



**RESEARCH ARTICLE**

**CEREBRAL VENOUS SINUS THROMBOSIS MASQUERADING AS GUILLIAN BARRE SYNDROME- A CASE REPORT**

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**ABSTRACT**

Cerebral venous sinus thrombosis is a rare condition accounting for 3-4 cases per million. This represents 0.5%-3% cases of all strokes predominantly in younger adults. Usually such cases present with headache, blurring of vision, diplopia, loss of control over movements in the body and at times seizures. Due to its varied presentation cerebral venous sinus thrombosis can be very difficult to diagnose. We present an interesting case of cerebral venous sinus thrombosis that masqueraded itself as Guillian Barre Syndrome.

**Key words:**

Cerebral venous thrombosis (CVT), Cerebral vein thrombosis, Cerebral venous and sinus thrombosis, Cerebral venous sinus thrombosis (CVST), Cerebral vein and dural sinus thrombosis, Sinus and cerebral vein thrombosis.

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**INTRODUCTION**

Ankle fractures have shown an exponential increase in incidence due to the high speed motorcycles on the roads. Thrombosis in cerebral brain or venous sinuses is much less common cause of cerebral infarction than that caused by arterial disease. The diagnosis is, unfortunately, sometimes initially missed on clinical assessment and The diagnosis is very much elusive even by a plain MRI or CT SCAN. However imaging study with contrast can demonstrate the obstruction of the venous sinuses<sup>1</sup>. The clinical presentation may be with features suggestive of transient ischemic attack (TIA) or at times an intracranial space-occupying lesion (ICSOL). High index of suspicion is required in appropriate clinical settings to diagnose this entity at the earliest. Prompt treatment with anticoagulants, in such cases is known to cause complete recovery.

**Case report**

A 26 year old male, driver by occupation, admitted with complaints of numbness of both upper extremities while driving. The numbness was followed 4 days later by a left sided

neck pain, headache-more in early morning worsened with position but not associated with any vomiting, was followed by numbness and weakness of both lower extremities also. The patient consulted a private practitioner who diagnosed him as Guillian Barre Syndrome on the basis of absent reflexes and progressive ascending weakness of the limbs. On admission to our hospital the patient complained of swallowing difficulty, pain on left side of neck as well as numbness and weakness of upper and lower extremities. MRI brain plain was done on admission which did not reveal anything. Admission blood pressure was 130/90 mmhg, heart rate was 63/min, oxygen saturation was 100% on room air. Patient was breathing comfortably and did not demonstrate any signs of respiratory distress or insufficiency.

On the nervous system examination he was found to be conscious, oriented to time place and person, and pupils were bilaterally equal reacting to light and his other cranial nerves examination was remarkably normal including the IX<sup>th</sup> and X<sup>th</sup> nerves. However he was unable to hold his neck upright or roll over to his sides. Power in upper and lower extremities was 2/5. Single breath count however was good. His bilateral plantars were equivocal and reflexes were present only on

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reinforcement .All modalities of sensations were preserved, There was no bowel or bladder involvement.

His laboratory investigation (complete blood count, serum electrolytes, creatinine phosphokinase, viral markers) and imaging of heart and chest did not reveal any abnormality. A neurological opinion was sought. Unfortunately we were unable to conduct a peripheral nerve conduction study as the machine had some technical problem. Cervical spine M.R.I was found to be normal. Lumbar puncture showed normal cerebrospinal fluid profile. However since it is well known that 10% of patients with G.B.S can present with normal proteins in the CSF the most likely clinical diagnosis was considered as Guillian Barre Syndrome. Treatment option of plasmapheresis versus intravenous immunoglobulin was discussed and patient was administered intravenous immunoglobulin (IVIG) for 5 days.

There was absence in the progress of his symptoms in the next 3 days. He did not develop any form of respiratory insufficiency. The patient was thus shifted to the ward with advice to continue extensive physiotherapy .However he did keep complaining of the head and neck pain that persisted right through the treatment and even when being shifted out of the ICU.

Next day the patient complaint of severe headache and was shifted back to ICU. He was noted to be irritable and mildly disoriented.

Preliminary CT brain done then showed no abnormality. The incessant headache prompted us to perform a MR venography which revealed loss of signal void in right sigmoid, transverse and superior saggital sinuses, suggestive of sinus thrombosis with no evidence of venous infarct .A funduscopy revealed papilloedema.

Patient was then started on anticoagulation and PT/INR was monitored and targeted to maintain between 2-3. During his stay with us patient showed signs of improvement , his headache had reduced, patient was well mobilised ,power in upper and lower extremities were 4/5. After observation and adjusting the dose of PT/INR and patient being hemodynamically stable he was discharged with the adjusted dose of warfarin.

## DISCUSSION

CSV T is a rare condition with an incidence of about 3-4 cases per million<sup>1</sup> with a predilection for women<sup>4</sup>.This condition is most commonly seen in pregnant women or patients on oral contraceptive pills. The incidence of cerebral venous thrombosis is 12 cases per 100 000 deliveries in pregnant women.

