LUPUS ERYTHEMATOSUS.

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Introduction: Systemic lupus erythematosus (SLE) is a complex and potentially serious autoimmune disease. Catatonia is a syndrome characterized by physical and behavioural abnormalities, which can result from psychiatric, neurological, or medical disorders. Attributing catatonia to SLE remains a challenge. Although it is not included in the diagnostic criteria for neuropsychiatric involvement associated with SLE, several cases have been reported in the literature, but only in patients with pre-existing, known SLE. We present here a case of catatonia revealing SLE.

Case Presentation With Investigation: This is a 21-year-old female patient with no significant medical history who presented several days prior to hospitalization with inflammatory arthralgia affecting both small and large joints, complicated by a catatonic syndrome associated with sepsis of pulmonary and urinary origin. This situation led to hospitalization in the intensive care unit for appropriate management. Clinical examination revealed a malar rash, negativism, and mutism. Laboratory results showed normochromic normocytic anemia (hemoglobin at 10.7 g/dl), lymphopenia (690/mm^3), a positive direct Coombs test, hypoalbuminemia, elevated alpha-1 globulins, and polyclonal hypergammaglobulinemia on protein electrophoresis (EPP). Autoimmune tests were positive, including antinuclear antibodies (ANA) at 1/160, strongly positive anti-SSA, anti-SSB, and anti-nucleosome antibodies, as well as positive anti-Sm, anti-histone, and anti-ribosome antibodies, and a borderline anti-RNP. The anti-glycoprotein antibody (Ig M) was strongly positive (58 U/mL). C3 and C4 complement levels were consumed. Hepatic and renal assessments were normal, and 24-hour proteinuria was negative. Serologies for hepatitis B and C viruses, HIV, and syphilis were negative. Thyroid function tests were also normal, as was the pregnancy test (BHCG). A lumbar puncture and cerebral MR angiography were performed, with no abnormalities detected. Deficiencies, toxic, and iatrogenic causes of catatonia were also ruled out. The diagnosis was established as a catatonic syndrome unveiling systemic lupus erythematosus (SLE). The patient was treated with hydroxychloroquine,

a methylprednisolone bolus followed by an oral dosage of 1 mg/kg/day, in addition to appropriate antibiotic therapy. After resolution of the infectious episode, a cyclophosphamide bolus was administered. Psychiatric care was provided alongside specific lupus treatment. The clinical outcome was favourable, with notable improvement in the catatonic syndrome, including resolution of negativism, as well as a gradual return of speech and eating.

Literature Review: Cases of catatonia associated with systemic lupus erythematosus have been reported in the literature, with the majority occurring in patients with a previously diagnosed systemic lupus. Although catatonia is not part of the official neuropsychiatric criteria for systemic lupus erythematosus, this case, along with those described in the literature, suggests that it may represent an important, though rare, manifestation of neuropsychiatric systemic lupus erythematosus (NPSLE).

Discussion: Distinguishing between NPSLE-induced catatonia and other causes, such as steroid-induced psychosis or antipsychotic-induced catatonia, can be challenging. However, the absence of steroid or antipsychotic use in our patient, alongside improvement with immunosuppressive therapy, strongly suggests that NPSLE was the underlying cause. The neurophysiological mechanisms behind catatonia in NPSLE remain unclear, and further research is needed to clarify these mechanisms and improve diagnosis and management.

PV299 / #287

Case Report Poster Topic: *AS23 - SLE-Diagnosis, Manifestations, & Outcomes*

AN UNUSUAL CASE OF LYMPHOCYTIC ENTEROCOLITIS AND VASCULITIS AS THE INITIAL MANIFESTATION OF SLE

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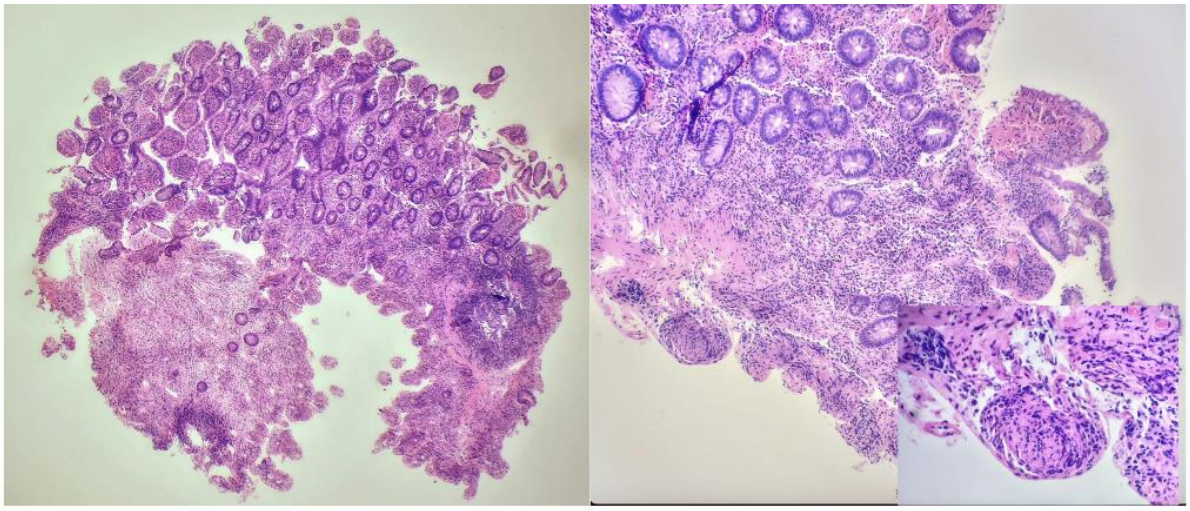
Introduction: Systemic Lupus Erythematosus (SLE) is a complex autoimmune disease with multi-systemic involvement, potentially affecting almost every organ. Lymphocytic enterocolitis (LE), or microscopic colitis, is a rare manifestation of SLE. We present an unusual case of a woman with persistent watery diarrhoea and LE as the initial presentation of newly diagnosed SLE.

