Case Report

Superior sagittal sinus thrombosis in a patient with systemic lupus erythematosus

D Flusser¹, M Abu-Shakra¹, A Baumgarten-Kleiner¹, G Flusser² and S Sukenik¹

¹ Department of Medicine D, Soroka Medical Center, Faculty of the Health Sciences, Ben-Gurion University of the Negev, Beer-Sheva, Israel; ² Department of Radiology, Shaarei Zedek Medical Center, Jerusalem, Israel

Cerebral venous thrombosis is a rare cause of focal neurological damage in patients with SLE. We describe a patient with SLE who presented with a focal neurological deficit and subsequently developed increased intracranial pressure. A superior sagittal sinus thrombosis was diagnosed by MRI. The association between SLE and superior sagittal sinus thrombosis is discussed. Early diagnosis is essential because treatment leads to a positive clinical outcome.

Keywords: venous thrombosis; SLE; MRI; superior sagittal sinus thrombosis

Introduction

Thrombotic events, including arterial and venous thromboses, occur in 10-20% of patients with systemic lupus erythematosus (SLE)¹. The majority of cases with venous thrombosis occur in the deep venous system of the extremities, although thrombosis of vena cava, renal, mesenteric and other veins has been reported. Superior saggital sinus thrombosis (SSST) rarely occurs in patients with SLE^{2-6} . The incidence of thrombosis in patients with SLE correlates strongly with the presence of lupus anti-coagulant^{7,8}. We describe a patient with SLE who presented with a focal neurological deficit and subsequently developed increased intracranial pressure and SSST. The association between SLE and SSST is discussed.

Case history

A 40-year-old woman was admitted to the hospital with right hemiparesis and dysphasia. She had been diagnosed as suffering from SLE four years earlier. The diagnosis was based on polyarthritis of the small joints of the hands, nephritis (mild proteinuria and red blood cell casts), a positive serological test for antinuclear antibody (ANA), high titers of anti-DNA and low levels of complement. She was treated with corticosteroids which were discontinued within a year when her disease

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was well-controlled, without signs of arthritis or nephritis.

During the week prior to admission, the patient suffered three episodes of transient right sided weakness, each lasting for less than 20 min. On the day of admission, she had a severe bitemporal headache and right sided weakness, which did not resolve.

On examination, the patient was without respiratory distress. The temperature was 37°C, the heart rate was 80/min, and the blood pressure was 170/100 mmHg. Examination of her head, ear, eyes and lung was unremarkable. The heart sounds were normal without murmurs, the abdomen was soft and non-tender and the musculoskeletal examination revealed no actively inflamed joints. The neurological examination demonstrated a mild right hemiparesis without sensory loss or hyperreflexia. The cranial nerves were normal.

The erythrocyte sedimentation rate (ESR) was 72 in the first hour, the hemoglobin concentration was 11.3 g/dl, the leukocyte count was $9000/ml^3$ and the platelet count was 166 000/ml³. Serum electrolyte, urea and creatinine levels were normal. Serological tests for antinuclear antibody were positive, the anti-DNA antibody titer was 83 units (Farr assay) (normal <10 units), the C_3 level was 88 mg/dl, (normal 80-120 mg/dl) and the C_4 level was 11 mg/dl (normal 20-40 mg/dl). The activated partial thromboplastin time (aPTT) was 27.1 s (control 30s), the serum titer of IgG anticardiolipin antibody (ACLA) was 4.3 units (normal < 23 units) and the titer of IgM ACLA was 11.2 units (normal <15units). Lupus anticoagulant was not detected with thromboplastin (dilutions of 1:50 and 1:500), Russell's viper venom, normal plasma (dilution 1:1) and Actin FSL. The VDRL test was negative.

Correspondence: Dr D Flusser, MD, Department of Medicine D, Soroka Medical Center, Beer-Sheva, Israel 84101



Figure 1 Axial 12 weighted image of the brain. An infarct is seen in the left parietal area. A 'delta' sign signifying a thrombus is seen in the sagittal sinus, posteriorly (arrow).

A computerized tomographic (CT) scan of the brain, an echocardiogram and an electroencephalogram (EEG) were normal at admission.

Neuro-psychiatric SLE was diagnosed and the patient was treated with intravenous methylprednisolone pulse therapy (500 mg daily for three days). One week after admission, she developed right hemiplegia and right Jacksonian fits were observed. An examination of the retinal fundus was normal, without signs of papilledema. A repeated CT scan of the brain showed a hemorrhagic infarct of the left hemisphere. An increased cerebral spinal fluid (CSF) pressure of 320 mm H₂O was found by lumbar puncture, with a protein level of 54 mg/dl. No cells were detected. Subsequently, the patient developed bilateral papilledema and right hemianopsia.

MRI demonstrated a left parietal infarct and the 'delta' sign, suggestive of cerebral venous thrombosis



Figure 2 The venous phase of cerebral angiography. A thrombus is seen in the sagittal sinus

(Figure 1). This was confirmed by the venous phase of a cerebral angiogram, which showed SSST (Figure 2).

The patient received anticoagulant therapy, with an incremental improvement in her motor strength and visual acuity. One month after admission, no neurological deficit was found. A repeat test of ACLA one year later was also within normal limits.

Discussion

Acute focal cerebrovascular deficits occur in less than 5% of patients with SLE. The most prevalent causes of these events are cerebral infarction due to thrombosis of cerebral blood vessels, in association with the antiphospholipid syndrome, accelerated atherosclerosis secondary to steroid therapy, or vasculitis of medium and small sized cerebral arteries. Rare causes of focal neurological damage include brain hemorrhage, malignancy, infection and cerebral venous thrombosis⁹.

