CASE REPORT

Cerebral venous thrombosis presenting with subdural haematoma as first presentation for systemic lupus erythematosus with negative antiphospholipid antibodies

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SUMMARY

We report the case of a 30-year-old woman, without any previous comorbidities presenting with acute onset headache, altered sensorium and unsteadiness of gait. Neurological evaluation revealed a drowsy patient with papilloedema, bilateral lateral rectus palsy, generalised hyper-reflexia and up going plantar responses. Urgent imaging performed showed extensive cortical venous sinus thrombosis. Workup for secondary causes of cortical venous sinus thrombosis revealed very high titres of antinuclear antibody and anti-dsDNA, but negative antiphospholipid antibodies (APLA). In hospital she started developing other complications of systemic lupus erythematosus (SLE). Urine evaluation revealed proteinuria and granular casts suggestive of glomerulonephritis. Cardiac evaluation revealed moderate pericardial effusion. We have discussed neurolupus as initial presentation of SLE and the rare occurrence of major neurovascular complications without secondary APLA syndrome.

BACKGROUND

Dural venous sinus thrombosis is a rare complication of systemic lupus erythematosus (SLE)-associated hypercoagulability and is often seen in association with antiphospholipid antibodies. SLE presenting as cerebral venous thrombosis (CVT) without secondary antiphospholipid antibodies (APLA) is quite rare. To the best of our knowledge, there are only three cases of SLE reported, with CVT presentation.¹⁻⁴ Cortical venous sinus thrombosis usually presents with headache, seizures, altered sensorium and focal neurological deficits. The most common neuropsychiatric manifestation of SLE is organic encephalopathy. Since the symptoms overlap, coexistent SLE encephalopathy is difficult to diagnose in the presence of CVT. Failure to consider, diagnose and treat this condition promptly may result in long-term neurological disability or death.

CASE PRESENTATION

A 30-year-old woman presented to the emergency department with 2 weeks history of holocranial throbbing headache associated with projectile vomiting, nocturnal worsening and non-responsive to analgesics. She also had progressive deterioration sensorium in the form of lethargy and irritability along with binocular horizontal diplopia, blurring of vision and unsteadiness of gait 2 days prior to the day of presentation. There was no history of weakness or

sensory symptoms in the limbs, faciobulbar symptoms, sphincter incontinence, loss of consciousness or seizures. There was no history of fever or features suggestive of vasculitis. There was no history of deep vein thrombosis or abortions in the past.

INVESTIGATIONS

Urgent CT of the brain performed in emergency showed hyperdensity in the region of straight sinus, Vein of Galen and internal cerebral veins, left transverse sinus, in addition to subdural haematoma (figure 1). Possibility of CVT appeared most likely and CT cerebral venography confirmed the same (figure 2). Which showed in three dimensional venogram (figure 3). Baseline blood investigations revealed normocytic normochromic anaemia, normal liver and renal functions. Vasculitic and thrombophilia work up was positive for antinuclear antibody and anti-dsDNA in very high titres, APLA was negative, rest of the thrombophilia screening was negative.

DIFFERENTIAL DIAGNOSIS

- ▶ CVT
- ▶ Subdural haematoma
- ▶ SLE

Figure 1 CT of the brain: hyperdensity in the region of straight sinus, vein of Galen and internal cerebral veins and subdural haematoma.

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Figure 2 CT venogram: superior sagittal and straight sinuses filling defects.

TREATMENT

The patient was started on therapeutic anti-coagulation with heparin with maintaining activated partial thromboplastin time between 60 and 90. However, she started having seizures with deterioration of her sensorium. Repeat imaging showed extension of thrombosis and hence mechanical clot removal was attempted, though unsuccessful. She was continued on heparin followed by warfarin along with anti-epileptic drugs and supportive care. In intensive care unit she started having moderate grade fever with persistent sinus tachycardia disproportionate to the degree of fever. Echocardiography showed moderate pericardial effusion. Initial routine urine profile was normal, but after a week of admission she started having proteinuria with granular casts in urine. Septic work up was negative.

In view of multiple major manifestations of SLE, in consensus with the rheumatology team, she was given pulse dose of intravenous methylprednisolone, followed by oral steroids and azathioprine. Following the steroid pulse, fever and tachycardia subsided, her sensorium improved to Glasgow Coma Scale score

Figure 3 Three-dimensional venogram: showing filling defect at the superior sagittal sinus, vein of Galen.

of 10. However, she contracted ventilator associated pneumonia and urinary sepsis because of which pulse cyclophosphamide was not started. Within a few weeks, she again started having tachycardia and fever with worsening of proteinuria.

OUTCOME AND FOLLOW-UP

Her neurological status remained the same despite adequate anticoagulation. She was weaned off ventilator after a month but had spastic quadriparesis as sequelae. She continued to have systemic manifestations of SLE in the form of proteinuria, myocarditis and developed punched out lesions in the inner aspect of the thigh typical of vasculitis. After augmenting immunotherapy, myocarditis and vasculitis ulcers improved as did her sensorium. She remains conscious and alert, but with spastic quadriparesis and incontinent sphincters.