Case Presentation With Investigation: A 69-year-old woman presented with a six-month history of watery, non-bloody diarrhoea, with mucus, progressively worsening from 1-2 episodes daily to up to 5 episodes. This was associated with a significant weight loss of 17 kg, though she had no abdominal pain, fever, or nocturnal symptoms. Her medical history included hypertension, Raynaud's phenomenon, and oral ulcers. Physical examination revealed normal bowel sounds without tenderness. Laboratory tests showed leukopenia (WBC = 3,300/ µl 4,500-10,500) with lymphopenia (Lymph=380/ µl 1,200-3,800) elevated ESR =78 mm/h, and mild hypoalbuminemia =3.2 g/dl. (3.5-5.2). Stool cultures and faecal calprotectin tests were unremarkable, as was the initial GI endoscopy. Celiac serology was negative. A repeat colonoscopy due to worsening symptoms revealed multiple shallow, intermittent mucosal ulcers throughout the bowel, while the terminal ileum remained unaffected. Upper GI endoscopy showed mild erosions in the antrum. Bowel histology showed predominant lymphocytic infiltration with mild neutrophilic aggregates, microbleeds, and some vasculitic changes (Picture 1). Faecal calprotectin remained within normal limits. Screening for connective tissue disorders performed due to the presence of mouth ulcers and Raynaud's phenomenon. This was positive for ANA at 1:1280 (homogeneous pattern), anti-dsDNA at 344 IU/mL (<100), anti-ENA IU/mL at 156 (<20), anti-RNP >100 IU/mL (<20), and anti-Sm at 82 IU/mL (<20), with low complement levels C3 = 35 mg/dl (90-180), C4 = 5 mg/dl (10- 40). An SLE diagnosis was confirmed, and the patient began treatment with methyprednisolone 32mg/daily (with gradual tapering) and Azathioprine (50 mg twice daily), resulting in rapid symptom relief. She remained asymptomatic at the two-year follow-up.



Picture 1:Left image profound lymphocytic infiltrate with neutrophilic aggregations and lymp, Right image, significant lymphocytic infiltrates with evidence of vasculitis (magnified spot) with microbleeds.

Literature Review: Approximately 40% of SLE patients experience gastrointestinal symptoms, including lupus enteritis, pseudo-obstruction, protein-losing enteropathy, hepatitis, pancreatitis, vasculitis, and mesenteric ischemia (1). However, lymphocytic enterocolitis is exceedingly rare, with only a few cases documented (2). Symptoms typically develop insidiously, with fewer, less severe bowel movements than inflammatory bowel disease, and generally respond well to SLE immunosuppressive therapy (2). (1)Sultan SM, Ioannou Y, Isenberg DA A review of gastrointestinal manifestations of systemic lupus erythematosus.Rheumatology (Oxford). 1999;38(10):917. (2)Hegazi MO, Owayed SF, Mourou M, Joneja M, Mashankar M.Lymphocytic Enterocolitis in Systemic Lupus Erythematosus Saudi J Gastroenterol 2009 Oct;15(4):274–276. doi: 10.4103/1319-3767.56100

Discussion: Microscopic enterocolitis can represent an initial manifestation of SLE, and if left untreated, may progress to complications such as vasculitis, as observed in this patient. A thorough medical history is crucial when diagnosing connective tissue diseases, given the broad range of possible clinical presentations.

PV300 / #635

Case Report Poster Topic: *AS24 - SLE-Treatment*

EXPERIENCE WITH ANIFROLUMAB IN SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS WITH NEUROLOGICAL MANIFESTATIONS

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Introduction: Anifrolumab, a biologic approved for systemic lupus erythematosus (SLE), excludes patients with active neurological manifestations in clinical trials. However, real-world clinical practice often necessitates its use in this population when other options are limited. This study analyzes the effectiveness of Anifrolumab in three SLE patients with neurological involvement.

Case Presentation With Investigation: We retrospectively reviewed three female patients treated with Anifrolumab who presented with neurological manifestations in addition to systemic disease. Clinical characteristics, treatment response, and safety outcomes were evaluated.

