Case Report

Superior Ophthalmic Vein Thrombosis with Cerebral Venous Sinus Thrombosis: A Rare Entity in a Child

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Abstract

Cerebral venous sinus thrombosis (CVST) is a rare stroke-like syndrome. Sometimes it may be rarely associated with ophthalmic vein occlusion. We present a 10-year-old girl who had severe headache, diplopia, severe congestion of eye, proptosis and orbital pain. She had no signs of meninigeal irritation, no focal deficit except left sided lateral rectus palsy and altered visual acuity. Investigations regarding vasculitis and thrombophillia were normal except thrombocytosis. MRI of brain showed 'dense triangle sign' and thickened superior ophthalmic vein. Magnetic resonance venography (MRV) showed occlusion of superior sagital sinus and transverse sinus. She was treated with paracetamol, acetazolamide, rivaroxaban and antibiotics followed by high dose pulse methyl prednisolone. She made a partial recovery within one week and at 3 months follow-up she was completely normal.

Key words*: Superior ophthalmic vein thrombosis; Cerebral venous sinus thrombosis; Rivaroxaban; Acetazolamide*

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Introduction

Cerebral venous sinus thrombosis (CVST) is a rare stroke-like syndrome occurring in only two to five cases per million per year.[1] It has a significant morbidity and mortality. This entity is associated with hypercoagulable states and can present with simple headaches to localizing neurologic deficits, frank seizures, and coma. Prompt recognition and treatment can prevent significant morbidity and life loss. The risk factors include infection and genetically acquired pro-thrombotic states.[2] Superior ophthalmic vein thrombosis (SOVT) is another clinical entity which is rare in pediatric population. The diagnosis is both clinical and radiological. It occurs due to congestion of the vein resulting into swelling of eye, proptosis, restriction of eye movement and visual impairment. It occurs due to infectious and noninfectious etiologies. In both cases confirmation of diagnosis is done with MRI and magnetic resonance venography (MRV) of the structures involved.[3] Here we present a case of a SOVT and CVST occurring in a 10-year-old girl.

Case report

A 10-year-old girl presented with headache for 2 months, redness of eye for 2 weeks and fever for 7 days. Prior to this illness she was in good health. Her headache was progressive, tightening in nature, usually more in morning, associated with vomiting which was non-projectile in nature. Later on, she developed redness of eye along with double vision associated with photophobia. She also developed orbital pain but her vision was intact. Her fever was low grade and irregular in nature. She had no history of seizure, loss of consciousness, trauma in head, palpitation, breathlessness, skin rash, photophobia, oral ulcer, contact with tuberculosis patient, joint pain, ear/eye/sinus infection and urinary problem. She was the first issue of non-consanguineous parents and there was no history of migraine or vasculitis in her family.

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The girl was conscious, ill-looking and co-operative. Eyes were congested and proptosis was present bilaterally (Fig 1). She had no pallor, cyanosis, jaundice, clubbing or edema. Anthropometry was within normal range. Vitals were within normal limit. There were no signs of meningeal irritation. Skin survey was normal and BCG mark was present. Ophthalmoscopic examination revealed hyperemic vessels, visual acuity was 6/18 in right eye and 6/12 in left eye unaided. Lateral rectus nerve palsy was present in left side. There was no other neurological deficit in the patient.

Her investigation reports showed Hb 12 gm%, leukocytosis (total count 13700, neutrophil 80%), thrombocytosis (total platelet count 6,93000), CRP 12 mg/dL, normal serum electrolytes and normal liver and renal function tests. Urine for routine examination was normal. PT was 14.9 (control 11.80), INR 1.25, APTT 32 seconds (control 28 seconds). Protein C and Protein S were normal. Antithrombin III, ANA and Anti-DS DNA were normal. Antiphospholipid antibody was IgG 4.1 U/mL (normal) and anticardiolipin antibody was IgG 5.23 U/L (normal). Homocystine level was 16.36 (high normal) and thyroid function tests were normal. Mantoux test was negative. CSF study revealed normal findings.

Magnetic resonance imaging (MRI) showed the classical 'dense triangle sign' in the posterior part of the superior sagittal sinus which corresponds the fresh thrombus (Fig 3). Thus it suggested thrombosis of superior sagittal sinus. In addition to that there was thickened superior ophthalmic vein with indistinct border due to perivascular edema of both sides. Magnetic resonance venography (MRV) of brain revealed features of dural venous sinus thrombosis involving superior venous sinus and transverse sinus without any evidence of hemorrhagic venous infarct (Fig 4). We diagnosed the case as bilateral ophthalmic vein thrombosis and cerebral venous sinus thrombosis.

