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Case Report

A case of systemic lupus erythematosus presenting with intracerebral hemorrhage: A rare entity

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Abstract

Systemic lupus erythematosus (SLE) is an autoimmune disorder of the connective tissue affecting multiple organs. Neurological involvement in SLE affects both the central and peripheral nervous system. Stroke is a common neurological presentation, including arterial and venous infarcts, and less commonly intracerebral hemorrhage (ICH). ICH presents with sudden onset headache, vomiting, focal limb weakness, seizure, altered sensorium, and coma. In this case report, we describe the clinical profile, laboratory work-up, treatment and outcome of a 35 year old male patient suffering from SLE, presenting with seizure and focal neurological deficits. Considering to evaluate a treatable cause like SLE is prudent in patients with stroke in young.

Keywords: Systemic lupus erythematosus, Intracerebral hemorrhage, Seizure, Outcome.

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1. Introduction

Systemic lupus erythematosus (SLE) is an autoimmune vasculitic disorder affecting multiple organs with protean manifestations. Neurological involvement is seen in 40 % SLE.[1] Intracerebral hemorrhage (ICH) is rare in SLE.[2] The incidence of cerebrovascular events are increasing in SLE.[3] In this case, we discuss the clinic-laboratory work-up and outcome of a young patient with stroke and its association with SLE.

2. Case Description

This 35 year non-hypertensive, non-smoker, non-alcoholic, male presented with severe headache, and sudden onset weakness of left side of the body for 1 day. There was no history of vomiting, altered sensorium, convulsion, blurring of vision, sensory complaints, difficulty in speech or swallowing, urinary or bowel difficulty. He had past history of multiple episodes of generalised tonic-clonic seizures 11 months back, along with history of acute-onset paraplegia with bladder involvement 6 months back. He was diagnosed as neurological-SLE based on antinuclear antibody (3+), anti-ds-DNA positivity, and short segment transverse myelitis and started on intravenous methyl prednisolone 1 gram/day for 5 days, followed by prednisolone 40 mg/day with gradual tapering upto 10 mg/day, levetiracetam 500 mg/day and immunomodulatory therapy (hydroxychloroquine 200 mg twice daily). At that time his brain imaging and electroencephalogram were normal. There was no history of trauma, head injury, use of blood thinners, recent history of significant weight loss, fever, hemoptysis, jaundice or significant travel history. Subcortical white matter changes, multiple small subcortical hemorrhages with normal angiogram and venogram.

General examination revealed pulse 82/min, blood pressure 108/70 mm Hg, respiratory rate 15 beats/min, temperature 97.6 ^{0}F, without rash, joint tenderness, pallor, icterus, edema, significant lymphadenopathy. Neurological examination showed GCS E4V5M6 with intact higher functions, normal cranial nerve examination including fundus, power of 4/5 on left and 5/5 on right side, hyperreflexia on left with normal reflexes on right side with bilateral extensor plantars, intact sensory and cerebellar functions. There was no carotid or renal bruit audible.

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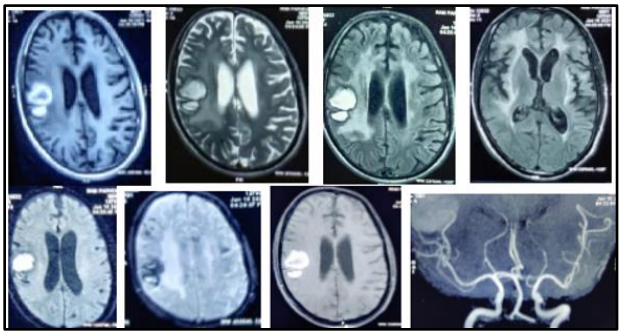

Figure 1: Brain imaging suggestive of right-sided frontal bleed with surrounding edema, symmetrical frontal