Literature Review: • Case 1: A 29-year-old woman with cutaneous and hematological SLE presented with acute psychosis. Extensive workup, including neuroimaging, lumbar puncture, and infection studies, was negative. She was treated with corticosteroid pulses followed by Anifrolumab, achieving disease remission and discontinuing corticosteroids after six months.
• Case 2: A patient with moderate-to-severe hematological SLE refractory to mycophenolate also presented with neurological involvement. Anifrolumab was initiated but discontinued after three months due to refractory thrombocytopenia and a cerebrovascular event.
• Case 3: A woman with articular, hematological, and cutaneous SLE, including significant alopecia and recurrent lupus headaches requiring steroid pulses, achieved disease remission with Anifrolumab. Additionally, no steroid pulses were required for six months after treatment initiation.

Discussion: Anifrolumab demonstrated effectiveness in some SLE patients with neurological manifestations, particularly in reducing corticosteroid use and managing lupus headache. However, safety concerns, such as thrombocytopenia and cerebrovascular events, warrant careful monitoring. These cases highlight the need for further studies to assess Anifrolumab's role in patients with neurological involvement.

PV301 / #634

Case Report Poster Topic: *AS24 - SLE-Treatment*

USE OF UPADACITINIB IN TWO CASES OF REFRACTORY SYSTEMIC LUPUS ERYTHEMATOSUS

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Introduction: Upadacitinib, a selective JAK inhibitor, is under evaluation in clinical trials alone and in combination with elsubrutinib for systemic lupus erythematosus (SLE). In our cohort, it has been employed off-label in two refractory cases of SLE with promising outcomes.

Case Presentation With Investigation: Two female patients with long-standing, refractory SLE were treated with upadacitinib 15 mg/day after multiple therapeutic failures. Clinical history, prior treatments, response to upadacitinib, and disease activity were analyzed.

Literature Review: • Case 1: A 54-year-old woman with SLE involving articular, hematological, cutaneous, and neurological (lupus headache) domains, alongside severe fatigue meeting chronic fatigue syndrome criteria. The patient failed conventional treatments, including belimumab. Upadacitinib 15 mg/day was initiated, resulting in complete disease remission and significant improvement in fatigue and headache. The patient has sustained remission for 18 months.
• Case 2: A 59-year-old woman with a 39-year history of SLE predominantly involving articular, cutaneous, hematological, and serosal domains. After multiple treatments, including methotrexate, leflunomide, mycophenolate, belimumab, rituximab, anifrolumab, and off-label baricitinib, she exhibited persistent disease activity. Upadacitinib 15 mg/day was started, leading to low disease activity within three months. However, the patient continues to require corticosteroids.

Discussion: Upadacitinib may represent a promising therapeutic option for refractory SLE, achieving disease remission or low disease activity in patients unresponsive to conventional and biological therapies. Further studies are needed to evaluate its safety and efficacy in broader SLE populations.

PV302 / #548

Case Report Poster Topic: *AS24 - SLE-Treatment*

ANTI-CD19 MONOCLONAL ANTIBODY FOR SYSTEMIC LUPUS ERYTHEMATOSUS AND NEUROMYELITIS OPTICA SPECTRUM DISORDER

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Introduction: Systemic Lupus Erythematous (SLE) is a multisystem autoimmune disease involving different immune pathways including B cell dysregulation. B cell-directed agents have shown efficacy in SLE and lupus nephritis, prompting the development of therapies targeting CD19, a transmembrane protein on pro-B cells, immature and mature B cells, memory B cells, plasmablasts and plasma cells. Anti-CD19 monoclonal antibodies have shown efficacy for neuromyelitis optica spectrum disorder (NMOSD) [1], a demyelinating disorder that may coexist with SLE. It is unclear whether anti-CD19 monoclonal antibody may also be effective for treating SLE.

Case Presentation With Investigation: 1) A 35-year-old Caucasian man developed slurred speech, paresthesia, and gait disturbance, found to have demyelinating lesions, aquaporin-4 immunolgoblin G (IgG) >1:100,000 and was diagnosed with NMOSD in the setting of concomitant SLE with pleuro-pericardial effusions, positive Anti-nuclear antibody (ANA), anti-double stranded DNA (dsDNA), anti-ribonucleoprotein(RNP), anti-chromatin, anti-Smith, anti-SSA, anti-SSB, anti-cardiolipin, anti-β2-glycoprotein antibodies, low complement 3 (C3) and C4. He was treated with high-dose steroids, plasma exchange, hydroxychloroquine and azathioprine which was discontinued due to cytopenia. He was then given inebilizumab with improved neurologic deficits and MRI findings. His steroid doses were tapered off and he remains on inebilizumab and hydroxychloroquine with no signs of SLE or NMOSD activity one year later. 2) A 25-year-old African American woman diagnosed with SLE, class V lupus nephritis, polyarthritis, with positive ANA, anti-chromatin, anti-RNP, anti-Smith, and anti-SSA antibodies, low C3 and C4, treated with steroid taper, hydroxychloroquine and mycophenolate dose reduced due to cytopenia. She initially presented with unilateral vision loss and paresthesia although her diagnosis of NMOSD was not made until she completed neurologic workup a year after her SLE diagnosis, which showed demyelinating lesions on spine MRI and aquaporin-4 IgG >1:100,000. She was then started on rituximab but due to inadequate response in her neurologic symptoms she was switched to inebilizumab. Six months after initiation she has no signs of SLE activity and continues to have visual disturbances and paresthesia but not attributed to active demyelination.