As the child presented after two months of onset, heparin was not given. The patient was treated with rivaroxaban (an oral anticoagulant) 10 mg daily. For headache paracetamol was given. Along with this, acetazolamide was also given. As the patient had fever with increased WBC count, injectable antiobiotic was given for 7 days. Her headache decreased. But congestion of eye was still there along with lateral rectus palsy. Methylprednisolone was started and continued for five days. After one dose of steroid, there was dramatic improvement of orbital pain and congestion of eye (Fig 2). After three months the girl was completely normal without any residual focal deficit, eye abnormalities or headache.

Fig 1. Congestion of eye and lateral rectus palsy in a girl with CVST and SOVT

Fig 2. Improvement of congestion after pulse methylprednisolone

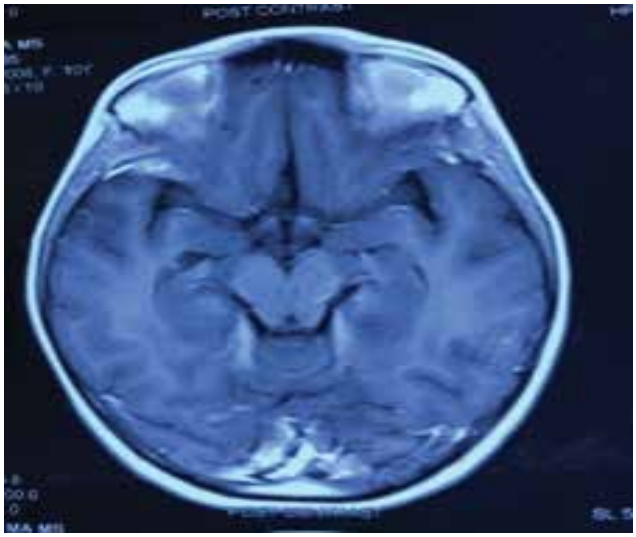

Fig 3. MRI of brain with contrast showing 'dense triangle sign'

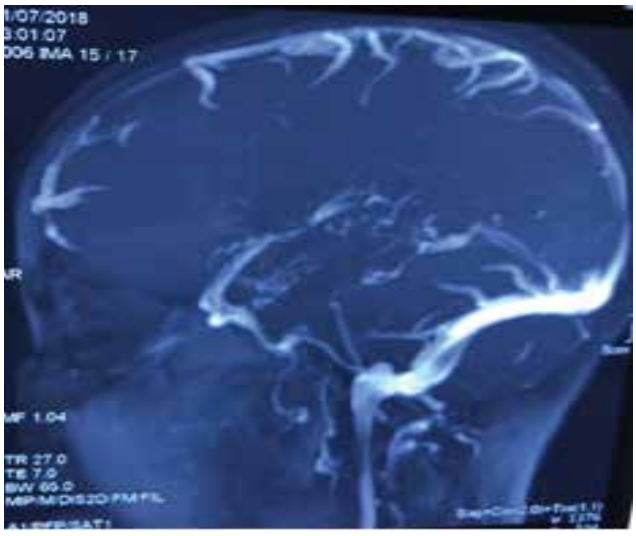

Fig 4. MRV of brain showing filling defect in superior sagittal sinus and transverse sinus

Fig 5. MRV of brain showing defect in superior sagittal sinus, transverse sinus and internal jugular veins

Discussion

CVST is an uncommon type of stroke, which may account up to 0.5 to 1% of all strokes.[4] Although CVST has been reported in adults, it has more commonly been observed in neonates and children in hospital-based studies.[2,5] The ratio of adult females to males is 3:1.[2,6] It can create diagnostic dilemma as the presentation is widely variable including isolated headache, mood changes, cranial nerve palsies, ocular complaints, aphasia, encephalopathy, TIA/stroke symptoms, seizure or frank coma. Headache is the most consistent complaint and is classically described as slow onset, dull, localized and worse with recumbent position.[2]

Traditionally CVST is of two types — septic and aseptic. Risk factors related to CVST are diverse. At least one risk factor could be identified in more than 85% of patients with CVST.[7] The common risk factors are pregnancy and puerperium, drugs like oral contraceptives, steroid, L-asparginase, hormone replacements, malignancy, systemic inflammatory disease like systemic lupus erythematosus, infectious diseases, hematological disorders like polycythemia, dehydration, coagulation disorders, congenital heart diseases, head trauma etc.[8] In this case patient had leukocytosis and thrombocytosis. These may be the risk factors in this case. All other blood tests and urine

investigation were within normal limits. Based on Bousser, four clinical patterns for CVST have been identified — a) Focal syndrome: presence of focal signs associated with headache, seizures or changes in mental state, b) Isolated intracranial hypertension (ICH): headache, nausea, vomiting and papilledema, c) Diffuse subacute encephalopathy: changes in mental state and d) Cavernous sinus syndrome: painful ophthalmoplegia, chemosis and proptosis.[9] Our case matches with the type 4, but here there was evidence of left cavernous sinus thrombosis (left 6th nerve palsy).

