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Evaluation and Management of Spinal Subarachnoid Hemorrhage in a Patient with Lupus Vasculitis

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Female, 37
Final Diagnosis: Lupus vasculitis
Symptoms: Back pain • headache • paralysis • sensory loss
Medication: Cyclophosphamide • prednisone • methylprednisolone
Clinical Procedure: CT/MRI thoracic spine • lumbar puncture • plasmapheresis • kidney biopsy
Specialty: Neurology

Objective: Challenging differential diagnosis





Background: Isolated spinal artery subarachnoid hemorrhage is a rare occurrence in the general population, but occurs more commonly as one of many neurologic sequela of systemic lupus erythematosus (SLE). The etiology of a neurologic deficit in an SLE patient is often multifactorial. Comorbid conditions, such as antiphospholipid antibody syndrome, predispose to stroke. Other diagnoses, including transverse myelitis, may also be attributed to local inflammation.

Case Report: A 37-year-old woman with SLE and antiphospholipid antibody syndrome experienced severe back pain followed by sudden paralysis and sensory loss below the T2 level. She remained alert and oriented on examination, with neurologic exam positive for diminished strength in the arms and with total loss of sensation and strength in the legs. Diagnostic workup was limited due to a contrast allergy and severe lupus nephritis; however, initial imaging showed increased cervical-thoracic spinal cord signal and concern for acute blood in the subarachnoid space. No neurosurgical intervention occurred, and the patient was treated with high-dose steroids and plasmapheresis for a possible transverse myelitis and non-aneurysmal subarachnoid hemorrhage. The patient received further neurologic and rheumatologic workup and remained neurologically stable, with improvement in proximal arm strength on physical exam.

Conclusions: We highlight the diagnostic challenges in treating a patient with SLE with acute paralysis and sensory loss. In this case, aggressive early treatment of the patient's myelitis and myelopathy were successful in leading to mild neurological improvement.

MeSH Keywords: Lupus Vasculitis, Central Nervous System • Myelitis, Transverse • Subarachnoid Hemorrhage

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/907001>

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Background

Isolated spinal artery subarachnoid hemorrhage (SAH) and transverse myelitis are rare occurrences in the general population but are considerably more common as neurologic complications of patients with systemic lupus erythematosus (SLE). In this report we present a patient with a history of SLE and antiphospholipid syndrome with an acute onset of spinal SAH followed by transverse myelitis. The etiologies and frequency of these complications of SLE are discussed as well as how to approach the diagnosis and treatment of these patients.

Case Report

We present the case of a 37-year-old African American female with a past medical history of SLE and antiphospholipid syndrome for 7 years, with multiple deep vein thromboses, pulmonary embolism, IVC filter, and 1 miscarriage secondary to hypercoagulability for which the patient was managed with Lovenox at baseline. Her chief complaint was acute-onset excruciating back pain characterized as sharp and occurring at 3: 00 PM as she ran up stairs, prompting hospital admission. Other associated symptoms included headache on the previous day with mild blurry vision. Upon hospital evaluation, she was given high-dose steroids to treat a presumed lupus nephritis due to the patient's back pain. At 11: 30 PM, Neurology was consulted due to sudden paralysis of the legs and arm weakness, with total loss of sensation below her chest. Physical exam revealed the patient to be afebrile and mildly hypertensive to 160/99, alert and oriented, with intact cranial nerves upon examination. The patient had diminished deltoid and biceps strength, with total loss of triceps, wrist, intrinsic hand muscle, and lower-extremity strength. Sensation was absent to vibration and pinprick below the T2 level. She also had mild hyperreflexia throughout the upper extremities. Notably, she also had rigors, and basic lab tests revealed an elevated WBC count, hypokalemia, creatinine of 3.23, INR of 1.0, and PTT of 95.

Initial imaging included CT and MRI, of the cervical and thoracic spine without contrast due to impaired renal function and a severe documented allergy to iodine (Figures 1, 2). The MRI without contrast demonstrated increased T2 signal throughout the C2 to mid-thoracic spine, with low T2 signal in the ventral thoracic subarachnoid space, which could represent SAH. Neurosurgery was consulted and decided against surgical intervention to evacuate blood due to low probability of cord compression and they agreed with medical management of an underlying subarachnoid hemorrhage or transverse myelitis. Medical management included high-dose steroids for 5 days, during which time the patient had improvements on physical exam including increased shoulder flexion and extension. At



Figure 1. CT scan revealed hyperdensity throughout the cervical and thoracic spinal canal surrounding the cord.

this point, a 5-day course of plasmapheresis with steroids was begun to further resolve an underlying inflammatory cord, but this treatment was stopped on the 9th clinical day due to lack of further clinical improvement. On the 7th clinical day, a lumbar puncture revealed xanthochromic CSF with 129 000 RBCs, protein 1471, IgG 310, 6 lymphocytes, 31 monocytes, and 63 segmented white blood cells, supporting inflammation and evidence of hemorrhage in the subarachnoid space.