Literature Review: There is limited data on the use of anti-CD19 monoclonal antibody for SLE. A trial with obexelimab for SLE did not meet the primary endpoint of proportion of patients reaching week 32 without loss of improvement [2]. However, there was a significant increase in time to loss of improvement in the obexelimab-treated patients. An ongoing trial is investigating the use of inebilizumab and blinatumomab, a bispecific CD19-directed CD3 T cell engager for SLE. One case series of 3 patients with SLE coexisting with NMOSD reported various therapies used for those patients including steroids, cyclophosphamide, rituximab, azathioprine, hydroxychloroquine [3]. There were no cases found in the literature of SLE and coexisting NMOSD treated with anti-CD19 antibodies.

Discussion: We present two cases of SLE with concurrent NMOSD controlled with anti-CD19 monoclonal antibody with remission of SLE activity. Compared to CD20, CD19 is expressed by more cells in the B cell lineage that may lead to greater inhibition of B cell signalling. Targeting CD19 may be effective for patients with coexisting SLE and NMOSD. References: 1. Cree BAC, Bennett JL, Kim HJ, et al. *Lancet*. 2019;394(10206):1352-1363 2. Merrill JT, Guthridge J, Smith M, et al. *Arthritis Rheumatol*. 2023;75(12):2185-2194 3. Ochi MGS, Shapiro SC, Melamed E. *Case Rep Rheumatol*. 2020;2020:8820071

PV303 / #378

Case Report Poster Topic: *AS24 - SLE-Treatment*

THERAPEUTIC CHALLENGES IN POLYAUTOIMMUNITY: ANIFROLUMAB FOR LUPUS ERYTHEMATODES IN OVERLAPPING AUTOIMMUNE SYNDROMES

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Introduction: Polyautoimmunity is defined as the presence of two autoimmune diseases in a single individual, while Multiple Autoimmune Syndrome (MAS) involves three or more coexisting autoimmune conditions.[1] Diagnosing specific diseases in patients with multiple autoimmune conditions can be challenging and treatment regimens are often complex. Here we present two patients with polyautoimmunity and refractory lupus erythematosus who showed significant symptom improvement after adding anifrolumab, a monoclonal antibody targeting the interferon alpha receptor 1 (IFNAR1), to their treatment regimen.

Case Presentation With Investigation: Case 1: A 57-year-old woman with a history of multiple sclerosis (MS), treated with glatiramer acetate, presented with recurrent facial and periungual erythema, fatigue, weight loss, fever episodes, and finger pain. Diagnostic work-up, including elevated ANA titer of 1:320, positive anti-dsDNA antibodies (42 U/mL), strongly positive anti-MDA5 and anti-RO-52 antibodies, led to an overlap syndrome of amyopathic dermatomyositis and lupus erythematosus. To address her full spectrum of autoimmune diseases, her MS treatment was switched to azathioprine and prednisolone, resulting in improved general condition and reduced facial erythema. However, painful periungual lesions with fissures persisted, greatly impairing her quality of life. Due to fine motor skill loss and significantly elevated Siglec-1 expression (16,347 antigens/monocyte), anifrolumab was added to her treatment regimen. This led to a rapid and marked improvement in the previously resistant periungual lesions, allowing corticosteroid tapering while maintaining MS stability. Case 2: A 60-year-old male patient with a history of psoriasis vulgaris and previous thromboembolic events presented with painful, scarring lesions on the fingers, nose, and scalp. Histology confirmed discoid lupus, and laboratory tests revealed a high ANA titer (1:1280), anti-dsDNA antibodies, and positive antiphospholipid antibodies, resulting in diagnoses of systemic lupus erythematosus (SLE) and antiphospholipid syndrome. Hydroxychloroquine was avoided due to concerns about exacerbating

psoriasis. Alternative therapies with methotrexate and mycophenolate mofetil provided limited benefit. Although apremilast effectively controlled psoriasis, lupus lesions continued to progress. The initiation of anifrolumab therapy led to a rapid improvement in cutaneous lupus symptoms within weeks, while psoriasis remained stable and was well-controlled with topical treatments. Phenprocoumon was added for thromboembolic prevention, with no further thromboembolic events reported.

