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CEREBRAL VENOUS SINUS THROMBOSIS IN CHILDREN - A SERIES OF SEVEN CASES

Neurology

Dr. Pawan Saraswat* MD Fellow Pediatric Neurology, Institute of Medical Sciences, Banaras Hindu University, Varanasi, India. *Corresponding Author

Dr. Sandeep Kumar Baranwal MD Fellow Pediatric Neurology, Institute of Medical Sciences, Banaras Hindu University, Varanasi, India.

Dr. Aditi Agarwal Junior Resident, Institute of Medical Sciences, Banaras Hindu University, Varanasi, India.

Dr. Rajniti Prasad MD, Professor, Incharge Division of Pediatric Neurology, Institute of Medical Sciences, Banaras Hindu University, Varanasi, India.

Dr Ankur Singh Associate Professor, Institute of Medical Sciences, Banaras Hindu University, Varanasi, India.

ABSTRACT

Cerebral venous sinus thrombosis in children is associated with a high incidence of morbidity and mortality. Clinical features are non specific and variable. We report seven children admitted in the pediatric neurology ward with a median age of 3 years. The presented symptoms were headache, weakness and abnormal body movement. On examination they had fever, papilloedema, and seventh cranial nerve palsy. The most common underlying conditions were sepsis followed by nephrotic syndrome and ear infection. All cases received low molecular weight heparin and warfarin and improved in follow up.

KEYWORDS

Cerebral venous sinus thrombosis, Low molecular weight heparin, Magnetic resonance venography, Nephrotic syndrome

INTRODUCTION

Cerebral venous sinus thrombosis (CVST) is a rare condition, which has partial or total blood flow obstruction in the superficial or deep veins with or without ischemic/ hemorrhagic lesions of brain parenchyma [1]. Under the age of 18 years, incidence is 0.67% per year [2]. Nearly 40% cases have neurologic sequelae and reported mortality is 10% [3]. Clinical presentation is nonspecific; it varies from being asymptomatic in some to others having neurological symptoms such as headache, vomiting, weakness and seizures. Most common cranial nerves involved are 6th and 7th cranial nerves. Due to the non-specific nature of clinical symptoms, early diagnosis of pediatric CVST is quite difficult, delaying treatment and prognosis[1].

Diagnosis of CVST can be confirmed radiologically only, through magnetic resonance imaging (MRI), especially magnetic resonance venography (MRV), or CT venography (CTV) [5, 6]. Absence of flow in venous sinuses can be seen in MRV and CTV. The most common causes of CVST are head and neck infections like mastoiditis or systemic infections such as nephrotic syndrome [7, 8].

Other causes include tumors and head injury with skull fractures [9]. Here, we present a case series of seven cases presented to our institution in a pediatric neurology division with different presentations that ended up being diagnosed as CVST.

Case series

A total of seven patients admitted in a pediatric neurology division with the symptoms and signs as mentioned in table 1. and brain imaging findings in table 2.

Table 1: Showing characteristics, symptoms and signs

Case no	AGE	SEX	RISK FACTOR	SYMPTOMS	SIGNS
1	3 Year	female	Sepsis and Anemia	Weakness of left half of body, difficulty in speech, and nasal twang of voice	Left sided hemiparesis
2	1.5 year	female	Polycythemia	Fever with generalized tonic-clonic seizure	Left sided hemiparesis
3	1.5 year	male	Nephrotic syndrome	Irritable, poor appetite	Grade 2 papilledema
4	8 years	male	Nephrotic syndrome	Headache, generalized tonic-clonic seizure, weakness of right half of body	Right sided hemiparesis
5	20 days	male	Neonatal sepsis	Delayed cry after birth, respiratory distress, subtle seizures	Macrocephaly, Low set ear, Depressed nasal bridge, retrognathia, poor GCS
6	12 years	male	Sepsis	Headache	Grade 3 papilledema
7	3 year	male	CSOM	Fever, headache, vomiting	Left upper motor neuron facial palsy

Table 2 showing brain imaging findings

Case	Modalities	Finding
1	MRV	Right straight sinus thrombosis with acute hemorrhagic infarct in right caudate nucleus and thalamus
2	CTV	Hypodensity in anterior two third and hypodensity in posterior one third of sagittal sinus
3	MRV	Superior sagittal and straight sinus thrombosis
4	MRV	Cerebral venous sinus thrombosis involving superior and inferior sagittal sinus, internal cerebral vein and straight sinus
5	MRI	Global cortical atrophy, cortical thickening with pachygyria and polymicrogyria,right transverse sinus thrombosis
6	MRV	Absent flow in left transverse sinus
7	MRV	Right transverse sinus thrombosis along with right internal jugular vein showing subacute thrombosis

Age ranged from 20 days to 12 years (median 3 years, mean 4.1 years). Male to female ratio was 5:2. Headache and abnormal body movements were common features. Headache most commonly occurred in combination with other features such as vomiting and visual disturbance. Two children who had visual disturbances were

found to have papilledema. Frequency of clinical features at the time of diagnosis is headache in four children, seizures in three children, unsteady gait in two children, visual disturbance in two children and vomiting in one child.