At least 1 risk factor can be identified in >85% of patients with cerebral venous thrombosis

In the International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVT) cohort, a thrombophilia was noted in

34%, and an inherited thrombophilia was detected in 22%. .Amongst these are deficiencies of antithrombin, protein C, and protein S . Factor V Leiden mutation. Prothrombin gene mutation 20210. Antiphospholipid antibodies, and Hyperhomocysteinemia.

Infections such as otitis, mastoiditis, sinusitis, Meningitis and Systemic infectious disorders are other causes of cerebral venous sinus thrombosis. Chronic inflammatory diseases ,cancer, trauma, hematologic disorders like polycythemia, essential thrombocytosis, can also causes this entity.

According to The International Study on Cerebral Vein and Dural Sinus Thrombosis, the most commonly affected site is the transverse sinus, followed by superior sagittal sinus and straight sinus <sup>5</sup>. Other less common sites are the cortical vein, jugular vein and internal cerebral vein. In most patients, thrombosis occurs in more than one sinus.

In the authors opinion our patient had cerebral venous sinus thrombosis patient had cerebral venous sinus thrombosis with raised intracranial pressure and papilloedema t presentation. at presentation .However the ascending areflexic paralysis with normal sensations diverted the diagnosis towards Guillain Barre Syndrome. His M.R.I brain and cervical spine were normal which could not explain any other cause of his symptoms. The incessant headache was the clue that the diagnosis was not Gulllian Barre Syndrome.

Increased venous pressure due to the thrombosis may cause reduction in capillary perfusion and thus cerebral perfusion causing ischemic injury and thus cytotoxic edema. Meanwhile such increase pressure may also lead to breakdown of the blood brain barrier and thus lead to vasogenic edema. In cases of extreme elevation of venular pressure due to the cerebral sinus thrombosis there could also be rupture of these vessels causing parenchymal hemorrhage. In effect all these pathophysiological changes may cause rise in the intracranial pressure and various manifestations including ascending weakness etc as noted above in the clinical case.

Up to 90% of the patients complaints of sub acute onset of headache 64% of time<sup>5</sup>.Headache maybe generalized or localized and it may worsen due to change of position or valsalva's maneuver.

Up to 44% of patients presents with focal neurological deficit and the most common finding is hemi paresis which may be seen in 40% of the patients <sup>5,6</sup>. In the elderly population many of these symptoms may not be seen leading to delay in the diagnosis.

30%-40% patients present with focal or generalized seizures<sup>5</sup>, which are generally features of sagittal sinuses and cortical vein thrombosis.

Raised ICP and papilloedema are rare but reported in GBS literature and this has been variously attributed to reduced CSF absorption and cerebral oedema <sup>8,9</sup>.Such papilloedema is usually

asymptomatic or may have mild visual defects and visual loss is very rare in such cases

Extensive radiculopathy due to raised intracranial pressure has been known to present with papilledema, marked visual impairment and flaccid areflexic quadriplegia with no parenchymal lesions on MRI of brain, brainstem and cervical spinal cord. However this could be diagnosed with nerve conduction studies. The high csf pressure may be causing distension of the subarachnoid spaces and thus causing compression of the nerve roots leading to the above manifestations. This phenomenon and clinical presentation has been documented in a few reports of idiopathic intracranial hypertension and cerebral venous sinus thrombosis as well<sup>10</sup>. Thrombotic complications following IVIG have been reported in literature<sup>11,12</sup>. These complication can occur anywhere from the start of therapy to two weeks later<sup>13</sup>. The mechanism of this complication is unknown. However it has been postulated that the hyperviscosity, platelet activation and arterial vasospasm caused by IVIG may be contributing to the same.<sup>14,15,16</sup> However the time course of the headache of the patient in our case did not suggest that the cerebral venous sinus thrombosis was caused by IVIG.

Diagnosis is always confirmed with CT, MRI and MRV. CT has poor sensitivity, only in 1/3rd of the patients it shows direct signs of cerebral venous thrombosis<sup>17</sup>. CT venography can be compared with MR venography for the diagnosis.<sup>18,19</sup> D-DIMER elevation is an extremely sensitive test when there is a suspicion of the entity however is insufficient as a standalone test for the diagnosis of cerebral venous thrombosis<sup>6,17,18</sup>

The treatment of cerebral sinus thrombosis is therapeutic anticoagulation to prevent thrombus propagation and prevent complications of deep vein thrombosis and pulmonary embolism.<sup>17</sup> The only relative contraindication would be in very large venous hemorrhagic infarct with significant mass effect. Oral anticoagulants should be overlapped with heparin and the latter discontinued once therapeutic INR of 2-3 is achieved.

In a meta-analysis of 1180 patients with cerebral venous thrombosis, the mean 30-day mortality rate was 5.6%. Recurrence of cerebral venous thrombosis is rare (2.8%). 30 Follow-up imaging to assess for recanalization 3-6 mo after diagnosis is recommended.

## CONCLUSION

Cerebral venous sinus thrombosis is an extremely rare but life threatening entity that can come in varied presentations. All cases of ascending areflexic paralysis are not G.B.S.

An extremely heightened sense of suspicion is required in order to diagnose this often missed diagnosis of cerebral venous sinus thrombosis.

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