Literature Review: Anifrolumab is an effective treatment for refractory SLE, particularly cutaneous manifestations. Evidence shows that blocking type I interferon pathways, central in lupus pathology, can effectively reduce inflammatory responses. Clinical trials (e.g., TULIP-1 and TULIP-2) have demonstrated anifrolumab's efficacy in decreasing both systemic and cutaneous disease activity.[2] While the use of anifrolumab in polyautoimmunity and MAS requires further research, its targeted mechanism of action and favorable safety profile already make it a promising option for patients with overlapping connective tissue diseases.[3]

Discussion: Both cases illustrate the potential of anifrolumab as an effective add-on therapy for targeting refractory cutaneous symptoms in patients with connective tissue diseases. Although anifrolumab is currently approved only for SLE, its positive effects may extend to other connective tissue diseases characterized by elevated interferon alpha activity. The successful and safe use in patients with polyautoimmunity underscores anifrolumab's suitability for patients with overlapping autoimmune diseases. [1.] Matusiewicz A. Int J Rheum Dis 2019;22:386-391 [2.] Morand EF. Lancet Rheumatol 2022;4:282-292 [3] Shaw KS.J Am Acad Dermatol 2024;doi:10.1016/j.jaad.2024.07.1491

PV304 / #276

Case Report Poster Topic: *AS23 - SLE-Diagnosis, Manifestations, & Outcomes*

THE IMPORTANCE OF TRANS-ESOPHAGEAL ECHOCARDIOGRAPHY IN DIAGNOSING LIBMAN-SACKS ENDOCARDITIS IN SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

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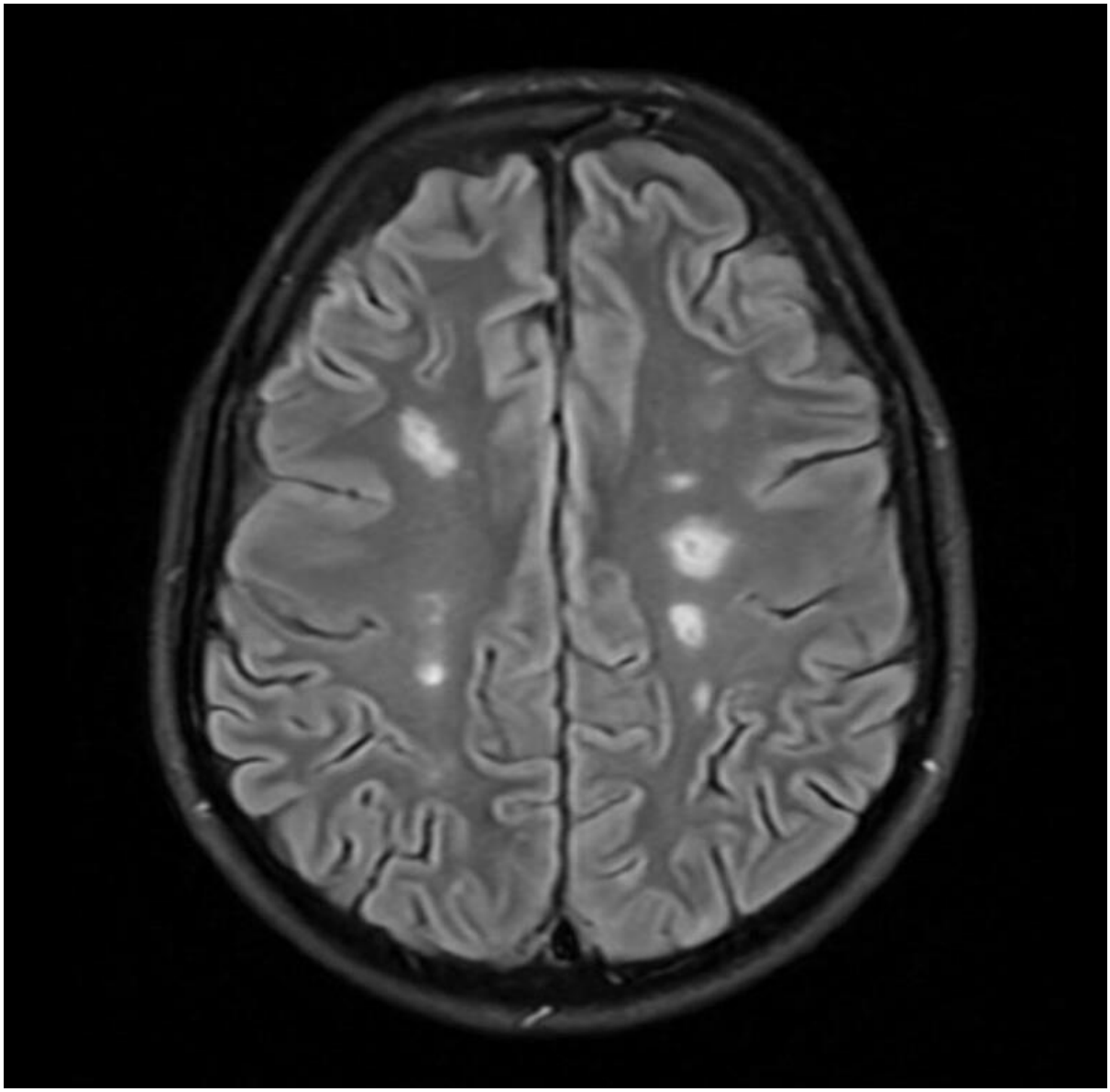
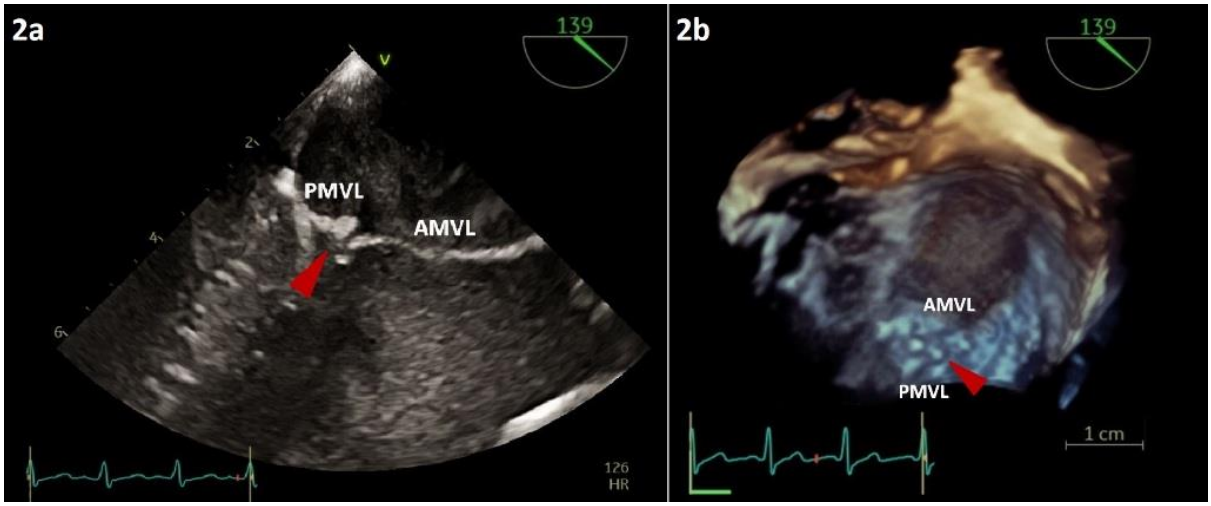
Introduction: Libman-Sacks endocarditis (LSE), a form of non-bacterial thrombotic endocarditis (NBTE), is characterized by fibrinous, sterile vegetations, favoring the mitral and aortic valves. LSE is observed in up to 11% of patients with systemic lupus erythematosus (SLE) as one of two major valvular manifestations, the other being thickening, with both mechanisms contributing to valvular dysfunction. LSE may typically be clinically silent, but predisposes to greater risk of cerebrovascular disease, underpinning the importance of its recognition. Complicating this, LSE may be clinically mimicked by infectious endocarditis; both may present with arterial embolism and may share elevated inflammatory markers and positive autoantibodies. Here, we present a complex first presentation of SLE with LSE resulting in cerebrovascular disease in a young patient. This case reinforces the importance of trans-esophageal echocardiography (TEE) in differentiating morphological features of NBTE from infectious endocarditis to guide timely management.

