

Multiple cerebral aneurysms in lupus

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Abstract

Systemic lupus erythematosus (SLE) is an autoimmune chronic multisystemic inflammatory disease of unknown etiology. It has variable course and prognosis. Its clinical manifestations may be constitutional or specific to organ/system involved. Immunological aberrations are the cause for inflammation in various tissues. Nervous system involvement in SLE is common and has diverse clinical and morphological manifestations. Diagnosis of neuropsychiatric involvement remains difficult in SLE. Intracranial vasculitis is rare and results in cerebrovascular accidents (CVA). Young patients are at risk of CVA. Neuropathological features on biopsy include infarcts characterized by fibrinoid necrosis of small intracranial arterioles and capillaries. Fibrinoid necrosis, a common histological finding in SLE, may be responsible for aneurysmal formation. The increased risk and associated mechanisms of SAH in SLE is less well understood. The prevalence of SAH due to rupture of intracranial aneurysms is higher in SLE patients than in the general population, a phenomenon that is thought to be due to focal transmural lupus angitis causing rupture. SAH secondary to aneurysmal bleed is very rare and requires prompt immunosuppression for better outcome.

Keywords: subarachnoid hemorrhage (SLE); lupus; cerebral aneurysms; cerebral vasculitis; neuropsychiatric lupus

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Introduction

Systemic lupus erythematosus (SLE) is an autoimmune chronic multisystemic inflammatory disease of unknown etiology. It has variable course and prognosis. Its clinical manifestations may be constitutional or specific to organ/ system involved. Nervous system involvement in SLE is common and has diverse clinical and morphological manifestations. Fibrinoid necrosis, a common histological finding in neuropsychiatric lupus, may be responsible for aneurysmal formation. The prevalence of subarachnoid haemorrhage (SAH) due to rupture of intracranial aneurysms is higher in SLE patients than in the general population. SAH secondary to aneurysmal bleed is very rare and requires prompt

immunosuppression for better outcome rather than emergency surgical intervention.

Case report

A 22-year old female, diagnosed as Systemic lupus erythematosus (SLE) in May, 2012, presented to us with erythematous macules over soles bilaterally, mononeuritis multiplex, left optic neuritis, left sixth cranial nerve palsy with positive antinuclear antigen and anti dsDNA antibodies. She had nephritis evidenced by WBC and granular casts, hematuria, 24 hour urine protein of 1.5gm, and hypertension. Serum for anti-cardiolipin antibodies and lupus anticoagulant was negative. In view of active disease she was given pulse steroids followed by cyclophosphamide. Her vision, sensory symptoms, motor weakness improved during the hospital stay. Two months later she presented to emergency with one day history of sudden onset of severe headache, convulsions, vomiting with no fever. Clinical examination revealed post ictal confusion with tachycardia, blood pressure of 150/90. Her cardiovascular, respiratory, gastrointestinal system examination was unremarkable. She had neck stiffness. Fundus examination revealed bilateral

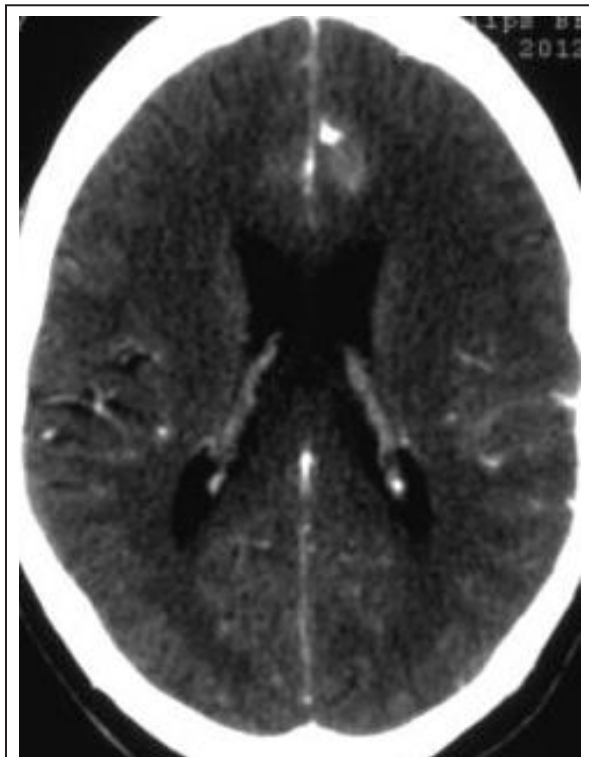


Figure 1: CT brain showing frontal and parasagittal hyperintensities s/o SAH.