SOVT is usually found in cases of orbital congestion such as orbital cellulitis, idiopathic orbital inflammation, vascular malformation, infection, trauma, tumor and coagulopathy.[10,11] Some systemic diseases have been reported in association with SOVT like systemic lupus erythematosus, Graves disease, ulcerative colitis etc.[12-16] Other causes are hematologic like antiphospholipid syndrome, sickle cell disease, hormonal causes like tamoxifen, oral contraceptive pills etc, Tolosa-Hunt syndrome, idiopathic orbital inflammatory disease.[17-25] In a case series by Nicolien et al[3] the majority of the patients were aseptic (seven out of nine) although all the patients in this case series were adults. The septic causes were sinusitis, dental, orbital and facial infection. The underlying cause in our case was CVST. The presenting symptoms are swelling of eyes, chemosis, proptosis, impaired eyelid motility, reduced visual acuity and increased intraocular pressure.[3] In our case we found proptosis, chemosis, impaired visual acuity and restricted motility of eyeball although here intraocular pressure was normal. The clinical presentations may be explained by congestion of the orbit due to impaired venous drainage. This mechanism is also present in CVST, thus both SOVT and CVST may occur together like in our case. In our case the SOVT was bilateral but in literature review most of the cases of SOVT found are unilateral.[3]

The main modality to confirm the diagnosis both CVST and SOVT is neuroimaging. Various imaging techniques are used; CT angiography and MRI/MRV being most sensitive and specific. In CVST the 'dense triangle sign' is a hyperdense triangular or round shape in the posterior superior sagittal sinus that has been described in non-contrast head CT scans. 'Empty delta sign' seen with contrast-enhanced CT scan is a triangular ring of enhancement with a central region lacking contrast in the posterior superior sagittal sinus.[26] In case of our patient, MRI showed the classical 'dense triangle sign' in the posterior part of the superior sagittal sinus which corresponds to fresh thrombus. Her MRV showed non-visualized superior sagittal sinus and transverse sinuses which confirmed the diagnosis.

The diagnosis of SOVT is confirmed preferably by contrast-enhanced CT or MR imaging. On contrast-enhanced CT scan SOVT is characterized by a thickened ophthalmic vein with irregular border. MRI is the modality of choice for confirming SOVT as it shows all stages of thrombus formation. Here a filling defect can be seen within the SOV itself, or a thin lining can be present close to the vessel wall, either due to vessel wall enhancement or a small lumen in between the clot and the vessel wall.[27,28] In case of a chronic SOV thrombosis, the central thrombus can also show enhancement opposite to the acute stage. MRI shows enhancement of intraorbital fat and swelling and enhancement of the eye musculature due to venous congestion. Similar to contrast enhanced CT, MRI can show intra-luminal filling defects in the SOV. Secondary signs include proptosis and thickening of the extraocular muscles.[29] MRI also excludes carotid-cavernous sinus fistula, cavernous sinus thrombosis and sino-orbital infection.[17] Orbital color Doppler imaging also allows noninvasive confirmation of SOVT.[28] In our case we have done MRI of brain with contrast with MRV which revealed thickened bilateral SOV with irregular border.

In CVST anticoagulant in the form of heparin in acute stage and warfarin in maintenance phase are suggested if there is no hemorrhage or contraindication. In case of transient risk factor such as infection, trauma or pregnancy, a treatment of three months is recommended while in prothrombotic states a prolonged duration of treatment is suggested. No definite data are there about the duration. However, mostly 6–12 months treatment is given.[31] As our patient presented late, we prescribed a substitute of warfarin, rivaroxaban which needed minimal monitoring. Addressing the etiology, as our patient had leukocytosis without definite focus of infection, we gave the patient a short course of injectable antibiotic. As there was severe headache initially, we gave her paracetamol and acetazolamide anticipating increased

intracranial pressure. For SOVT antibiotics, steroids and anticoagulants are the mainstay of treatment. Surgery may be needed for an underlying sinusitis, orbital abscess or dental infection.[3] Our patient had severe congestion of both eyes, thus we treated the patient with pulse methylprednisolone which caused dramatic improvement of the congestion and orbital pain.

Early detection and treatment and identification of risk factor are very important for favorable outcome in both CVST and SOVT. Most patients experience complete resolution over several weeks with treatment. Our patient improved in hospital with treatment and at three month follow-up she was completely normal without any residual clinical feature.

Conclusion

Our case had some exceptional criteria found rarely — pediatric onset, SOVT coexisting with CVST and dramatic response to steroid and anticoagulant. The underlying cause was thrombocytosis which is nonspecific. Thus in a pediatric patient with new onset severe headache along with proptosis and congestion of eye a suspicion of SOVT and CVST can be made as early diagnosis has positive implication in management and prognosis.

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