A Concomitant Myriad of Neurological Symptoms in a Patient with SLE Ending up in a Rare Complication of Osteonecrosis of the Jaw

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ABSTRACT

Systemic lupus erythematosus (SLE) can have a legion of multi-organ manifestations, whereas neuropsychiatric SLE refers to the neurological and psychiatric features in the context of active SLE. Neurological features constitute diverse central and peripheral symptoms. Patients who experience a flare of SLE receive immunosuppressants for disease control. At one end of the spectrum, these immunosuppressive medicines are life-saving, and at the other end, they can have several undesirable effects. We present the case of a young female admitted with a flare of SLE who exhibited multiple peripheral and central nervous system manifestations during her hospital course. She received multi-disciplinary care for her complaints; however, she ended up with an unusual complication of osteonecrosis of the jaw.

Key Words: *Systemic lupus erythematosus, Neuropsychiatric SLE, Osteonecrosis of the jaw*.

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INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune rheumatic condition with heterogeneous presentations, marked by flares, relapses, and remissions.[1] Neuropsychiatric SLE (NPSLE) can manifest as one of the 19 distinct clinical syndromes according to the American College of Rheumatology (ACR). It often presents as a diagnostic and therapeutic dilemma owing to its elusive presentations.[2]

In this case report, the case of a young female is illustrated who experienced a flare of NPSLE and was managed as per guidelines.[3] She had a multitude of peripheral and central neurological symptoms in addition to the psychiatric complaints. Her hospital course was full of multiple disease-related and treatment-related complications. She ultimately developed osteonecrosis of the jaw (ONJ)¾a potential complication rarely reported in the context of SLE.[4]

CASE REPORT

A 23-year married female was diagnosed as a case of NPSLE five years ago, when she presented to the outpatient department with choreiform movements of the right side of the body, followed by fever, arthralgias, and a malar rash.

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Her diagnosis was validated by the laboratory parameters, which showed high titres of anti-nuclear antibody (ANA), strongly positive anti-dsDNA, and low complement levels. MRI of the brain was suggestive of CNS vasculitis (Figure 1). She received induction therapy with Cyclophosphamide (CYC), followed by maintenance therapy with Mycophenolate mofetil (MMF), which was switched to Azathioprine (AZA) due to intolerance to MMF. Her disease remained quiescent for four years on AZA. In July 2024, she was admitted due to suspicion of disease flare as she had fever, arthralgias, oral ulcers, dysuria, and hypo-complementaemia.

During the initial days of admission, she developed behavioural changes, left lateral rectus palsy, followed by paresthaesias of the extremities. Fundoscopy and CSF analysis were unremarkable. Considering neuropsychiatric features and a history of two miscarriages, an antiphospholipid antibody (APLA) profile was sent that turned out to be negative. However, MRI and MRV of the brain revealed multiple acute infarcts in the midbrain and pontine areas, and the nerve conduction study was suggestive of a sensorimotor axonal polyneuropathy.

For the flare of NPSLE, she was administered a pulse therapy with three doses of methylprednisolone (500 mg), followed by CYC as per the Euro-lupus protocol. After a week of receiving CYC, she developed severe neutropenia followed by macrophage activation syndrome (evident on laboratory parameters and confirmed on bone marrow biopsy) (Table I). These complications were managed with a neutropenic regimen consisting broad-spectrum antibiotics and granulocyte-monocyte colony-stimulating factor (GM-CSF). The macrophage activation syndrome was managed with high-dose glucocorticoids to which she responded well.

Table I: Relevant investigations.

Lab parameters	Results	Reference ranges
Hb	8.9 g/dl	11 - 13 g/dl
MCV	72 fl	80 - 95 fl
TLC	0.5 × 10^3/uL	4 - 11 × 10^3/uL
ANC	0.35 × 10^3/uL	1.5- 8 × 10^3/uL
ALC	0.15 × 10^3/uL	1 - 4.5 × 10^3/uL
ESR	43mm/1st hour	0-15 mm/1st hour
Platelets	75000/uL	150000-450000/uL
ALT	50 IU/L	5-50 IU/L
Creatinine	0.7 mg/dl	<1.2 mg/dl
Ferritin	10597 ng/ml	10-204 ng/ml
Pro-calcitonin	5.82 ng/ml	<0.5 ng/ml
CRP	21 mg/L	<5 mg/L
Triglycerides	690 mg/dl	<150 mg/dl
Urine RE	4-6 pus cells, blood + protein negative, nitrite negative	No pus cells and blood
Urine CS	No growth	Normal

Hb, Haemoglobin; MCV, Mean corpuscular volume; TLC, Total leucocyte count; ANC, Absolute neutrophil count; ALC, Absolute lymphocyte count; ESR, Erythrocyte sedimentation rate; ALT, Alanine aminotransferase; CRP, C-reactive protein; Urine RE, Urine routine examination; Urine CS, Urine culture and sensitivity.

