Abstract: Cortical venous thrombosis is a rare type of cerebrovascular disease that mostly affects children and young adults. Superior sagittal sinus thrombosis is the most common type. CVT in children is multifactorial. A prothrombotic risk factor or direct cause is identified in about 85% of cases with sinus thrombosis. The widespread use of neuroimaging now allows for early diagnosis and treatment. CVT is more common than previously thought with favourable outcome and low mortality rate. MRI with T1, T2, fluid-attenuated inversion recovery and T2 sequences with MRV are the best diagnostic methods. Heparin is the first line treatment, but in few cases more aggressive treatments such as local intravenous thrombolysis, mechanical thrombectomy and decompressive hemicraniectomy may be required.

Keyword: CVT, superior sagittal sinus thrombosis, protein C and protein S deficiency.

Introduction: CVT in childhood is a serious disease that is being increasingly diagnosed mainly because of more sensitive diagnostic procedures and increasing clinical awareness of the disease (1). Incidence is 7 cases per million among children. We report a 5 yr old child with superior sagittal sinus thrombosis with protein C and protein S deficiency as a prothrombotic factor.

CASE REPORT: 5 yr old female child Jasmine seevi second born to nonconsanguineous marriage parents from tanjore admitted in the hospital with headache and vomiting for 4 days. One episode of GTCS with ALOC for 10 mts and low grade fever one day duration, no history of trauma, dog bite or recent vaccination or no history of eardischarge. No history of similar illness in the past, previous hospitalisation or family history of seizure. Child treated in near by hospital and referred to our hospital as a case of acute CNS infection. On examination child drowsy, febrile, responds to painful stimuli, no pallor, no neurocutaneous markers, no focal neurological deficit. System examination normal except fundus examination shows bilateral papilloedema. Child treated with antiepileptics, antibiotics and steroids. All basic investigations are within normal limits. CSF analysis done was normal. CT brain taken 24 hrs after admission and reported as sagittal sinus thrombosis with bilateral maxillary sinusitis and ethmoidal, sphenoidal sinusitis. Neurologist advised MRI with MRV and to continue antiepileptics and antibiotics. Hematologist advised to do protein C, protein S and antithrombin levels and start LMWH. MRI and MRV taken shows superior sagittal sinus thrombosis and right lateral sinus thrombosis. LMWH started and continued for 7 days and child put on oral warfarin. Child responded well. PT and INR monitored. INR maintained 1.5 to 2 times normal. Protein C and S levels reduced, child having 38.2% protein C (normal 70 to 140%) and protein S 10% (normal 55 to 123%). Antithrombin levels are normal. Follow up MRI with MRV taken after 14 days shows recanalisation within the thrombus which is the sign of resolution. Child discharged after 3 wks and advised to continued oral warfarin for 6 months and antiepileptics.