Blood investigations including complete hemogram, blood glucose, renal function test, liver function test, thyroid function test, serum electrolyte, serological tests for HBsAg, anti HCV, HIV-1,2 were normal, urine showed 50 RBC/HPF in absence of infection. His C reactive protein level were elevated (12 mg/L). The coagulation profile including Prothrombin time (PT), Internationalised Ratio (INR), activated tissue thromboplastin time (aPTT), bleeding time and clotting time were within normal range values. Renal and chest imaging were normal. Electrocardiogram and 2D trans-thoracic echocardiogram were normal. Brain imaging revealed right-sided frontal lobar bleed with surrounding edema, symmetrical frontal subcortical white matter changes, multiple small subcortical hemorrhages with normal angiogram and venogram.(**Figure 1**) The carotid and renal doppler studies were normal.

Differentials included hypertensive disorders, coagulative disorders, drugs causing thrombocytopenia or bleeding diasthesis, ruptured aneurysm, arterio-venous malformation of the brain, cerebral venous thrombosis, haemorrhagic neuroinfections, trauma (head injury), vasculitis (primary or secondary), etc.

He was managed with pulse therapy of methyl prednisolone (1 g/day for 5 days), followed by prednisolone (40 mg/day), azathioprine (25 mg twice daily) and anti-seizure medication (levetiracetam 500 mg/day). Patient improved with medications over a week and was discharged within 10 days of admission. Physiotherapy was advised in addition to dietary support for nutrition. On follow-up at 1 month, she regained back his normal power, and was able to her daily work independently.

3. Discussion

SLE is a common immunological disease with diverse presentations. SLE can involve renal, skin, cardiac, respiratory, immune, locomotor, gastrointestinal, ocular and nervous system with guarded prognosis. Neuro-lupus presents with diverse manifestations of the central (headache, seizure, stroke, cognitive deficits, optic neuropathy, movement disorders, myelopathy), and peripheral nervous system (peripheral neuropathy, and cranial neuropathy). Similar manifestations can be observed in neuro-infections like tuberculosis, dengue infection, HIV, neurosyphilis, Hansen's disease, and neuro-inflammatory disorders like neurosarcoidosis.

Stroke in SLE is invariably ischaemic due to endothelial injury-vasculitis. However, cerebral hemorrhage can also be seen, the exact aetiopathogenesis of which is not properly known.[4] It might be postulated that vasculitis-related vessel wall damage and vasomotor tone dysregulation might precipitate such events in SLE. In addition, the use of immunomodulatory drugs like steroids might lead to sodium and water retention and cerebral intravascular hypertension in already weakened vessels leading to sudden rupture and hemmorrhage.[5] A thorough study of cerebral autoregulation and vessel integrity in SLE can enlighten concepts in future.

The management of ICH in SLE depends upon the severity of the lesion, site of involvement, signs of raised intracranial pressure and cerebral herniation, and importantly the disease activity status. Supportive treatment in the form of cerebral decongestants, hydration, blood pressure control, anti-seizure medications (in seizure), nutrition, and limb physiotherapy help in the management and better outcome in ICH.[6] In refractory cases, there is requirement of surgical procedures like decompressive hemicraniectomy, evacuation of hematoma, and ventriculo-peritoneal shunting in hydrocephalus. In certain studies, an upgradation of immunotherapy in the form of cyclophosphamide, and rituximab have been tried successfully in SLE flare.

Another important association in such presentations of proven SLE presenting with ICH is Anti-phospholipid antibody syndrome, which should be kept into consideration.[7-8] Interestingly, ICH can be the heralding manifestation of SLE, and hence lies the importance of a consideration of SLE in ICH. [9]

4. Conclusion

SLE is a multi-systemic inflammatory disorder capable of inflicting stroke in young people. ICH is an uncommon presentation in SLE with good outcome if treated timely. A high index of clinical suspicion and diagnostic work-up is essential in treating stroke in young patients. Furthermore, one should be aware of the associations of SLE with similar presentation like Anti-phospholipid antibody syndrome.

5. Source of Funding

None.

6. Conflict of Interest

None.

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