# Cerebral Venous Sinus Thrombosis Presenting Feature of Systemic Lupus Erythematosus

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*Background:* CVST (cerebral venous sinus thrombosis) may sometimes be associated with autoimmune disorders that require specific treatment. The clinical and magnetic resonance imaging (MRI) findings of systemic lupus erythematosus (SLE) patients with CVST are presented and contrasted with CVST without SLE. *Methods:* Consecutive patients with CVST admitted in neurology service during 2012-2016 were included. The diagnosis of CVST was confirmed by MR venography or digital subtraction angiography. SLE was diagnosed according to American College of Rheumatology criteria. The clinical and MRI findings of CVST with SLE and those without SLE were compared. *Results:* Forty-three consecutive patients with CVST were included during the study period, 3 of whom had SLE .Their age ranged between 20 and 35 years and all were females. The clinical markers of SLE were present in all and included oral ulceration in 3 patients, serositis in 2 patients, and arthritis and psychosis in 1 patient. The SLE patients did not have antiphospholipid antibodies or lupus anticoagulant. The manifestation of CVST in SLE was similar to the other patients with CVST. The CVST in SLE required long-term anticoagulation and immunosuppression with cyclophosphamide pulse in 1 patient and oral prednisolone in 2 patients. The outcome was good, partial, and poor in 1 patient each. *Conclusions:* CVST may be the presenting feature of SLE, but these patients often have clinical clues to SLE. These patients need prolonged anticoagulation and immunosuppression. **Key Words:** Cerebral venous sinus thrombosis—systemic lupus erythematosus—antiphospholipid antibodies—lupus anticoagulant.


## Introduction

Cerebral venous sinus thrombosis (CVST), although a rare cause of stoke (.1%), is common in pregnancy, puerperium, and in females on oral contraceptive. CVST may sometimes be associated with autoimmune disease such as systemic lupus erythematosus (SLE). In such patients, CVST is usually associated with antiphospholipid antibodies (APLA) and lupus anticoagulant (LA). Such patients have been reported in literature but seldom does CVST occur in SLE without APLA and LA. We managed 3 patients with SLE in whom CVST was a presenting feature. It is important to consider the possibility of SLE because of specific therapeutic requirements.

## Patients and Methods

Consecutive patients with CVST admitted in neurology service during 2012-2016 were included. This study was approved by the Institute Ethics committee. The diagnosis of CVST was confirmed by magnetic resonance

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venography (MRV) or digital subtraction angiography. SLE was diagnosed according to American College of Rheumatology criteria.[1]

## Clinical Evaluation and Investigations

The demographic information (age, gender), clinical presentation, and their outcome were noted. The duration of disease and presenting features like seizure, headache, encephalopathy, and focal signs were also noted. The risk factors such as use of oral contraceptive drugs, pregnancy or puerperium, past thrombotic events such as deep venous thrombosis, abortion, dehydration, infection, or evidence of vasculitis or SLE (joint pain, rash, or oral ulcer) were noted. General and systemic examinations were done. Fundus was examined for papilledema, optic atrophy, or retinopathy. Focal weakness was noted and categorized as hemiparesis or quadriparesis. Consciousness was evaluated by Glasgow Coma Scale and mentation by Mini-Mental State Examination.

The laboratory investigations included blood count, thyroid function test, enzyme-linked immunosorbent assay D dimer, autoimmune and vasculitis markers (antinuclear antibody, anti-double stranded DNA, extractable nuclear antigen antibody, antineutrophil cytoplasmic antibody, C-reactive protein, and C3, C4), fasting homocysteine, Vitamin B12, and serum folate level. Abdomen ultrasonography was done and 24-hour urinary protein was measured. Prothrombotic conditions like functional assay of protein C, protein S, and antithrombin III; methylene tetrahydrofolate reductase mutation; Factor V Leiden mutation; LA; APLA; fibrinogen level; and paroxysmal nocturnal hemoglobinuria profile were screened.

