ORIGINAL RESEARCH PAPER

Neurology

KEY WORDS: cerebral venous thrombosis, children.

AETIOLOGICAL FACTORS OF CEREBRAL VENOUS SINUS THROMBOSIS IN CHILDREN

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ABSTRACT

Objectives: To determine the clinical profile, aetiology and prognosis of cerebral venous sinus thrombosis in children beyond neonatal age group.
Design: Descriptive study.
Setting: Tertiary care teaching hospital.
Method: Children with cerebral venous thrombosis beyond neonatal age group were studied regarding age, sex, symptoms and signs, site of thrombosis, aetiology and prognosis.
Results: There were 16 children with cerebral venous thrombosis. Males (62.5%) predominated the study. Headache (62.5%) was the commonest symptom followed by vomiting, seizures, drowsiness, fever and focal deficits. Multiple venous sinuses were involved in the majority. 44% had septic causes. Nearly two thirds had a recovery.

INTRODUCTION:

Cerebral venous sinus thrombosis (CVST) is a rare form of venous thrombo embolism (VTE).It is an uncommon form of stroke affecting predominantly younger people[1] . It represents ~0.5-1% of all strokes.[2] CVST is characterized by varied clinical picture, diverse risk factors and prognosis. Literature is limited from our part of the country. Considering the paucity of data in our country, this study was conducted to determine the clinical profile, aetiology and prognosis of children with cerebral venous sinus thrombosis.

SUBJECTS AND METHODS:

The study group comprised of all children from 2 months to 12 years age group with a diagnosis of cerebral venous sinus thrombosis proved by Magnetic Resonance Imaging from 2014 to 2016.

The data was analyzed for age and sex distribution, clinical symptoms and signs, history of ear sepsis, underlying medical illnesses, sinuses involved in neuroimaging, possible cause and outcome. Relevant investigations such as complete haemogram, sepsis screening, CSF analysis, prothrombotic work up were done.

RESULTS:

During the study period, there were 16 children with cerebral venous sinus thrombosis. 8 (50%)were in more than 5 years age group, 5 belonged to 1-5 years, and there were 3 infants. There were 10 (62.5%) males and 6 females.

Headache 10(62.5%), vomiting 8(50%) were the common symptoms followed by drowsiness 6(37.5%), fever 6(37.5%),seizures 5(31%), irritability 4(25%). Focal deficits were noticed in 25%patients, papillodema in 6(37.5%), cranial nerve palsy in 4(25%), meningeal signs in 2 (12.5%).

There were 2 children with chronic serous otitis media, one child with acute lymphatic leukemia, one with nephrotic syndrome and one with cyanotic congenital heart disease.

The commonest sinus involved was transverse sinus in 12 (75%),followed by sigmoid sinus in 10(62.5%),superior saggital sinus in 4(25%), internal jugular vein in 4(25%).Majority presented with involvement of multiple sinuses. Intraventricular haemorrhage with hydrocephalus was noticed in one child. Parenchymal involvement in the form of infarcts were seen in the frontal region in 2, parietal region in 4, occipital region in 2. One had a cerebellar abscess and subdural empyema was noticed in one child. The infarcts were bland in 2 and haemorrhagic in 6 children.

The etiological risk factors identified in this cohort were infection in 7(43.7%), which included meningitis in 3,systemic sepsis in 2, mastoiditis in 2 children. Acute lymphatic leukemia on L-asparaginase was the risk factor in one, nephrotic syndrome in one, Tetrology of Fallot with polycythemia in one child. Prothrombotic conditions such as Protein C deficiency and Protein S deficiency were the causes in 2 children each. The cause could not be found out in 2 patients. The various causes of cerebral venous sinus thrombosis is depicted in table I.

Children were managed with antibiotics alone in 3(18.7%), antibiotics and anticoagulants in 4(25%), anticoagulants alone in 43.7%, One child with cyanotic heart disease with polycythemia needed a partial exchange. One child with hydrocephalus underwent ventriculo peritoneal shunting. Supportive measures included anticonvulsants and antiodema measures. 10 (62.5%) had a complete recovery. Remaining had residual neurological deficits in the form of hemiparesis in 2, quadriparesis in one, cognitive deficit in one, speech delay in one, developmental delay in one.

DISCUSSION:

The common age of children with CVST in our cohort was above 5 years. 50% of them belonged to this age group. Males predominated the study (62.5%).This is in accordance with other studies.[3,4]

We noticed that in our patients headache 10(62.5%), vomiting 8(50%) were the common symptoms followed by drowsiness 6(37.5%), fever 6(37.5%),seizures 5(31%), irritability 4(25%). Focal deficits including hemiparesis, monoparesis, cerebellar signs were noticed in 25% patients, papillodema in 6(37.5%), cranial nerve palsy in 4(25%), meningeal signs in 2 (12.5%). Wasay et al[5] in their study which included neonates noticed seizures as the most common symptom (59%), followed by drowsiness in 50%, headache in 18%, cranial nerve palsy in 33%, papillodema in 18%, hemiparesis in 29%.

