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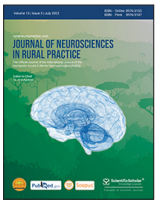

Case Report

Aneurysmal subarachnoid hemorrhage in a patient with dual autoimmune disorders. Perioperative challenges and management

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ABSTRACT

Autoimmune disorders (AIDs) are known to be associated with intracranial aneurysms; however, the coexistence of dual AIDs is a rare entity. Perioperative neuroanesthetic management of aneurysmal subarachnoid hemorrhage (aSAH) is typically complicated and challenging in such patients. In this report, we describe the successful management of a case of aSAH complicated by coexistent multiple sclerosis and systemic lupus erythematosus. A multidisciplinary team approach is warranted to manage such complicated cases.

Keywords: Aneurysm, Multiple sclerosis, Subarachnoid hemorrhage, Systemic lupus erythematosus

INTRODUCTION

Autoimmune disorders (AIDs) are known to be associated with intracranial aneurysms; however, the coexistence of dual AIDs is a rare entity. We describe the perioperative management for an intriguing case of aneurysmal subarachnoid hemorrhage (aSAH) in a middle-aged woman with concurrent multiple sclerosis (MS) who later also developed lupus nephritis while in the hospital. Written informed consent was taken from the patient for reporting this case for publication.

CASE REPORT

A 43-year-old debilitated lady presented with complaints of sudden onset severe headache associated with vomiting, seizures, and neck stiffness. Her initial Glasgow coma scale was 10/15 (E2V3M5). Computed tomography (CT) scan of the brain demonstrated SAH in the anterior interhemispheric fissure [Figure 1]. Her breathing was laborious; hence, tracheal intubation was done. Further, digital subtraction angiography revealed a rupture of the anterior communicating artery (ACoA) aneurysm. High-resolution CT scan of the thorax revealed mild pleural (bilateral) and pericardial effusions. She was a known case of MS for the past 6 years with extensive areas of demyelination in the brain [Figure 2] and spine. In addition, her bowel and bladder were also involved. However, she was not on any immunotherapy for MS due to financial constraints. She was also diabetic and hypertensive on medications.

In the operation theater, craniotomy and clipping of the ruptured ACoA aneurysm was done uneventfully under total intravenous anesthesia. The patient was then shifted to the intensive care unit (ICU) for further management. In the ICU, the patient developed lactic acidosis (lactate level increased from 3.3 to 7 mmol/L). The urine output was normal but serum creatinine increased from 1 to 2.2 mg/dL along with the rise in serum phosphate (5.2 mg/dL) and calcium levels (10.7 mg/dL). The pupillary reaction became sluggish. An urgent CT scan of the brain was done which showed no obvious postsurgical complication and the clip was *in situ*. The diagnosis of metabolic encephalopathy was made to correct the acidosis and normalize the metabolic milieu. All nephrotoxic drugs were discontinued. Few episodes of nystagmus were also noticed presumably due to non-convulsive status epilepticus. A neurology opinion was taken and the dose of anti-epileptic drugs was escalated. The metabolic acidosis improved gradually though there was no improvement in the sensorium. Sepsis was excluded by negative cultures, normal leucocyte count, and serum procalcitonin levels. Blood samples were also sent

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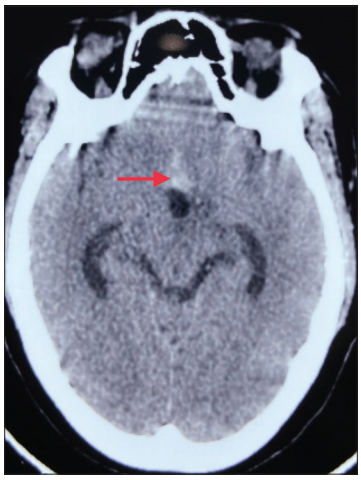

Figure 1: Non-contrast computerized tomography scan of the brain showing blood in the interhemispheric fissure suggestive of subarachnoid hemorrhage.

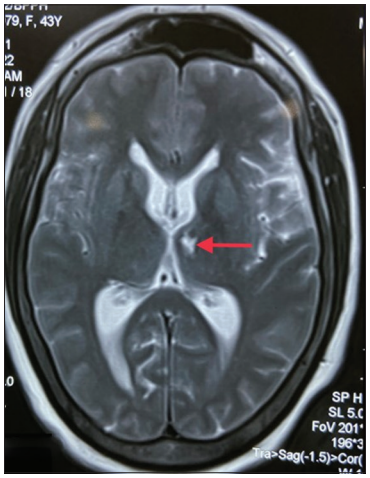

Figure 2: Non-contrast magnetic resonance imaging of the brain (axial, T2WI) showing demyelinating plaque in the left thalamic area.

to investigate autoimmune panel (antinuclear antibody, cytoplasmic anti-neutrophil cytoplasmic antibodies, perinuclear anti-neutrophil cytoplasmic antibodies, anti-double stranded DNA antibodies [anti-dsDNA], and complement levels). Ultrasound of the abdomen showed bilateral small kidneys with loss of corticomedullary differentiation.

