Central Nervous System Lupus: Case Report

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Abstract

Cerebral infarction is a presentation of systemic lupus erythematosus (SLE) and generally appears during advanced, active phases of the disease. A thirty-two year old female was admitted to hospital with a history of diarrhea, agitation, headache and left hemiparesis. On physical examination her body temperature was high, lymphadenopathy and pretibial edema were noticed. Cranial diffusion weighted images were performed and acute bilateral disseminated multiple cortical and subcortical areas of high signal intensities on T2 and T1-weighted images compatible with vasculitis were detected. On laboratory tests, low complement levels, thrombocytopenia, neutropenia, lymphopenia, positive antinuclear antibodies (ANA) (1/320), persistently elevated anti-DNA titres and proteinuria were found. Renal biopsy confirmed lupus nephritis. Chest X-ray showed cardiomegaly and pleural effusion. This case presentation shows evidence of ischemia in patients with active SLE and also shows that the first symptom of SLE can be an acute ischemic stroke.

Introduction

Stroke is an uncommon complication of SLE, attributed to many different etiopathogenetic mechanisms. Strokes have been reported in up to 19% of patients with SLE (1, 2). Cerebral infarction in relation to SLE is low and generally appears during advanced, active phases of the disease. Presentation with neurological features at the onset of the disease is regarded as rare, occurring only in approximately 3% of the patients (3).

Case Report

A thirty-two year old female presented with a history of diarrhea, agitation, headache and left hemiparesis that significantly improved in a week during hospitalization. Physical examination revealed sinus tachycardia (105/min), hypertension (200/95 mm Hg) and pretibial edema. On neurological examination she was confused, agitated and partially cooperating with an altered mental state and bilateral hyperactive deep tendon reflexes. Motor examination revealed left-sided hemiparesis (MRC grade 3). Computed tomography of brain showed patchy low attenuation areas in the right temporal area and close to the frontal horn of lateral ventricle (Figure 1). Cranial MRI scans revealed acute bilateral disseminated multiple cortical and subcortical areas of high signal intensities on diffusion MR images compatible with vasculitis (Figure 2). MR-venography and cervical MR-angiography were normal. Neuroimaging changes were confined to the bilateral cerebellum, cortical, subcortical and deep white matter with multiple hyperintense signals suggesting acute infarctions due to vasculitis (Figure 3).

Laboratory evaluations showed proteinuria (protein/creatinine ratio> 45 mg/mmol), high serum creatinine level (2.8 mg/dL), telescopic urine sediment findings, thrombocytopenia (52000/mm³) and lymphopenia (52000/mm³). Serological testing was positive for antinuclear and anti-DNA antibodies. Complement levels of C3 (0.776, 0.75-1.4) and C4 (0.066, 0.1-0.34) were low. Antiphospholipid antibody (APA) titer was within normal limits and lupus anticoagulant assay was negative/normal. Transthoracic echocardiography showed dilated left atrium, thickened calcified mitral valve, severe mitral insufficiency and excluded infective endocarditis. The patient was complaining of dyspnea and coughs for nearly a month. Chest X-ray showed cardiomegaly and pleural effusion. EEG and CSF analysis was unremarkable, and CSF oligoclonal bands were absent.

In view of her neurological status and evidence of active SLE, she was diagnosed as having definite SLE on clinical and serologic grounds with fever, lymphadenopathy, and photosensitivity, a malar rash and proteinuria with renal functional impairment which gradually progressed to the degree of renal replacement therapy requirement. Daily plasmaphresis with 15ml/kg plasma exchange for five consecutive days and every other day thereafter was applied simultaneously with intravenous pulse prednisolone 1 g/day for 3 days, followed by maintenance dose of 1 mg/kg/day gradually tapered to 10 mg at about 3 months and cyclophosphamide 750 mg/day monthly infusions. Antihypertensive therapy was started initially. Her mental status dramatically improved following therapy. At discharge one month later, she had near-total recovery and she was off dialysis.

She received various immunosuppressant drugs for treatment of lupus nephritis, including pulsed...
methylprednisolone, oral prednisolone, hydroxychloroquine, and azathioprine with moderate response. A beneficial response was only eventually seen with mycophenolate treatment which appeared to improve renal function and systemic disease more than CNS features.

Discussion

SLE is a syndrome of variable clinical and immunological expression, which may affect patients at any age. SLE has a tendency for widespread organ involvement. The autoimmune disregulation plays probably a central role in the pathogenesis of SLE but the molecular targets and the mechanisms involved in brain damage have not been fully elucidated. Diagnostic criteria of SLE is based on American College of Rheumatology criteria (4). Although these criteria do not necessarily happen simultaneously, they may occur at any time during the disease course. Neurologic and psychiatric symptoms occur in 10 to 80% of patients either prior to the diagnosis of SLE or during the course of their illness (5). The psychiatric aspects of this disorder are anxiety, cognitive dysfunction, mood disorders and psychosis. Neurologic manifestations of SLE may be cognitive dysfunction; headache, cerebrovascular events, seizures and polyneuropathy. Nervous system involvement in SLE was initially thought to be due to vasculitis. The pathogenesis of the vasculopathy and the vasculitis are not known. Certain autoantibodies have been associated with different aspects of CNS lupus but not with the vascular disease itself. Stroke is attributed to many different etiopathogenetic mechanisms in SLE. Direct injury due to vasculopathy may affect blood-brain barrier, thereby allowing antibodies enter the nervous system. Patients with SLE have an increased risk of stroke. SLE is generally considered in the differential diagnosis of stroke in young patients and occurs as an etiology in 3.5% of stroke before the age of 45 (6). SLE is relatively an uncommon etiology of stroke and the incidence of recurrence in patients with SLE is much higher than other stroke patients (7). Cerebral infarction in relation to SLE occurs rarely and generally appears during advanced, active phases of disease. Strokes have been reported in up to 19% of patients with SLE (1,2) and may involve small, medium, or large vessels by a variety of mechanisms. Baseline disease activity, hyperlipidemia and hypertension are predictive factors for ischemic stroke and stroke severity in systemic lupus erythematosus (2). Strokes may be ischemic or hemorrhagic due to thrombocytopenia. Antiphospholipid antibodies also appear to increase the risk of stroke syndromes (8). As an example, hypertension and accelerated atherosclerosis, both associated with chronic steroid therapy and valvular heart disease due to SLE are common risk factors for stroke (9). Elevated plasma homocysteine levels have been identified as a risk factor for stroke and other thrombotic events in patients with SLE (10). Most strokes occurred within the first five years of illness. Prospective studies suggest that from 50 to 78% of neurologic episodes are caused by secondary factors (5,11), including infections associated with immunosuppressive therapy, metabolic complications of other organ system failure, such as uremia, hypertension, toxic effects of therapy (particularly corticosteroids).

Conclusion

As a conclusion, SLE may be associated with a significant increase in the risk of stroke and of premature death due to cerebrovascular disease. Represented case with SLE presented without clinically suspcible symptoms of stroke but found to have stroke after neurological assessment and allowed identifying already known but also rare clinical picture in SLE.

References

Archives of Neurology 1995; 52: 491- 495.
Illustrations

Illustration 1

Unenhanced cranial CT on admission: showing patchy low attenuation areas in the right temporal area and close to the frontal horn of lateral ventricle.

Illustration 2

Diffusion weighted MRI on admission: showing bilateral disseminated multiple cortical and subcortical areas of high signal intensities on diffusion MR images compatible with vasculitis.
Illustration 3

T2 and Flair-weighted images showing subcortical areas of high signal intensities compatible with vasculitis.
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