The causes of cerebral venous sinus thrombosis are diverse in children. Acute infections of head and neck are the most common cause followed by chronic underlying disease like nephrotic syndrome, cancer, inflammatory bowel disease etc.[6] We also found that infection was the commonest cause of CVST in our cohort constituting 44%, of which bacterial meningitis in 3(43%), mastoiditis in 2 (29%),and systemic sepsis in 2(29%). Vieira et al[4] found that infection as the probable cause in 51%, mastoiditis in 43%, meningitis in 13%. The systemic underlying illnesses included acute lymphatic leukemia on L-asparaginase, nephrotic

syndrome, Tetrology of Fallot with polycythemia in one child accounting for 6% each. Wasay et al noticed nephrotic syndrome as the risk factor in 1%, leukemia in 2%, systemic lupus in 2%, leukemia in2%, sickle cell disease in 1%. Prothrombotic conditions accounted for 25% of the risk factors in our study which included Protein C deficiency and Protein S deficiency in 2 children each comprising 25% of all causes. Similar findings were reported by Mallick et al in their cohort of 21 patients.[7] However Heller et al have identified a higher percentage in their study which included 149 children less than 18 years of age.56.4% of their study population had atleast one prothrombotic risk factor.[8] The cause could not be identified in 12.5%. This is in accordance with other studies.[4,5]

Teksan et al described that the most common location for CVST was transverse sinus followed by superior saggital sinus, sigmoid sinus in older children and straight sinus in neonates.[9] We also noticed that the commonest sinus involved was transverse sinus in 75% of the children followed by sigmoid sinus in 62.5%,superior saggital sinus in 25%,and internal jugular vein in 25%.Haemorrhagic infarcts were noticed in 37.5% which is in accordance with other studies [3,5]

In cases of otitis media related CVST, children receive parenteral antibiotic therapy. The role of surgery such as mastoidectomy, myringotomy is unclear. Treatment regimens vary between centres , but older infants and children recceive anticoagulation in the acute setting with either unfractionated heparin or with low molecular weight heparin followed by chronic anticoagulation with low molecular weight heparin or oral anticoagulants.[10] Our patients with mastoiditis with CVST received parenteral antibiotics. 75% of our children required anticoagulation of which 83% were managed with low molecular weight heparin.

Nearly two thirds had a complete recovery which is in accordance with other studies. [4,5,7,11]

CONCLUSION:

Cerebral venous sinus thrombosis is an important cause of childhood stroke. Head and neck infections are the common risk factors followed by prothrombotic conditions. Transverse sinus is commonly involved and majority had involvement of multiple sinuses. Two thirds had a complete recovery.

Table I: Aetiological factors of cerebral venous sinus thrombosis in children

Etiology	No.(%)
1. Infections(7)	43.8%
Meningitis	3
Mastoiditis	2
Sepsis	2
2. Systemic illness(3)	18.8%
Acute Leukemia	1
Nephrotic syndrome	1
Cyanotic congenital heart disease	1
3.Prothrombotic conditions(4)	25%
Protein C deficiency	2
Protein S deficiency	2
4. Unidentified	2 (12.5%)

REFERENCES:

1. Bousser MG, Ferro JM. Cerebral venous thrombosis: An update. Lancet Neurol. 2007;6:162–70.
2. Ruiz-Sandoval JL, Chiquete E, Bańuelos-Becerra LJ, Torres-Anguiano C, González-Padilla C, Arauz A, et al. Cerebral venous thrombosis in a Mexican multicenter registry of acute cerebrovascular disease: The RENAMEVASC study. J Stroke Cerebrovasc Dis. 2012;21:395–400.
3. Sébire G, Tabarki B, Saunders DE, Leroy I, Liesner R, Saint-Martin C, Husson B, Williams AN, Wade A, Kirkham FJ. Cerebral venous sinus thrombosis in children: risk factors, presentation, diagnosis and outcome. Brain. 2005 Mar; 128(Pt 3):477-89.
4. Vieira JP, Luis C, Monteiro JP, Temudo T, Campos MM, Quintas S, et al. Cerebral sinovenous thrombosis in children: Clinical presentation and extension, localization and recanalization of thrombosis. Eur J Paediatr Neurol. 2010;14:80–5.
5. Wasay M, Dai A, Ansari M, Shaikh Z, Roach ES. Cerebral venous sinus thrombosis in children: A multicenter cohort from the United States. J Child Neurol. 2008;23:26–31.
6. Hashmi M, Wasay M. Caring for cerebral venous sinus thrombosis in children J Emerg Trauma Shock. 2011 Jul-Sep; 4(3): 389–394.
7. Mallick AA, Sharples PM, Calvert SE, Jones RW, Leary M, Lux AL, et al. Cerebral venous sinus thrombosis: a case series including thrombolysis. Arch Dis Child (2009) 94:790–4.
8. Heller C, Heinecke A, Junker R, Knöfler R, Kosch A, Kurnik K, et al. Childhood stroke study group.Cerebral venous thrombosis in children: A Multifactorial origin. Circulation. 2003;108:1362–7.
9. Teksam M, Moharir M, Deveber G, Shroff M. Frequency and topographic distribution of brain lesions in pediatric cerebral venous thrombosis. AJNR Am J Neuroradiol. 2008;29:1961–5.
10. Dlamini N, Billinghurst L, Kirkham FJ. Cerebral venous sinus thrombosis in children. Neurosurg Clin N Am. 2010;21:511–27.
11. deVeber G, Andrew M, Adams C, Bjornson B, Booth F, Buckley DJ, et al. Canadian pediatric ischemic stroke study group. Cerebral sinovenous thrombosis in children. N Engl J Med. 2001;345:417–23.