Case Presentation With Investigation: A 22-year-old male of fit-and-well background presented with progressive lower limb swelling and right upper limb weakness. Broad-spectrum intravenous antibiotics were commenced for suspected infectious endocarditis, given trans-thoracic echocardiography (TTE) findings of moderate mitral regurgitation, magnetic resonance imaging (MRI) brain evidence of multiple embolic strokes [figure 1], and progressive kidney injury with microscopic hematuria. Characteristic findings of LSE unseen on TTE were visualized by TEE: sessile posterior mitral valve leaflet tip lesions and thickening of the leaflets [figure 2]. This supported the clinical suspicion of NBTE, reinforced by persistent culture negativity. Laboratory tests confirmed positive anti-nuclear and double-stranded DNA antibodies, compatible with a first presentation of SLE, featuring cerebrovascular disease, LSE and lupus nephritis. Early TEE findings supported multi-disciplinary team agreement to switch from intravenous antibiotics to high-dose corticosteroids.





Literature Review: LSE is an under-reported manifestation of SLE and is one of two mechanisms whereby lupus may affect the heart valves, alongside valvular thickening.

SLE patients with neurological sequelae and high index of suspicion for NBTE should first undergo TTE to assess for valvular thickening or vegetations, as their presence relates to three-fold greater numbers of circulating cerebral microemboli and subsequent cerebrovascular disease[1.]. Trans-thoracic imaging, however, underestimates lupus-associated valve disease, and poorly differentiates NBTE from infective endocarditis. No individual biochemical test can confirm LSE; imaging must therefore play a vital role in differentiation. Importantly, three-dimensional TEE possesses greater sensitivity and specificity than TTE and two-dimensional TEE for visualization of LSE vegetations[2.]. LSE-related valvular masses may be differentiated from bacterial vegetations based on their appearance, mobility, and location. Libman-Sacks vegetations are sessile, typically smaller (<10mm), are heterogeneous in echotexture, may show central calcification, and are visualized at the leaflet base. Conversely, bacterial lesions are homogenous in echotexture, are found at the line of leaflet closure, and move independently from the motion of the valve[3.]. In our patient, we observed typical features of NBTE, namely valvular thickening [figure 2a] and multiple small, sessile lesions [figure 2b], correlating with his extra-cardiac manifestations of lupus including cerebrovascular disease [figure 1].

