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Case Report

Multiple cerebral aneurysms: a rare presentation in systemic lupus erythematosus

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ABSTRACT

Systemic lupus erythematosus is a multisystem autoimmune disorder and its complications include cerebral vasculitis and vasculopathy which can be the first manifestation of SLE. Subarachnoid haemorrhage due to rapid aneurysm growth and rupture is a major neurosurgical emergency associated with significant morbidity and mortality. There is need for more rapid diagnosis and aggressive treatment of SLE patients with unruptured aneurysms. Authors report a case of 23 years old female, a newly diagnosed case of SLE complicated by rupture of cerebral aneurysms.

Keywords: Systemic lupus erythematosus, Aneurysms, Subarachnoid haemorrhage, Optic neuropathy, Anterior cerebral artery

INTRODUCTION

SLE is a chronic autoimmune disease with multiple pathologies that can affect every organ system of body including central nervous system (CNS).[1] CNS systemic lupus erythematosus (SLE) occurs in 24-51% of patients with psychosis and seizures which are among the diagnostic criteria of SLE.[2,3] Intracerebral aneurysms and subarachnoid haemorrhage (SAH) are one of the comparatively rare manifestations of CNS-SLE occurring in upto 3.9% of cases.[4]

CASE REPORT

A 23 years old female, mother of three normally delivered children was presented to emergency department with high grade fever, severe headache and projectile vomiting followed by clouding of consciousness with no significant past medical illness. On examination, patient was febrile, drowsy with hypotonia in left upper and lower limb with diminished reflexes and nystagmus towards left although Babinski and Hoffman's were negative. No signs of cranial nerve palsies and no signs of meningeal irritation were present. Bilateral pupils were sluggishly reacting to light. Other systems examination was normal other than nontender hepatomegaly. No signs of infective endocarditis were present. Patient was started on empirical antibiotic therapy till reports were awaited. Routine lab reports were all within normal limits with no evidence of infection and CSF analysis was also normal. On the third day of admission, patient became fully conscious with normal orientation but still was febrile with ataxia towards left side and complaining of diffuse headache and blurring of vision. MRI revealed multiple blooming in different areas with thrombotic infarct in left lateral medulla. CT angiography of brain vessels showed bilateral multiple saccular aneurysms in anterior cerebral artery and anterior communicating artery junction, right posterior cerebral artery, right superior cerebellar

artery, anterior cerebellar artery, bilateral posterior inferior cerebellar artery and internal carotid artery (ICA) with dolichoectasia (Figure1). Perimetry showed bilateral altitudinal visual field defects and fundus examination revealed bilateral optic neuropathy with disc oedema (Figure 2).

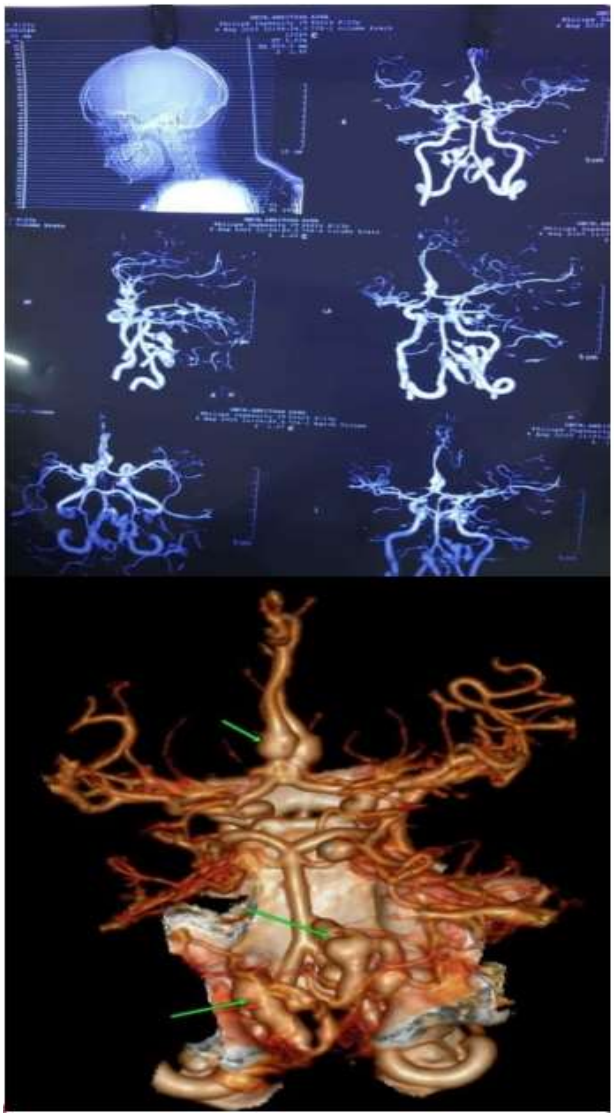

Figure1: CT angiography of patient demonstrates multiple bilateral saccular aneurysms in anterior and posterior circulation.

Autoimmune profile was sent in view of cerebral vasculitis leading to dilatation and aneurysm formation which showed strongly positive ANA (90.32 units; normal <20 units), anti Smith (23.28 units; normal <20 units) and U1RNP (81.05 units; normal <20 units) antibodies suggestive of SLE. In view of life threatening SLE, patient was started on pulse steroid therapy and cyclophosphamide. Despite that patient deteriorated and developed seizures with signs of meningeal irritation with NCCT head showing bilateral intraventricular haemorrhages due to ruptured aneurysm. Hence, she was referred for urgent neurosurgical intervention.

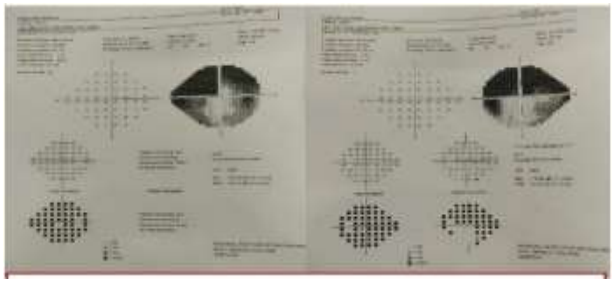

Figure 2: Perimetry of patient shows bilateral altitudinal visual field defects.

DISCUSSION

The cerebral aneurysms in SLE patients are more common in younger age group compared with the normal population and tend to appear as multiple with rapid growth.[5] There is higher incidence of SAH in SLE due to lupus vasculitis or fragility of vessels. Various contributors such as coagulopathy, angiopathy due to premature atherosclerosis and hypertension are also considered. These patients bear worse prognosis, increased mortality and worse HUNT and HESS grades on presentation of SAH.[6,7] Risk factors for rupture includes location on ICA, middle cerebral artery or basilar artery, large size, multiple lobes, family history of SAH, smoking and female sex.[8] The greatest risk of rupture occurs during rapid growth with inflammation and ischemia produces endothelial remodeling and increases aneurysm growth via collagen and fibrin deposition in walls with vascular smooth muscle deficiency and elevated elastase and collagenase activity.[9] The natural history of aneurysm rupture is characterized by periods of intermittent, unpredictable vulnerability.[10]

CONCLUSION

The rare initial presentation of SLE with rapid growth and rupture of aneurysm in this case study highlights major deficiencies in current understanding of relationship between SLE and aneurysm biology. It should be considered in patients presenting with fever, headache and other neurological symptoms. They should be treated aggressively with endovascular embolization or open surgical clipping.

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