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Rare presentation of cerebral venous sinus Thrombosis

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RARE PRESENTATION OF CEREBRAL VENOUS SINUS THROMBOSIS:

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ABSTRACT:

We report case of a 16 year old girl with no known co-morbidities, presented with headache, quadriparesis and sudden vision loss. She had pallor and BMI was 33.09kg/m². She had left 6th nerve palsy, bilateral papilledema and no perception of light. Motor examination showed power of 4/5 in all four limbs with bilateral extensor plantars. Neuroimaging was suggestive of superior sagittal sinus thrombosis. CSF opening pressure was 72 cm of water. 20 ml CSF was drained leading to improvement in headache and visual acuity. CVST presenting with quadriparesis and visual loss is a rare entity. Common presentations of CVST include headache, seizures, focal neurological deficit and altered conscious level. In our case risk factors for CVST like anemia and high body mass index (BMI) were present, but presenting clinical features were unusual. CVST is treatable so early diagnosis and knowledge of whole spectrum of clinical presentation is of extreme importance.

KEY WORDS: Cerebral venous sinus thrombosis, quadriparesis, visual loss

INTRODUCTION:

CVST is a distinct cerebrovascular disorder that most commonly affect young adults and children.^{1,2} It has a strong female preponderance with female to male ratio of 3:1.^{3,4,5} It is less common than arterial stroke but with advancement of neuroimaging modalities it is being more frequently diagnosed. Due to its variable clinical spectrum its diagnosis is often challenging.³ Annual incidence of CVST is 0.22-1.57/100,000 of population.^{3,6} The median age of patients presenting with CVST is 37 years.⁴ Raised intracranial pressure is a common complication of CVST, occurring more frequently if superior sagittal sinus (SSS) is involved.³ Signs and symptoms of CVST are grouped into three major syndromes:

1. Isolated intracranial hypertension syndrome (IIHS), consisting of headache with or without vomiting, papilledema and visual disturbance.⁷
2. Focal syndrome (focal deficits, seizures, or both)³.
3. Encephalopathy (multifocal signs, mental status changes, stupor or coma)⁶.

With SSS thrombosis motor deficit, bilateral deficit and

seizures are frequent, however; IIHS is infrequent.³

We report a case of CVST with unusual presentation of quadriparesis and visual loss in whom SSS thrombosis presented unusually with features of IIHS.

CASE REPORT:

We report the case of a 16 year old female, who presented with headache for 2 months, episodic, involving the whole head, throbbing in character with no other associated feature and relieved with analgesics. This was followed by 10 days history of acute onset non progressive, painless, symmetrical quadriparesis and 4 days history of bilateral visual loss. There was no past medical history of drug use, upper respiratory tract infection, vomiting, diarrhea, fever, seizures or sphincter involvement while gynecological history was normal. Patient had a Body Mass Index (BMI) of 33.09 kg/m². She has pallor of conjunctiva, with stable vitals and Glasgow coma scale (GCS) of 15/15. She had no perception of light bilaterally and pupils were dilated and non-reactive to light. Fundus examination showed

bilateral papilledema with hemorrhages. She had left 6th nerve palsy. Sensory system examination was normal. Motor system examination showed normal tone, power of 4/5 in all four limb, reflexes were grade 2 and plantars were bilaterally extensor. Functional mobility scale (FMS) was 3. Important differential diagnosis were multiple sclerosis and NMOSD. Blood complete picture showed Hb: 5.3, MCV 69. Serum peripheral film showed microcytic hypochromic red cells with Hemoglobin electrophoresis was normal.

Lumbar puncture was done and showed CSF opening pressure of 72 cm of water. 20 ml of CSF was drained. CSF routine examination was normal. CSF for oligoclonal bands was negative. Anti-NMO antibodies were negative. Thyroid function tests were normal. CT scan brain was normal.

MRI brain and orbit showed left transverse and superior sagittal sinus flow void signals while MR Venography (MRV) revealed superior sagittal sinus thrombosis (fig 1 and 2). CSF opening pressure checked after 3 days showed opening pressure of 35 cm of water. Injection Methylprednisolone 1 gm once daily was given for 3 days, acetazolamide 500 mg twice daily and injection Enoxaprin 1mg/kg twice a day subcutaneously, later shifted to tablet Rivaroxaban. 2 pints of Red cell concentrates were transfused. Ventriculoperitoneal shunting was planned. Visual acuity improved to perception of moving fingers and FMS was 1.