DISCUSSION: Cortical venous thrombosis (CVT) in childhood is a serious disease that is being increasingly diagnosed, mainly because of more sensitive diagnostic procedures and increasing clinical awareness of the disease(1). The clinical presentation shows a wide spectrum of symptoms, eg, seizures, papilledema, headache, lack of consciousness or lethargy, and focal neurological deficits(1,2). According to ISCVT frequency of symptoms follows: headache 89%, papilloedema 49%, seizures 37%, neurological deficit 34%, altered mental status 30%, multiple cranial nerve palsies 12% and cerebellar symptoms in 3% (16). The origin and pathophysiology of CVT in the pediatric population is still poorly understood, mainly because of its low incidence, which is estimated at 0.67 per 100 000 children(1). The disease is serious, and predisposing and influencing factors should be unraveled to identify patients at risk and to establish treatment regimens in children. Local or systemic infections, vascular trauma, cancer, acute lymphoblastic leukemia, drug toxicity, lupus erythematosus, nephrotic syndrome, dehydration, asphyxia, maternal problems during pregnancy, Behcet’s disease and metabolic disorders, have been described as predisposing factors. Recently published data have suggested that multiple additional factors including prothrombotic risk factors contribute to the symptomatic onset of CVT. Prothrombotic factors are factor V leiden mutation, protein C, protein S deficiency, anti-thrombin deficiency, prothrombin mutation and homocystinemia. High factor VIII levels appear to be associated with CVT in adults (Cakmak et al., 2003), but factor VIII is not commonly performed in children (Kurecki et al., 2003).
According to recent study on CVT (ISCVT), mostly more than one sinuses involved. Frequency of involvement of sinuses follows: superior sagittal sinus 72%, lateral sinus 70%, straight sinus 14.5%, cavernous sinus 2.7%, cerebral vein 35% and cerebellar vein 3% (16). Frequent predisposing condition in children follows: prothrombotic disorders 41%, dehydration 25%, head and neck infection 18%, systemic infection 8%, haematological disorders 12%, malignancy 8% and cardiac causes 5% (16). In our case superior sagittal sinus and right lateral sinuses are involved and child having protein C and S deficiency as a risk factor.

As clinical diagnosis is difficult in CVT, neuroimaging usually required. MRI with T1 T2 fluid-attenuated inversion recovery and T2 sequences with MRV are the best diagnostic methods. CT and MRI may show focal oedema (63%), infarct (47%) and hemorrhage (39%) (16).

Currently, the standard care for patients with a first episode of CVST is anticoagulation therapy, which is usually prescribed for 3 to 6 months in the absence of an identifiable cause. However, controlled data about the benefit and optimal duration of oral anticoagulation in patients with CVST are missing. In the recent ISCVT study, 58.8% of the patients with a recurrence of thrombotic event were not undergoing anticoagulation therapy (16). The significance of these observations was underscored by the fact that recurrence of cerebral venous thrombosis after stopping anticoagulation therapy can occasionally run a fatal course (16).

The need for more chronic treatment is further supported by recent observations on the effects of prolonged anticoagulant therapy in patients with idiopathic venous thromboembolism (either pulmonary or deep venous thrombosis). The practical implication was that clinical practice should presumably consist of this prolonged, low-intensity anticoagulant regime, at least for patients with a prior event of pulmonary or deep venous thrombosis. So to reduce recurrence of thrombotic events after CVST, it may be prudent to treat these patients with long-term anticoagulant therapy. Future research should determine the optimal duration and intensity of oral anticoagulation therapy that is necessary to optimally reduce the risk for recurrence of thrombotic events.

Few cases require local intravenous thromblysis, mechanical thrombectomy or hemicraniectomy. Along with antithrombotic therapy adequate hydration should be maintained. Intracranial tension should be monitored. Antioedema measures should be started if child having increased ICT. If child having threatened vision lumbar puncture or lumboperitoneal shunt or VP shunt should be done to relieve ICT.

Poor prognostic factors are coma (worst), altered mental status, hemorrhage on admission, male child and underlying malignancy. Untreated cases mortality occurs >50% patients and in treated cases 67% become normal and remainder have mild neurological deficit. Child can have long term sequela in the form of hemiplegia, monoplegia, quadriplegia or optic atrophy.

REFERENCES:

picture 1: CT Brain showing superior sagittal sinus thrombosis

picture 2: CT Brain showing right transverse sinus thrombosis

picture 3: CT Brain showing B/L maxillary sinusitis
picture 4: CT Brain showing ethmoidal and sphenoidal sinusitis

picture 5: MRI Brain showing superior sagittal sinus thrombosis

picture 6: MRI Brain showing right transverse sinus thrombosis

picture 7: MRV Brain showing superior sagittal sinus thrombosis

picture 8: MRV Brain showing recanalisation in thrombus after anticoagulant therapy

picture 9: MRV Brain showing recanalisation in thrombus after anticoagulant therapy

picture 10: the child after recovery