Discussion: Our case highlights the importance of trans-esophageal echocardiography in the diagnosis of SLE-related LSE and differentiation from infective endocarditis, which may mimic NBTE clinically and biochemically. TEE may complement trans-thoracic imaging in diagnosing NBTE to identify a subset of patients with SLE at greatest risk of cerebrovascular disease who may benefit from intensive immunosuppression.
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PV305 / #413

Case Report Poster Topic: *AS24 - SLE-Treatment*

ANIFROLUMAB IN THE MANAGEMENT OF LUPUS HEADACHES: A CASE REPORT

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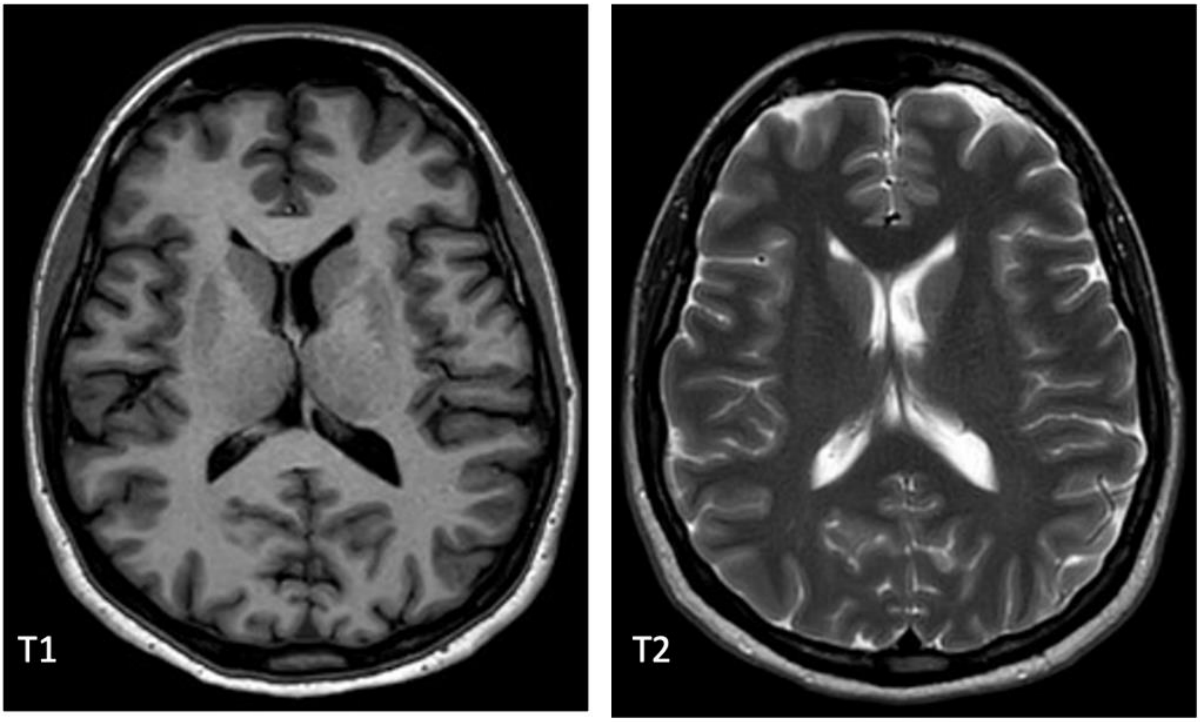
Introduction: We present a 52-year-old woman with a 10-year history of systemic lupus erythematosus (SLE) characterized by systemic, articular, cutaneous, and neurological involvement (lupus headache). Her condition was refractory to treatment with glucocorticoids, hydroxychloroquine, and belimumab. Despite treatment, her headaches remained resistant to nonsteroidal anti-inflammatory drugs (NSAIDs), triptans, and amitriptyline. After initiating treatment with anifrolumab 300 mg monthly for five months, she experienced significant symptom resolution, including improvement of her lupus headaches.

Case Presentation With Investigation: The patient was diagnosed with SLE in 2014 after presenting with pericarditis, polyarthritis, photosensitive skin lesions, recurrent oral aphthae, and positive ANA, with no anti-dsDNA elevation or complement consumption. She also had secondary Sjögren's syndrome (anti-Ro >320 U) and Raynaud's phenomenon. Initial treatment included glucocorticoids and hydroxychloroquine (200 mg/day). Methotrexate was trialed in 2019 but was stopped due to ineffectiveness. In 2018, belimumab was added for persistent polyarthritis. In 2022, she developed persistent unilateral headaches resistant to migraine therapies (NSAIDs, triptans, amitriptyline, and flunarizine). By April 2024, her systemic symptoms worsened with chronic fatigue, polyarticular pain, and photosensitive rash, requiring glucocorticoids. Neurologically, she had refractory headaches and depressive symptoms (normal MRI (Image 1); SLEDAI-2k 14, SLE-DAS 31.04). Belimumab was stopped, and anifrolumab 300 mg IV monthly was started. Image 1. MRI scans with T1 and T2 sequences showed no abnormalities.



