Case Report

Neonatal Cerebral Sinovenous Thrombosis: A Case Report

Dr. Snehal Jagtap[1], Dr. Abhijit Shinde[2], Prof. Dr. Sunil Natha Mhaske[3], Dr. Suresh Waydande[4], Dr. Sushrut Kumar[2]

[1]Junior Resident, [2]Assistant Professor, [3]Professor & Head, Department of Paediatrics, DVVPFs Medical College & Hospital, Ahmednagar-414111, Maharashtra, India

[4]Dean & Professor Paediatrics, DVVPFs Medical College & Hospital, Ahmednagar-414111, Maharashtra, India

Abstract:

Neonatal cerebral sino-venous thrombosis (CSVT) is a rare disorder, associated with long-term neurological sequelae. Cerebral sino-venous thrombosis (CSVT) is a focal or diffuse disruption of cerebral blood flow secondary to occlusion of cerebral veins and/or sinuses. The most commonly presented clinical manifestations were feeding difficulties, lethargy, respiratory distress, loss of consciousness, and seizures. Here we report one such rare case of Neonatal Cerebral Sinovenous Thrombosis in a full-term male admitted to the NICU in view of respiratory distress due to meconium-stained liquor and cried immediately after birth. The baby had convulsions on day 5 of life, MRI Brain was done and reported as multifocal areas of diffusion on restriction and large areas of diffusion restriction involving various parts of brain. Subcutaneous low molecular heparin was started. There is limited literature describing outcomes: 60-80% of all infants have some neurological deficits; cognitive impairment is reported in 10-60%, and epilepsy in 30-40%.

Key words: Cerebral Sinovenous Thrombosis, Stroke, Infarction, Convulsion, Neuroimaging

Introduction:

The incidence of CSVT in neonates is 0.6-15 per 100,000 newborns per year, which is higher than the incidence in the childhood period (0.67 per 100,00 children per year).[1] Among the different classes of perinatal stroke, CSVT is less common than arterial ischemic stroke (AIS). The reported incidence of CSVT is probably underestimated due to various reasons: non-specific clinical presentations, lack of clinician awareness, and the relative difficulty of radiological diagnosis in a neonate. Cerebral sino-venous thrombosis (CSVT) is a serious disease, which leads to long-term neurological sequelae in majority of survivors. There is uncertainty with regards to age specific individual risk factors, the impact of hereditary thrombophilia or underlying diseases on development of thrombosis or outcome. Previously published cohort studies have described epidemiology, treatment practices and outcomes, but those studies are limited due to small sample size or diversity of the populations included. The incidence of thrombotic disorders in children is increasing with advances in the diagnostic modalities, supportive care, and management of many health conditions. There is a bimodal peak in the paediatric age group, with the highest incidence in neonates and adolescents.[2] The developing coagulation system, need for intensive care, including catheterization, and co-morbid conditions are responsible for relatively high risk of thrombosis in neonates compared to older children.[3] Prevention and appropriate treatment are key to minimizing morbidity and balancing the adverse effects of treatment.

Seizures and neonatal encephalopathy are frequent clinical signs, non-specific signs and symptoms such as poor feeding, dehydration, and sepsis can also precede the diagnosis.[4] The management of neonatal CSVT, including the use of anticoagulation therapy (ACT), remains controversial because of the lack of efficacy and safety data and increased risk of intracranial haemorrhage (ICH).[5] There are some observational studies highlighting the safety of ACT; however, they are small and limited to single centers.[6,7]

Corresponding Author: *Dr. Snehal Jagtap*

Email ID: *snehal.jag95@gmail.com*

Address: *Department of Paediatrics, DVVPFs Medical College & Hospital, Ahmednagar-414111, Maharashtra, India*