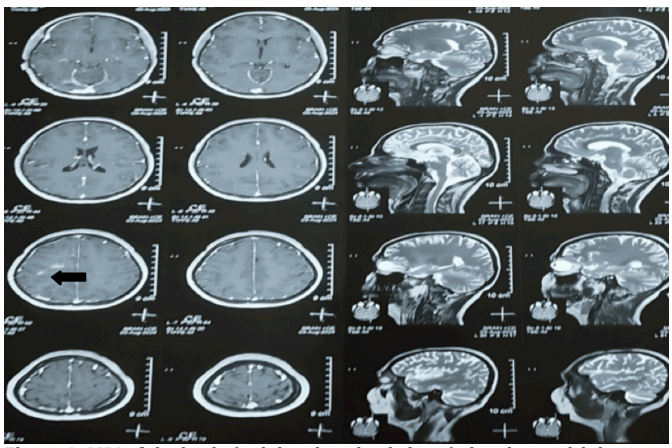

Figure 1: MRI of the brain (axial and sagittal views) showing multiple acute ischaemic infarcts (arrows), suggestive of CNS vasculitis in the setting of SLE.

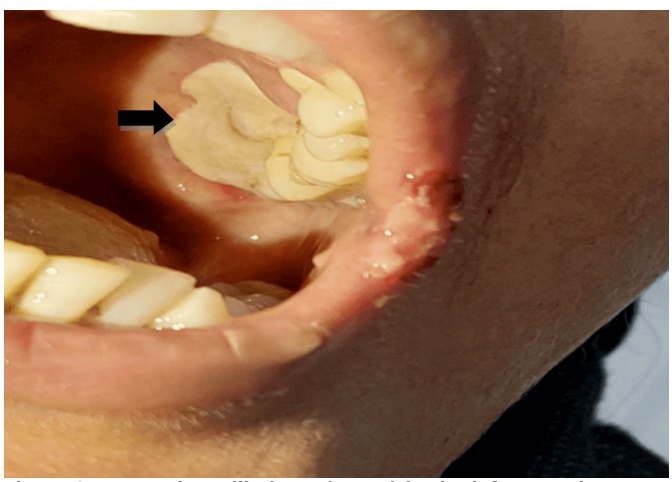

Figure 2: Exposed maxilla bone (arrow) in the left upper jaw, suggesting a necrotic area and highly indicative of osteonecrosis of the jaw.

Meanwhile, the patient complained of pain in the lower dentition. A dental consultation was obtained, and she was referred to the oral and maxillofacial surgery (OMFS) department for a thorough assessment. A three-phase bone scan revealed ONJ involving the right haemimandible and left maxilla (Figure 2). According to the OMFS department, it could be a complication of her underlying disease or a consequence of immunosuppressive therapy (glucocorticoid/non-glucocorticoid).

Following the diagnosis of ONJ, her steroids were tapered down to the lowest possible dose, and CYC was also stopped after three pulses to allow her to undergo surgical repair for ONJ. She underwent extensive debridement of the right mandible and left maxilla, along with the left nasal antrostomy. The patient is currently on MMF as maintenance therapy along with hydroxychloroquine and antiplatelet, off steroids, and has recovered well postoperatively with good wound granulation.

DISCUSSION

In this case report, a case of SLE diagnosed as per the 2019 ACR-EULAR classification criteria for SLE, is presented.[5] NPSLE can manifest as 19 different clinical syndromes as per 1999 ACR NPSLE case definitions. Twelve out of these are attributed to CNS, while seven are related to the peripheral nervous system and psychiatric features.[6] Among these case presentations, this patient fulfilled the criteria for movement disorders, cerebrovascular disease, polyneuropathy, and psychosis. Manifestations of the peripheral nervous system are highly variable and inexplicable in the context of NPSLE.[7] Her disease flare was successfully managed with immunosuppressants as per the latest guidelines for organ-threatening SLE, and she started to show remarkable improvement. The array of complications can be attributed to the disease process itself, or they could be iatrogenic (secondary to immunosuppressive medicines). Non-traumatic osteonecrosis is a debilitating and irreversible complication of SLE with a prevalence of 7.6% as per a Chinese cohort study.[8] Systemic steroids, CNS involvement, and APLA are considered significant risk factors for multiple sites of osteonecrosis in patients with SLE. Medication-related ONJ has been reported frequently with antiresorptive (bisphosphonates and denosumab) and antineoplastic medications.[9] However, this patient, despite not being on the aforementioned medicines, suffered from ONJ, thus advocating for SLE, high-dose glucocorticoids, and CNS involvement as the major culprits.

SLE and its neuropsychiatric presentation portend a significant risk of morbidity to the patient due to the plethora of presentations and high incidence of complications associated with both the disease process and treatment modalities. Patients and their families should be critically counselled in this regard. ONJ is a rare feature in patients with SLE, thus making it a unique case presentation.

PATIENT'S CONSENT:

Informed consent was obtained from the patient to publish the data concerning this case.

COMPETING INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

QUAA: Drafting of the work and acquisition.
TK: Final approval of the work.
SAS: Conception.
SS: Critical revision of the manuscript for the important intellectual content.
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