Superior sagittal sinus thrombosis is a distinct clinical entity characterized by an acute onset of severe headache, focal neurological deficit, cranial nerves palsy, visual impairment and increased intracranial pressure $(ICP)^{10}$. Our patient presented initially with a severe headache and a focal neurological loss, without clinical features of increased ICP, and with high levels of anti-DNA antibodies and low titers of complement. The neurological findings were highly suggestive of an immune mediated mechanism (vasculitis). However, the lack of response to steroid therapy, the worsening of the neurological finding, and the development of increased ICP raised suspicion of a cerebral venous thrombosis.

Superior sagittal sinus thrombosis in association with SLE was first noted by Gibson and Myers². Eight case reports of SSST in patients with SLE have appeared in the literature²⁻⁶. In one case SSST was diagnosed in the postpartum period². In another case the thrombosis occurred prior to the diagnosis of SLE⁴, while in two other cases the two conditions were diagnosed at the same time^{5,6}. In four cases SSST was diagnosed in patients known to suffer from SLE. In these the interval between the diagnosis of SSST and SLE ranged from one to 36 years. A history of venous/arterial thromboses was elicited from one patient⁶. Serological tests for lupus anticoagulant were positive in two of six patients tested⁶. However, the levels of anticardiolipin antibodies were not measured and more sensitive assays of the lupus anticoagulant were not performed. In our patient we found no evidence for either anti-cardiolipin antibodies or lupus anticoagulant, despite rigorous testing with several assays, including the RVV, Actin FSL, thromboplastin and normal sera, suggesting that the development of SSST is not restricted to SLE patients with the antiphospholipid syndrome.

In two cases^{5,11} increased ICP was the only clinical feature of SSST. Previous cases have been described with benign intracranial hypertension, or pseudotumor cerebri (PTC), a clinical entity with increased ICP not related to obstruction of cerebrospinal fluid flow or to an intracranial mass¹². However, SSST and PTC differ in their clinical course, treatment and prognosis and it is important to distinguish between them.

Magnetic resonance imaging (MRI) is indicated in all cases of unexplained increased ICP. MRI is a sensitive procedure for the diagnosis of SSST. The MRI findings indicative of SSST include the absence of flow void on T1 weighted images and the appearance of the thrombosed vein as a hypodense cord in T2 weighted images. Secondary findings include cerebral edema, venous infarction and enhancement of the dural sinus wall around the clot ('delta' sign)¹³. Cerebral angiography is indicated in patients with a non-diagnostic MRI study. The venous phase of cerebral angiography is a highly sensitive tool for visualization of the thrombus in the sagittal sinus, and is considered the gold standard for diagnosing SSST.

The treatment of SSST includes anticoagulant therapy and steroids. The role of steroids in SLE patients without major organ involvement is unclear. There are published reports of patients treated only with steroids who had a favorable outcome⁵. However, this positive outcome may represent the natural course of the disease, or a true response to steroid. The use of anticoagulants in patients with SSST associated with SLE or other diseases has led to positive clinical responses. Our patient received a high initial dose of steroids, without clinical improvement. Shortly after heparin was added to the therapeutic regimen, she experienced a significant clinical recovery, suggesting that anticoagulation is an effective therapeutic strategy for SSST. The prognosis of treated SLE patients with SSST is usually favorable. In all six cases of SSST reported in patients with SLE, the patients had a complete resolution of their neurological deficit.

In summary, SSST is a rare clinical manifestation of SLE. It may occur in patients with and without antiphospholipid antibodies. The development of increased ICP is a clinical marker of the disease. Early diagnosis is essential because treatment leads to a positive clinical outcome.

References

- 1. Gladman DD, Urowitz MB. Systemic lupus erythematosus clinical features. In: Klippel JH, Dieppe PA (eds). *Rheumatology*. St. Louis: Mosby, 1994: 6.2.1–6.2.24.
- Gibson T, Myers AR. Nervous system involvement in systemic lupus erythematosus. Ann Rheum Dis 1975; 35: 398-406.
- Molle D, Guillevin L, Herreman G, Godeau P. Thrombophlebite du sinus longitudinal superior et lupus erythemateux dissemine. Sem Hop 1982; 58: 1215-1219.
- 4. Shiozawa Z et al. Superior sagittal sinus thrombosis and systemic lupus erythematosus. Ann Neurol 1986; 20: 272.
- Parnass SM et al. Dural sinus thrombosis: a mechanism for pseudotumor cerebri in systemic lupus erythematosus. J Rheumatol 1987; 14: 152-155.
- 6. Vidailhet M et al. Cerebral venous thrombosis in systemic lupus erythematosus. Stroke 1990; 21: 1226-1231.
- Love PE, Santoro SA. Antiphospholipid antibodies and the lupus anticoagulant in SLE and in non-SLE disorders. Ann Intern Med 1990; 112: 682-698.
- Glueck HI et al. Thrombosis in systemic lupus erythematosus: relation to the presence of circulating anticoagulant. Arch Intern Med 1985; 145: 1389-1395.
- 9. Bluestein HG. The central nervous system in systemic lupus erythematosus. In: Lahita RG (ed). Systemic Lupus Erythematosus. Churchill Livingstone, 1992: 639-655.
- 10. Enevoldson TP, Russel RW. Cerebral venous thrombosis: new causes for an old syndrome? Q J Med 1990; 77: 1255-1275.
- 11. Li EK, Chan MSY. Is pseudotumor cerebri in SLE a thrombotic event? J Rheumatol 1990; 17: 983-984.
- Li EK, Ho PCP. Pseudotumor cerebri in systemic lupus erythematosus. J Rheumatol 1989; 16: 113-116.
- Jacobs BC, Brant-Zawadzki M. Ischemia. In Stark DD, Bradley WG (eds). Magnetic Resonance Imaging. Mosby, 1992: 636-669.

336