Comorbid factors identified at diagnosis were Septicemia in three ,nephrotic syndrome in two,mastoiditis in one and anemia in one child . Three children had investigations for markers of thrombophilia, although the range of markers tested varied amongst these cases. Only one child was found to be anemic.

On MRV/CTV three children had superficial venous sinus thrombosis, out of which two had transverse sinus thrombosis and one had superior and inferior sagittal sinus thrombosis, one child had deep venous thrombosis involving straight sinus(figure 1) Three children had both superficial and deep sinuses involved including sagittal sinuses with deep cerebral veins or straight sinuses, two children had non-hemorrhagic infarcts found on MRI. One child had infarcts of the right parietal lobe and one child had a right thalamic infarct. No child had intracranial hemorrhage (ICH).

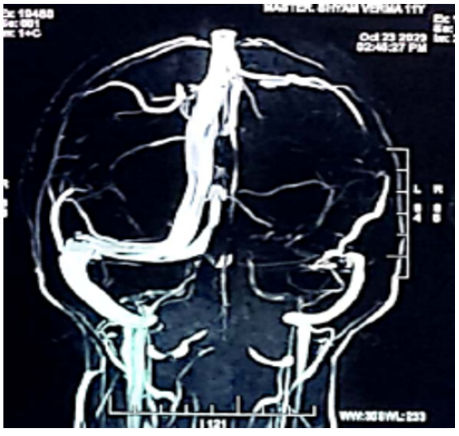

Figure 1 showing absent flow in left transverse sinus-suggestive of left transverse sinus thrombosis.

All 3 children with infection received antibiotics as per hospital protocol. All 7 children received low molecular weight (LMW) heparin (enoxaparin) during the acute phase, Which was overlapped with warfarin for 5 days and was later on shifted to oral warfarin for 3 months. None of the children required thrombolysis.

Outcome

Follow-up details were available for all patients. Outcome was good in all of them with no residual neurological deficit.

DISCUSSION

CVST is an important cause of stroke in the pediatric age group, causing morbidity and mortality. CVST affects more frequently male patients, which may be due to the protective role of estrogen, but few studies are available to confirm this[10, 11]. In our study male to female ratio was 5:2. Clinical presentation of this condition is extremely variable. The most common symptoms are symptoms of raised intracranial pressure (ICP), such as headache, vomiting, and drowsiness. Other symptoms include diplopia, strabismus, exophthalmos, papilledema, seizures and focal neurological deficits. In children below 1 year of age, there is increased compliance of the cranial compartment because of open cranial sutures and fontanelles, hence they have more nonspecific symptoms, such as decreased level of consciousness and seizures.

Pediatric CVST occurs due to both intravascular and vascular factors, and infection is among the common risk factors for all ages.This condition could present with an acute, subacute, or chronic onset.CT shows characteristic features such as the high-density shadow. Head MRI and MRV are utilized to diagnose CVST with high sensitivity and specificity. During the subacute stage, T1WI and T2WI sequences may directly identify the thrombus. MRV is characterized by a lack of cerebral venous sinus blood flow signal or marginal blurring, irregular lower signals, and venous collateral formation [12]. Therefore, head MRI/MRV is the "gold standard" for the diagnosis of CVST.

The American Society of Hematology 2017 Guidelines for Management of Venous Thromboembolism: There are 3 main anticoagulant drugs used in neonates and children, unfractionated heparin (UFH), low molecular weight heparin (LMWH), and oral vitamin K antagonists (VKAs)[13].

UFH is a commonly used anticoagulant in pediatric patients. It has the advantage of a short half-life and rapid onset and offset of action, making it ideal for cardiopulmonary bypass, circuit prophylaxis, procedural prophylaxis (e.g., cardiac catheterization), and for maintaining vascular access patency [13].LMWH has become the anticoagulant of choice in pediatric patients[13].

Warfarin is the most commonly used and studied VKA worldwide. The current therapeutic international normalized ratio (INR) ranges for children with VTE are inferred from recommendations for adult patients, because no clinical trials have assessed the optimal INR range for children. The therapeutic target INR is 2.5 (range, 2.0–3.0). Warfarin usually starts at 0.1 to 0.2 mg/kg and can be given maximum up to 5 mg [14]. Monitoring oral anticoagulant therapy in children is difficult and requires close supervision, with frequent dose adjustments. First, therapy should be started during hospitalization, and INR should be monitored daily or every few days. However, even after stable long-term therapy is achieved, only 10 to 20% of children are safely monitored monthly. The treatment duration is usually 6 weeks to 3 months. Summarizing the current consensus is that older children diagnosed with CVST without bleeding should receive anticoagulant therapy. It is safe and effective to administer subcutaneous injections of LMWH (two times a day, 180 U/kg BID) for 2 weeks, then switch to oral warfarin for 3 months.

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