DISCUSSION

SLE may be associated with a high incidence of cerebrovascular disease, and upto 19% of patients with SLE may have arterial stroke.⁵ However, venous sinus thrombosis is a rare manifestation of SLE-associated hypercoagulability and is often seen in association with antiphospholipid antibodies.⁶ There are only a very few case reports where SLE presented as cerebral venous sinus thrombosis without associated APLA.^{3–6} Inhibition of vascular tissue prostacyclin production, antiphospholipid (including anticardiolipin) activity,⁷ prekallikrein inhibition,⁸ changes in antithrombin III function, platelet activity and aggregation⁹ and direct injury to the vessel wall through an antibody antigen complex are the proposed mechanisms of thrombotic tendency in SLE.¹⁰

Clinical and neuroimaging features of CVT in patients with SLE are similar to those of patients with cerebral venous thrombosis of other origins.

In a recent Iranian study they reported the incidence of CVT associated with SLE is 6.6%, which is higher compared to normal population.¹¹ It was also reported that some patients with SLE have special tendency to involve cerebral vessels, which is not related to the duration of SLE, but the nature of SLE.¹²

MR venous angiography, CT venogram or conventional angiography are the recommended imaging modalities for the diagnosis of cerebral venous sinus thrombosis. Cerebral oedema or haemorrhagic venous infarcts may be obvious on CT scans. MR venous aids direct visualisation of thrombus within a vessel. Venous infarctions and haemorrhagic foci can be seen with gradient-echo or susceptibility-weighted images. Susceptibility-induced signal loss from deoxyhaemoglobin provides a basis for detection of even small foci of haemorrhage, which tend to occur in the subcortical white matter, thalami and basal ganglia. Rarely subdural haematoma may occur consequent to CVT.

Aggressive management of CVT and SLE simultaneously is recommended to improve outcomes as the expected rate of neurological complications with both is very high.

Learning points

- ▶ Neurological involvement is a very important complication of systemic lupus erythematosus (SLE).
- ▶ Cerebral venous thrombosis is a rare complication of SLE, especially as first presentation.
- ▶ Cerebral venous thrombosis is a treatable cause of death and morbidity and should be in our mind in any patient with SLE with neurological involvement.

Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Vidailhet M, Piette JC, Wechsler B, *et al.* Cerebral venous thrombosis in systemic lupus erythematosus. *Stroke* 1990;21:1226–31.
- 2 Shiozawa Z, Yoshida M, Kobayashi K, *et al.* Superior sagittal sinus thrombosis and systemic lupus erythematosus (letter). *Ann Neurol* 1986;20:272.
- 3 Cardona-Portela P, Casasnovas-Pons C, Moral-Torres M, *et al.* Servicio de Neurología, Hospital de Bellvitge, L'Hospitalet de Llobregat, Barcelona, Spain. Cerebral venous thrombosis as the presenting symptom of systemic lupus erythematosus. *Rev Neurol* 2004;39:30–4.
- 4 Agreda-Vásquez GP, Galván-Plata ME, Nellen-Hummel H, *et al.* [Cerebral venous thrombosis in systemic lupus erythematosus. Case report and literature review]. *Rev Med Inst Mex Seguro Soc* 2006;44:365–9.
- 5 Mikdashi J, Handwerker B, Langenberg P, *et al.* Baseline disease activity, hyperlipidemia, and hypertension are predictive factors for ischemic stroke and stroke severity in systemic lupus erythematosus. *Stroke* 2007;8:281.
- 6 Merkel PA, Chang Y, Pierangeli SS, *et al.* The prevalence and clinical associations of anticardiolipin antibodies in a large inception cohort of patients with connective tissue diseases. *Am J Med* 1996;101:576–83.
- 7 Thiagarajan P, Shapiro SS, Demarco L. Monoclonal immuno- globulin M-X coagulation inhibitor with phospholipid specific- ity. Mechanism of a lupus anticoagulant. *J Clin Invest* 1980;66:397–405.
- 8 Sanfelippo MJ, Drayna CJ. Prekallikrein inhibition associated with the lupus anticoagulant. *Am J Clin Pathol* 1982;77:275–9.
- 9 Harris EN, Gharavi AE, Hughes GRV. Antiphospholipid anti-bodies. *Clin Rheum Dis* 1985;11:591–609.
- 10 LeRoux G, Wautier M-P, Guillemin L, *et al.* IgG binding to endothelial cells in systemic lupus erythematosus. *Thromb Haemost* 1986; 56:144–6.
- 11 Saadatnia M, Sayed-Bonakdar Z, Mohammad-Sharifi G, *et al.* Prevalence and prognosis of cerebrovascular accidents and its subtypes among patients with systemic lupus erythematosus in Isfahan, Iran: a hospital clinic-based study. *Int J Prev Med* 2014;5:123–6.
- 12 Ferro JM, Correia M, Pontes C, *et al.* Cerebral Venous Thrombosis Portuguese Collaborative Study Group (Venoport) cerebral vein and dural sinus thrombosis in Portugal: 1980-1998. *Cerebrovasc Dis* 2001;11:177–82.

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