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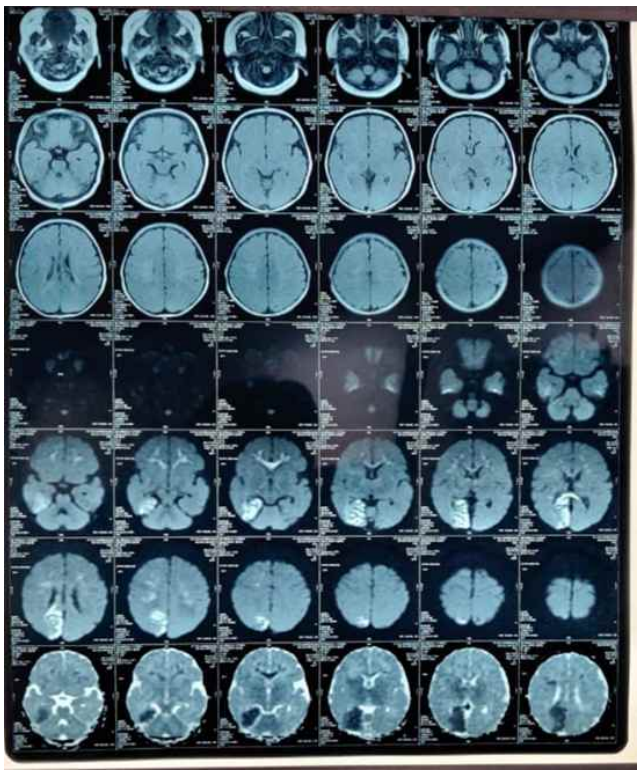
Case report:

A full-term male born to a primi mother via lower segment cesarean section in view of meconium-stained liquor with birth weight of 3kgms, cried immediately at birth and delivered at Civil Hospital Ahmednagar and then referred to our hospital in view of respiratory distress. On admission the Downe score was 2, so the baby was taken on O2 by nasal prongs and intravenous fluids were started. As the distress settled feeds were started via oro-gastric tube and increased gradually upto full feeds after which baby was breastfed and then shifted to motherside. On day 5 of life the mother complained of convulsions in the baby and so baby was admitted in the NICU and intravenous anticonvulsants were started. MRI Brain plain was done in view of the same, reported as multifocal areas of diffusion on restriction involving bilateral centrum semioval, corona radiata, thalami and all parts of corpus callosum suggestive of Acute Hypoxic Ischemic changes.(Pic no.1) Large areas of diffusion restriction involving right occipital and medial temporal regions suggestive of Acute Infarcts.(Pic no.1) Prominent vein of Galen with dilated Straight Sinus and bilateral Internal Cerebral Vein.(Pic no.2) Neuropaediatrician opinion was taken and as per advised- 2d echo, sickling test, serum homocysteine, PT/APTT done were all normal. MRI Angiography and MR venography done; reports were same as that of the previous MRI report. Subcutaneous low molecular heparin was started. No repeat convulsion was observed. Oral antiepileptics were started and the baby was discharged with continuation of subcutaneous heparin to be administered by the local paediatrician.

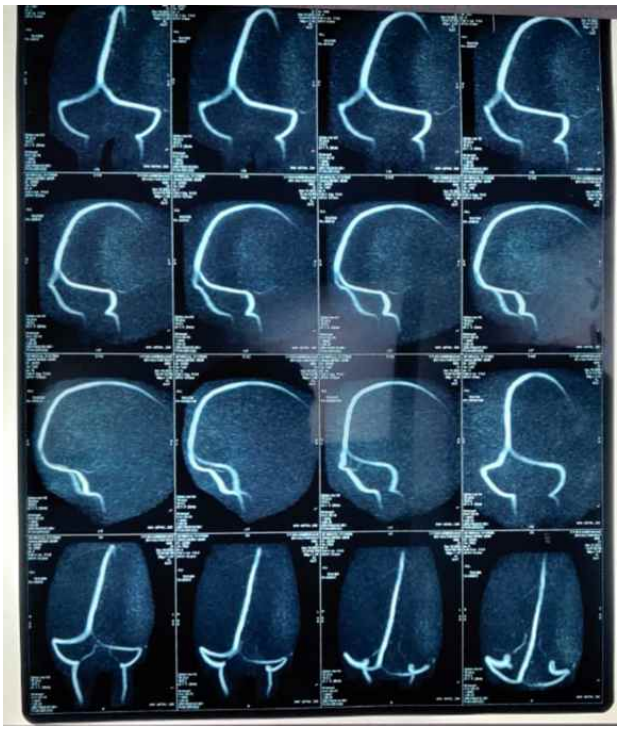
Discussion:

A venous clot leads to occlusion and impairment of drainage of cerebral veins. The result is, increased venous pressure, diminished capillary blood flow, vasogenic oedema, and infarction, usually haemorrhagic in nature. Thrombosis can be diagnosed by imaging with or without parenchymal involvement.[8, 9]

Pic no. 1: Multifocal areas of diffusion restriction involving bilateral Centrum semoovale, corona radiata, thalami and all parts of corpus callosum and Large areas of diffusion restriction involving the right Occipital and Medial Temporal region.