Cranial magnetic resonance imaging (MRI) with MRV was done and parenchymal lesions (infarct, hemorrhagic transformation, or hematoma) with evidence of venous sinus thrombosis were noted. On MRV, the location, extent of venous sinus thrombosis, number and type of sinuses involved, and development of anastomosis and its type were noted.

Outcome was evaluated by modified Rankin Scale (mRS) on discharge and at 3 months. The outcome was categorized into poor (mRS > 2) and good (mRS ≤ 2).

## Results

There were 43 patients with CVST, of whom 3 have SLE. The median age of CVST patients without SLE was 29.65 (range 3-55) years and 18 (45%) were females. The median duration of hospitalization was 16.50 (range 0-74) days. The presenting symptoms were headache in 38 (95.0%), vomiting in 33 (82.5%), and altered sensorium in 23 (57.5%). Seizures were present in 30 (75%) patients and 10 (25%) of whom had status epilepticus. Seizure was presenting in 26 (65%) patients, early in 3 (7.5%) patients, and late in 1 (2.5%) patients. Refractory status was present in 3 patients and it was recurrent in 3 patients. Focal weakness on presentation was noted in 21 (52.5%), monoparesis in 3, hemiparesis in 12, and quadriparesis in 6. Papilledema was present in 11 (27.5%) patients and median GCS score was 14 (5-15) and was ≤8 in 6 (15%) patients. Eight patients required mechanical ventilation.

**Table 1.** *Comparison of CVST patients with and without SLE*

| | CVST without SLE (n = 40) | CVST with SLE (n = 3) |
|---|---|---|
| Age (mean + SD) | 29.7 ± 11.8 | 27.00 ± 7.6 |
| Female | 18 (45%) | 3 (100%) |
| Duration of hospital stay (mean + SD) | 17.2 ± 11.8 | 22.00 ± 4.6 |
| Clinical features | | |
| Papilledema | 11 (27.5%) | 1 (33%) |
| Seizure | 30 (75%) | 3 (100%) |
| GCS at admission (mean) | 12.8 ± 3.1 | 12.7 ± 1.2 |
| Encephalopathy | 23 (57.5%) | 3 (100%) |
| Focal deficit | 21 (52.5%) | 2 (66.75) |
| MRI | | |
| Parenchymal involvement | 31 (77.5%) | 3 (100%) |
| Infarction | 5 (12.5%) | 1 (33.3%) |
| Hemorrhagic lesions | 26 (65%) | 2 (66.7%) |
| Bilateral lesions | 12 (30%) | 1 (33.3%) |
| Supratentorial lesions | 31 (77.5%) | 3 (100%) |
| Number of sinuses involved (mean + SD) | 2.3 ± 1.2 | 2.4 ± 1.2 |
| Both superficial and deep sinus involvement | 5 (12.5%) | 1 (33.3%) |
| Outcome at 3 months | | |
| Good | 32 (80%) | 2 (66.7%) |
| Poor | 8 (20%) | 1 (33.3%) |

Abbreviations: CVST, cerebral venous sinus thrombosis; GCS, Glasgow Coma Scale; MRI, magnetic resonance imaging; SLE, systemic lupus erythematosus.

Three of 43 patients with CVST had SLE, all of whom were female with median age of 26 years. The onset of disease was subacute (1-6 weeks) in the first patient, chronic (>6 weeks) in the second patient, and acute (<1 week) in the third patient. The median duration of hospitalization was 21 days (range18-27) and none required mechanical ventilation. Headache, vomiting, altered sensorium, and seizure were present in all of them and focal weakness was present in 2 (66.7%) patients. All had presenting seizure and status epilepticus was present in 2 (66.7%) patients. Papilledema with later optic atrophy was present in 1 patient. Median GCS score was 12 (12-14) and was ≤8 in none of them (Table 1).

### *MRI Findings*

In the patients with CVST without SLE, MRI revealed parenchymal lesion in 31 (77.5%), infarction in 5