DISCUSSION:

CVST is a multicausal disorder that affects the venous part of neurovascular system. The incidence, clinical presentations, imaging and outcome are variable. Various risk factors are associated with different age groups.^{8,9,10,11,12} Risk factors associated with CVST are numerated in table 1.⁸

INFECTIONS	<ul style="list-style-type: none"> ●) sinusitis ●) Central nervous system infections
TRAUMA	<ul style="list-style-type: none"> ●) Head injury
OTHER MEDICAL CONDITIONS	<ul style="list-style-type: none"> ●) Dehydration ●) Pregnancy related
	<ul style="list-style-type: none"> ●) Hematologic disorders: Polycythemia, sickle cell disease, Thrombotic thrombocytopenic purpura, Polycythemia, Paroxysmal nocturnal hemoglobinuria ●) Hereditary thrombophilia ●) Collagen vascular disorders
MEDICATION	<ul style="list-style-type: none"> ●) Oral contraceptive medications

Table 1. Risk Factors for CVST

However 20% of cases do not show any known risk factors.^{11,12,13,14} The risk of CVST is increased 20 folds in females with inherited disorder of hemostatic system who are using contraception.¹³ Pregnancy and puerperium causes a fourfold increase in the risk of venous thrombosis. Females below the age of 40 years have a twofold higher risk of having venous thrombosis as compared to arterial infarcts.¹⁴

With increased awareness and advancement of diagnostic tools CVST is more frequently diagnosed.¹⁴ Clinical presentations of CVST are in table 2. Headache is the most common presenting symptom in CVST.¹⁴

Anemia is not very uncommon in CVST. 76 % of female patients with CVST has anemia and 66% of them had microcytic hypochromic anemia.¹⁵ In our case patient had microcytic hypochromic anemia which also contributed to development of CVST.

Symptoms	<ul style="list-style-type: none"> ●) Headache ●) Blurred vision ●) Altered sensorium ●) Nausea, vomiting ●) Seizures
Signs	<ul style="list-style-type: none"> ●) Papilledema, ●) Focal neurologic deficit ●) Cranial nerve palsies

Table 2. Clinical Presentation of CVST

A rare syndrome is described comprising of blindness, ophthalmoplegia and extensive radiculopathy in 2 patients of CVST and cause in them was raised ICP.^{16,17} Bilateral motor weakness is common with SSS thrombosis. Both clinical features, i.e. visual impairment and bilateral motor weakness are present in our patient which is a rare presentation of CVST. Cerebral venous sinus thrombosis is treatable disease, with prompt diagnosis and treatment, risk of complications and disability can be reduced.

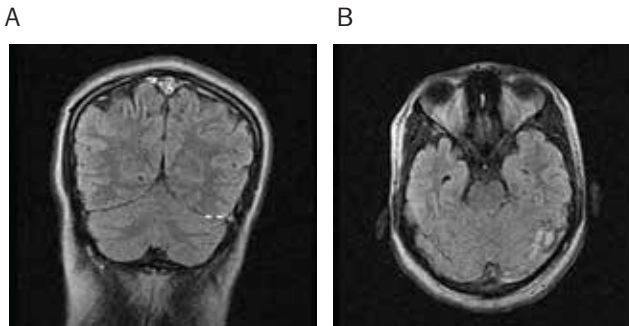


Figure 1. MRI brain plain FLAIR images. A and B show coronal (A) and axial images (B) with hyper intense signals within superior sagittal, left transverse and sigmoid sinuses

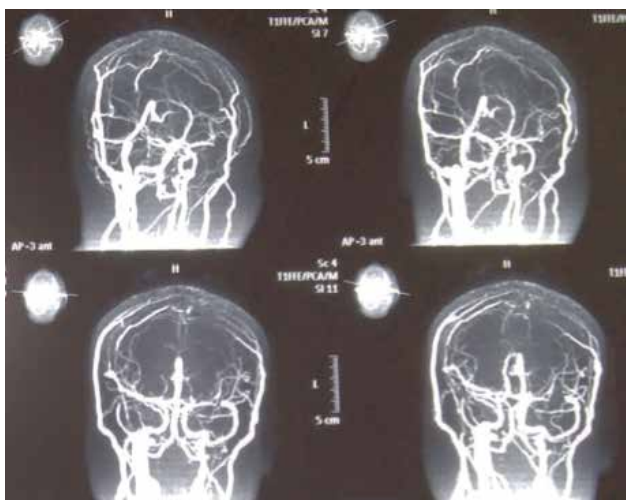


Figure 2. MRV brain. MRV shows signal void in superior sagittal sinus, left transverse and sigmoid sinuses, suggestive of superior sagittal sinus, left transverse and sigmoid sinuses thrombosis.

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Author's contribution:

Haris Majid Rajput; concept, data collection, data analysis, manuscript writing, manuscript review

Anam Anis; data collection, data analysis, manuscript writing, manuscript review

Iqra Athar; data collection, data analysis, manuscript writing, manuscript review

Neelma Naz Khattak; data collection, data analysis, manuscript writing, manuscript review

Hanin Tanich; data collection, data analysis, manuscript writing, manuscript review

Mazhar Badshah; data analysis, manuscript writing, manuscript review