After one month of anifrolumab, she had fewer, less intense headaches and improved cutaneous and articular symptoms. By five months, systemic symptoms resolved, and headaches diminished. Laboratory tests showed normalized lymphopenia, stable hemoglobin, and normal CRP, with no anti-dsDNA elevation or complement consumption (SLEDAI-2k 0, SLE-DAS 0.37).

Literature Review: Anifrolumab, a human monoclonal antibody targeting type I interferon receptor subunit 1, was approved in March 2022 for SLE treatment. It is included in the updated 2023 EULAR recommendations for managing SLE, particularly for patients not responding to hydroxychloroquine or those unable to reduce glucocorticoids to acceptable doses. Phase 3 trials (TULIP 1 and TULIP 2) demonstrated that Anifrolumab leads to rapid and sustained reductions in global and organ-specific disease activity, facilitating glucocorticoid tapering. Recently, new real-world data have been published demonstrating the activity of Anifrolumab and observed rapid effectiveness in severe and refractory patients, particularly in those with cutaneous, joint, and hematological involvement. Additionally, other manifestations, including renal or neuropsychiatric symptoms, are also studied also responded to treatment. Neurological manifestations in systemic lupus erythematosus (SLE) encompass a wide range of neurologic and psychiatric symptoms with varying severity, often complicating the differentiation from unrelated conditions. Lupus headache is a term used to describe a severe headache that is directly attributed to SLE with no secondary cause, and the pathogenic mechanism is unclear. The prevalence of headache in SLE varies from 24 to 72% with no clear association with disease activity, and the management typically involves a combination of treatments targeting both the neurological complications and the underlying SLE. However, the efficacy of these treatment

protocols has not been rigorously studied, highlighting the need for further investigation.

Discussion: Anifrolumab may prove beneficial for SLE patients with neurological or neuropsychiatric involvement, including lupus headaches, showing a positive response and sustained improvement from the first month of treatment.

PV306 / #753

Case Report Poster Topic: *AS03 - Antiphospholipid Syndrome*
Late-Breaking Abstract

ECULIZUMAB IN THE MANAGEMENT OF CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME WITH MULTIORGAN INVOLVEMENT: A CASE REPORT

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Introduction: Catastrophic antiphospholipid syndrome (CAPS) is a severe and rapidly progressive form of antiphospholipid syndrome that results in multiorgan failure due to thrombosis in the microvasculature. It is rare, occurring in only 1% of patients with antiphospholipid syndrome, but it carries a high mortality rate and can be the first manifestation of the disease. Eculizumab, a C5-blocking monoclonal antibody, is effective in the treatment of thrombotic microangiopathy by controlling complement system hyperactivation. Due to the rarity of CAPS, there is limited literature regarding the
appropriate management of these patients, and there is little clinical experience with Eculizumab treatment. This case report aims to provide clinical insight to improve the management of patients with catastrophic antiphospholipid syndrome and multiorgan involvement.

Case Presentation With Investigation: We present the case of a 62-year-old female patient with a debut diagnosis of CAPS secondary to systemic lupus erythematosus. She exhibited renal involvement with significant deterioration of kidney function, which required hemodialysis. Renal biopsy revealed thrombotic microangiopathy affecting small and medium-sized vessels and glomeruli. Additionally, she exhibited cardiopulmonar involvement, including non-ischemic dilated cardiomyopathy and pulmonary hypertension. Laboratory findings showed hemolytic anemia, thrombocytopenia, renal failure, complement consumption, and positivity for ANA, anti-cardiolipin IgM, and beta-2 glycoprotein antibodies.In the first weeks following the initial diagnosis, the patient received treatment with hydroxychloroquine (200 mg every 12 hours), methylprednisolone boluses (500 mg for 3 days), followed by prednisone at a tapering dose, immunoglobulin therapy (weight-adjusted for 5 days), mycophenolate (500 mg every 12 hours), and 3 cycles of Rituximab 1g. Anticoagulation was managed with bemiparin. Additionally, treatment with Eculizumab was initiated with 4 weekly doses, followed by Eculizumab every 2 weeks for 3 months. It was interrupted due to the patient's admission for bacteremia, with a good clinical outcome. Afterward, it was not reintroduced as the patient remained stable with her disease and showed very good

clinical progress, as well as normalization of laboratory values, with no significant incidents during 18 months.

Literature Review: Almeida C. Autoimmun Rev 2015;14:1087-96. Guerra, N., García, J et al. Acta Colombiana de Cuidado Intensivo 2019;.19:154-9

Discussion: Given the severity and high mortality of CAPS, early diagnosis and appropriate treatment are critical to prevent irreversible structural damage to target organs. Early treatment with anticoagulation and intravenous corticosteroids, followed by plasmapheresis, rituximab, and eculizumab in the early stages, is crucial to achieve disease remission and prevent irreversible structural damage to target organs.