The autoimmune panel was positive for anti-dsDNA (+++). Hence, a diagnosis of lupus nephritis superadded to MS was made. The patient was started on prednisolone 30 mg twice daily. An elective tracheostomy was also done. The patient showed a gradual response to the therapies and started to follow commands (M6). The patient was advised to start immunotherapy and thereafter discharged with an intact sensorium.

DISCUSSION

The coexistence of MS and systemic lupus erythematosus (SLE) is rare. Till date, only 17 cases have been reported in the literature.[1] Fanouriakis *et al.* reported nine cases of coexisting MS and SLE in the European subcontinent; generally, these patients were on immunotherapy and the less dominant AID was quiescent, unless there was a stress response.[2] There may be a possibility that the latent SLE got unmasked either by the aSAH or the stress of surgery or both. The prevalence of MS in India as reported by Singhal and Advani is estimated to be approximately 1.33/100,000.[3] It is important to determine if the neurologic lesions of the brain and spine were due to SLE or as a result of MS, because neurological manifestations of SLE can present years before the systemic manifestations.[4] In our patient, there were no prior manifestations of SLE as evident from the fact that she had a successful pregnancy without complications. Furthermore, she responded to disease modifying treatment with cyclophosphamide and steroids when she was first diagnosed with MS around 6 years back.

Intracranial aneurysms are not uncommon in patients with MS as well as in central nervous system (CNS) lupus; however, its association in a patient with both MS and SLE is rarely reported. MS is caused by immune cell infiltration across the blood–brain barrier, which promotes inflammation, demyelination, gliosis, and axonal degeneration of the white matter in the CNS. The pathogenesis includes molecular mimicry with myelin, vasculopathy, and autoimmune vasculitis.[1,2,4] Furthermore, there are reports of genetic correlation between MS and cerebral aneurysms.[5] On the other hand, SLE is a chronic autoimmune inflammatory disease with a wide spectrum of clinical and serological manifestations caused by autoantibody production, complement activation, and immune complex deposition. Collagen tissue alteration in SLE leads to the weakening of vessel walls. Furthermore, the increased prevalence of hypertension and dyslipidemia in SLE patients could increase the vulnerability to arterial dissection by atherogenesis and degenerative artery changes.[6]

Important anesthetic challenges in such cases include increased risk of aneurysmal re-bleeding, difficult airway due to underlying associated atlantoaxial dislocation, underlying myocarditis and pericarditis, deep vein thrombosis, and renal involvement in SLE.[2,4,6] In our case, we encountered mild pleural (bilateral) and pericardial effusions which did not warrant any intervention. Other major concerns include autonomic disturbances and

consequent exaggerated hypotension with vasodilators and drug-drug interactions. We used invasive arterial and central lines to monitor and administer vasopressors. Intraoperatively, we used propofol infusion for neuroprotection and systolic blood pressure was maintained at around 140–150 mmHg before clipping.

Adequate hydration should be ensured throughout the perioperative period. Titrated dose of induction agent should be used to prevent hypotension. There is also a need for stress dose steroids during induction as these patients receive long-term corticosteroids. Succinylcholine should be avoided due to the risk of hyperkalemia secondary to the proliferation of extrajunctional cholinergic receptors. For non-depolarizing neuromuscular blocking drugs (NMBDs), the lowest dose is advisable as the muscles become more sensitive due to wasting. Ideally, the use of NMBDs should be guided by neuromuscular monitoring. Pain should be adequately controlled using multi-modal analgesia. In poor grade aneurysms, it is always better to electively ventilate the lungs after the surgery until adequate neurological recovery has occurred.

CONCLUSION

The existence of dual AID is a rare occurrence; the latent disorder has the propensity to flare up under the effect of stressors. Ruptured aSAH should be secured as early as possible to prevent re-bleeding. The multidisciplinary team approach is warranted to manage such complicated cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Sánchez EC, Castillo MA, González VP, López FG, Díaz EP. Coexistence of systemic lupus erythematosus and multiple sclerosis. A case report and literature review. Mult Scler J Exp Transl Clin 2018;4:2055217318768330.
2. Fanouriakis A, Mastorodemos V, Pamfil C, Papadaki E, Sidiropoulos P, Plaitakis A, *et al*. Coexistence of systemic lupus erythematosus and multiple sclerosis: Prevalence, clinical characteristics, and natural history. Semin Arthritis Rheum 2014;43:751-8.
3. Singhal BS, Advani H. Multiple sclerosis in India: An overview. Ann Indian Acad Neurol 2015;18 Suppl 1:S2-5.
4. Kakati S, Barman B, Ahmed SU, Hussain M. Neurological manifestations in systemic lupus erythematosus: A single center study from northeast India. J Clin Diagn Res 2017;11:OC05-9.
5. Albrekkan FM, Bachir S, Jumaa MA, Zaidi SF, Medhkour A. Is there a genetic correlation between multiple sclerosis and cerebral aneurysms? World Neurosurg 2016;95:624.e1-4.
6. Prasad C, Khandelwal A, Patel S, Mulaokar MG, Chaturvedi A. Lupus-associated internal carotid artery dissecting aneurysm: An occult association. Ann Indian Acad Neurol 2021;24:276-7.

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