Other imaging studies included an MRI and MRA of the head, which were negative for hemorrhage but revealed FLAIR hyperintensity in the subcortical white matter in the bilateral occipital lobes. This finding was consistent with a differential diagnosis of either lupus vasculitis or posterior reversible



Figure 2. MRI revealed increased T2 signal intensity within the central cord at the C2–T3 levels.

encephalopathy syndrome (PRES), which was less likely due to lack of chronic headaches, altered mental status, or visual changes. MRI of the lumbar spine was also performed and ruled out possible AV fistula causing a thoraco-lumbar ischemia. Overall, this clinical picture was consistent with an underlying spinal cord ischemia due to lupus vasculitis with a possible component of transverse myelitis. Rheumatology was consulted and a 3-month trial of cyclophosphamide (Cytoxan) was started, and a subsequent kidney biopsy revealed a suspected class 4 lupus nephritis with crescents. The patient was transferred to physical rehabilitation and was given 6-month (long-term) cyclophosphamide therapy to further resolve inflammation from lupus vasculitis and lupus nephritis, with close neurology follow-up.

Discussion

Lupus vasculitis

The incidence of stroke in lupus patients is greater than in the general population, but only 1–4% of SLE patients experience a SAH, which can be of aneurysmal or non-aneurysmal subtypes. SAH is associated with a worse prognosis than other forms of stroke and has a worse prognosis when occurring in younger patients with SLE [1].

Patients with SLE without other common risk factors for SAH have high mortality and frequently have no visible aneurysm, suggesting a non-aneurysmal SAH [2]. A worse prognosis was noted particularly in patients that had medical non-compliance to steroid therapy. Therefore, aggressive treatment of active lupus disease with steroids is of high priority to reduce the underlying disease process. Prior to the neurological event, the patient received outpatient prednisone and our initial treatment began with high-dose IV methylprednisolone, with which some neurological improvement was noted. Further treatment with plasmapheresis and Cytoxan therapy yielded no further clinical improvement until the time of discharge. This lack of further short-term clinical improvement is likely due to the irreversible nature of a myelopathy due to cord ischemia and was unrelated to the resolving spinal cord vasculitis.

Transverse myelitis

While neuropsychiatric symptoms such as headaches, seizures, and CVAs occur in up to 60% of patients with SLE, transverse myelitis is a very rare complication occurring in only 1–2% of patients [3]. Transverse myelitis involves rapidly progressive motor, sensory, and autonomic dysfunctions that can include loss of bowel and bladder control. In patients with SLE, the presence of antiphospholipid antibodies is associated with higher incidence of transverse myelitis, which could indicate that spinal cord ischemia or necrosis secondary to arterial thrombus is to blame for its development. However, data to support this theory has been sparse. A review by C. G. Katsiaria et al. showed that the use of anticoagulation in lupus patients with antiphospholipid syndrome has not been shown to decrease the incidence of transverse myelitis in these patients [4]. Indeed, the patient being discussed was on Lovenox at baseline and still experienced an episode of transverse myelitis, leaving the true cause of transverse myelitis unclear.

Patient management

SAH commonly presents with headache, altered mental status, focal neurological deficits, and seizure, with focal neurological deficits and seizure found more commonly in non-aneurysmal SAH than in aneurysmal SAH [5]. Etiologies of SAH can include PRES, coagulopathy, and vasculopathy. One reason why spinal hemorrhage is less frequently present with nuchal rigidity and thunderclap headache has been proposed by Spitzer et al. as either a smaller volume of blood or a non-arterial source of bleeding [6]. In our case, a backache occurred just prior to onset of neurological deficit with a mild headache 1 day prior to the event. This variable onset and severity of headache compared to the classic presentation of a SAH is consistent with a contained bleed.

While initial imaging of the cervical and thoracic spinal cord indicated SAH with possible transverse myelitis, further radiographic evaluation was limited due to the patient's inability to tolerate contrast. A spinal angiogram was not pursued due to severe renal dysfunction in the patient limiting further cervical or thoracic contrast imaging. Rather, this unique presentation showed that aggressive neurological management with anti-inflammatory and lupus treatment were effective for mild short-term improvement.

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Conclusions

When managing any patient with severe, acute-onset back pain, spinal SAH is an important part of the differential. A spinal SAH should particularly be considered as a possible sequela of SLE flare-up. Due to diagnostic limitations preventing a contrast CT scan or MRI in lupus nephritis and allergy, we treated this patient with high-dose anti-inflammatory and plasma exchange therapy. A treatment strategy that targets both vasculitis and myelitis is crucial to prevent progression of symptoms and achieve a mild improvement in neurological deficit.