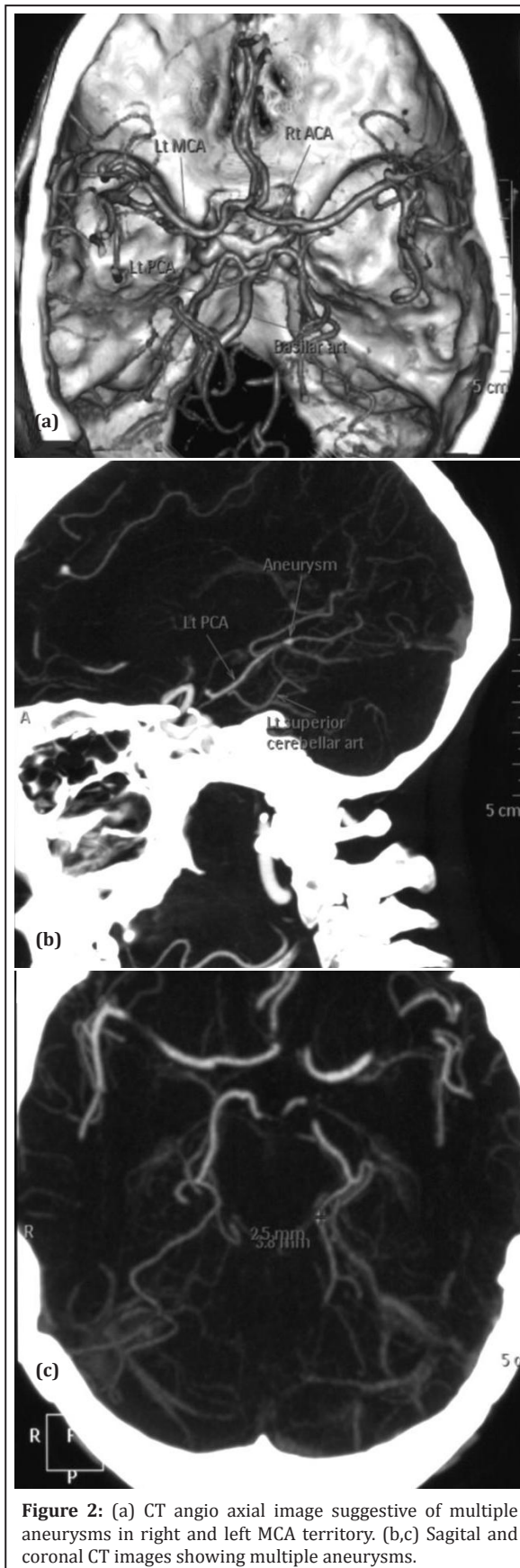


Figure 2: (a) CT angio axial image suggestive of multiple aneurysms in right and left MCA territory. (b,c) Sagittal and coronal CT images showing multiple aneurysms.

papilledema. Her deep tendon reflexes were normal. CT brain (Figure 1) showed frontal and parasagittal hyperintensities which were focal on left side, s/o subarachnoid haemorrhage. CT angiography (Figure 2a, b) revealed multiple aneurysms in left anterior cerebral artery, posterior cerebral artery, right middle cerebral artery, with bleeding from distal anterior cerebral artery aneurysm. Cerebral vasculitis secondary to active lupus was considered and planned for immediate ligation of aneurysms along with immunosuppression. In view of multiple aneurysms and as none of them was easily accessible surgically, pulse steroids were given. Her consciousness improved gradually. As she was on immunosuppression with cyclophosphamide, it was planned to continue the same along with high dose steroid. She has recovered from the crises and is reported to be asymptomatic now.

Discussion

In adults, manifestations of neuropsychiatric systemic lupus erythematosus (NPSLE) can develop before or around the time of the diagnosis of SLE in up to 28%-40% [1]. The pathogenic aetiologies of NPSLE manifestations are multifactorial which include autoantibody production, microangiopathy, intrathecal production of proinflammatory cytokines and premature atherosclerosis [2]. Fibrinoid necrosis, a common histological finding in SLE, may be responsible for aneurysmal formation, since it produces local weakness in the walls of small arteries. Aneurysmal dilatation of cerebral arteries is due to vasculitis. Subarachnoid haemorrhage (SAH) is a rare but associated complication of SLE with a high mortality rate. The increased risk and associated mechanisms of SAH in SLE is less well understood. The prevalence of SAH due to rupture of intracranial aneurysms is higher in SLE patients than in the general population, a phenomenon that is thought to be due to focal transmural lupus angitis causing rupture [3]. SAH secondary to aneurysmal bleed is very rare in SLE as occurred in our patient. Aneurysm with SAH has varying outcomes. In seriously ill cases, immediate treatment with immunosuppressants is essential. Neuropsychiatric events have high morbidity and mortality and warrant aggressive management [4]. Our patient was immediately started on high dose steroids along with cyclophosphamide which has led to dramatic clinical improvement.

Conclusion

Neuropsychiatric events, one of the major complications of SLE, have high morbidity and mortality and warrant aggressive management. Aneurysms resulting from vasculitis can rupture and lead to mild to massive haemorrhage associated with high mortality. Early intervention may benefit the patient with good outcome.

Acknowledgements

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Conflicts of interest

Authors declare no conflicts of interest.

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