Pic no.2: Prominent Vein of Galen measuring maximum diameter of 6mm with dilated single Straight Sinus and bilateral Internal Cerebral Veins.



The risk factors for CSVT overlap with those for neonatal AIS. The following factors are reported most commonly .[10,11]

Maternal/neonatal factors	Prothrombotic factors	Other risk factors
Primiparity	a. Abnormal levels of prothrombotic factors are found in 10 -20% of infants with CSVT but the abnormalities are usually minor and occur in the context of other risk factors.	a. Male sex [12]
Multiple births		
Gestational Diabetes		
Pre-eclampsia		
Preterm birth		b. Mechanical compression during intrapartum events and occipital bone.[12]
Chorioamnionitis		
Complicated Delivery (vacuum, forceps), a. Meconium Aspiration b. Hypoxia c. Acidosis d. Asphyxia		
Acute Systemic Illness a. Meningitis b. Sepsis c. Dehydration		
Congenital Heart Disease		
Extracorporeal Membrane Oxygenation (ECMO)		

Infants who require ECMO are at risk of CSVT due to retrograde thrombosis following and occlusion of Right Jugular flow. The neonate suspected of having a CSVT should be evaluated with neuroimaging to make the diagnosis prior to detailed assessment of risk factors and formulation of treatment. Intraventricular haemorrhage in term infants or haemorrhagic infarction, especially involving parasagittal cerebrum, thalamus or basal ganglia should arouse suspicion of CSVT.

Seizures are the most common presentation (60-70%) of neonatal CSVT. Generalized and focal seizures are both common in CSVT, which is different than neonatal seizure with AIS, which are predominantly focal. Also, seizures with neonatal CSVT occur slightly later (later in first week or beyond) than with AIS (first day). Encephalopathy which can mimic hypoxic ischemic encephalopathy, is another common presentation. Respiratory distress, apnoea, tone abnormalities, and feeding difficulties have also been reported, especially in preterm infants and other features includes focal EEG abnormality, unexplained thrombocytopenia.[12]

Neuroimaging is mandatory to diagnose neonatal CSVT. Ultrasound: the presence of intraventricular haemorrhage, particularly when combined with thalamic haemorrhage, suggest the presence of CSVT. Colour flow Doppler ultrasound may be used to assist in the diagnostic process by demonstrating decreased or absent flow in venous sinuses and major cerebral veins. MRI: is the modality of choice to detect neonatal CSVT by demonstrating thrombus and parenchymal involvement. Diffusion Weighted imaging (DWI) combined with apparent diffusion coefficient (ADC) mapping detects infarcts within minutes of occurrence. Routine sequences, T1weighted and T2 weighted imaging, are useful for evaluation of blood products and oedema. Susceptibility weighted imaging (SWI) sequences are most sensitive to detect blood products and haemorrhagic transformation. Magnetic resonance venography (MRV) (time of flight) is the gold standard diagnostic approach to detect thrombus in the cerebral veins and sinuses.[13]

Low Molecular Weight Heparin (LMWH) or Unfractionated Heparin (UH) in neonates with severe thrombosis or clot propagation despite supportive treatment, but without intracerebral haemorrhage. Clot propagation occur in 25-30% of cases; anticoagulation therapy reduces the rate to about 3%.[14]

Conclusion:

The major factors influencing outcome of neonates following CSVT included comorbid medical conditions, abnormal neurological examination at presentation, location of venous thrombosis, and type of cerebral injury. These results can help guide further studies in neonatal CSVT aiming to decrease morbidity and mortality with the goal of improving long-term neurological outcomes. There is limited and discordant literature regarding outcomes and predictors of outcomes in neonates with CSVT. Furthermore, lack of standardized neurodevelopmental follow-up influence outcome data, but the most likely explanation is the spectrum of lesions sustained.

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