

Cerebral Venous Thrombosis in Neonates: Two Case Reports

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Abstract- Cerebral venous thrombosis is an uncommon disorder in children however it can be associated with serious clinical consequences. As it is a rare condition without any specific clinical presentation, it is usually undiagnosed. Clinical manifestation of cerebral thrombosis in children is relatively different from adult because of age-related differences in the vascular and neurologic systems. We present a case of a twenty days boy with a history of one-week lethargy and poor feeding and grand mal seizure. He had hypernatremia and severe dehydration. Non-contrast CT scan of the brain revealed superior sagittal and both transverse and sigmoid sinus thrombosis. The patient was treated in neonatal ICU with a favorable clinical response. We also present a fourteen days girl who presented with three days of fever, poor feeding, tachycardia, respiratory distress, and grand mal seizure. She had hypernatremia and severe dehydration. Non-contrast CT scan of the brain revealed superior sagittal sinus thrombosis and right temporal lobe infarction. She was also treated in neonatal ICU with noticeable improvement in symptoms. These cases demonstrate the importance of dehydration in neonatal and clinical suspicion of cerebral venous thrombosis (CVT). We present these two cases to enhance the awareness of clinical practitioners. CVT is an unusual and serious condition in neonates and due to its nonspecific clinical presentation often remains unrecognized. Early neuroimaging, including transfontanel doppler ultra-sonography, and non-contrast CT scan in all neonates with neonatal seizures will improve detection.

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Introduction

Cerebral venous thrombosis (CVT) was first described in the 19th century and was considered a rare condition with a fatal prognosis (1). The diagnosis can be difficult, because of its nonspecific clinical manifestations and radiological findings. Delay in diagnosis may lead to venous congestion, venous infarction, and even death. Early and accurate diagnosis of this condition is important (2).

Case Report

Case 1

A twenty days boy who was the first child of the family and born with cesarean section.

He presented with one-week lethargy and poor feeding. This was followed by one attack of generalized

tonic-clonic seizure. Relevant family history of disease did not exist. During the visit to the emergency room, he was lethargic. There was no fever, chills or diarrhea. Physical examination revealed tachycardia, decreased skin turgor, and severe dehydration with completely dry oral mucosa, and his reflexes were decreased.

Laboratory studies showed hypernatremia, hyperglycemia, increase of creatinine (Cr), and blood urea nitrogen (BUN), anemia and finally thrombocytopenia (Platelets<10000). The neonate under suspicion of septic shock was treated with appropriate hydration and antibiotics, platelets, FFP, insulin, phenytoin, and phenobarbital. Lumbar puncture was negative for signs of infection or hemorrhage, and arterial blood gas showed metabolic acidosis. Non-contrast CT scan of the brain showed high density in biventricular posterior horn representing acute IVH (Figure 1). There is also high density in superior sagittal

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and both transverse and sigmoid sinus suggestive of acute thrombosis without midline shift (Figure 2). Hypercoagulability studies in the emergency department were normal. Two days later neonatal brain sonography showed partial thrombosis of superior sagittal sinus and mild ventriculomegaly. The color Doppler sonography

showed increased echogenicity with no color in the vein. The patient was treated for two weeks in neonatal ICU. She experienced a good recovery without further seizure and was discharged. At one month follow-up, all the neurological and systemic examinations and child development were normal.

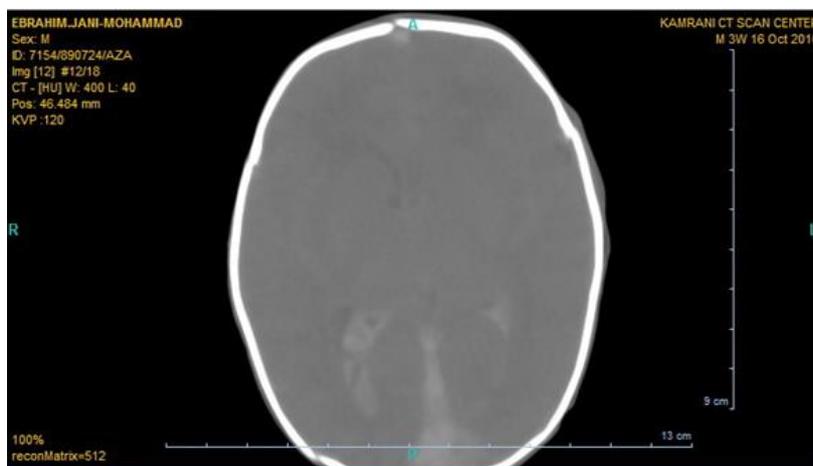


Figure 1. Unenhanced CT scan of the brain shows a high density in biventricular posterior horn representing acute IVH

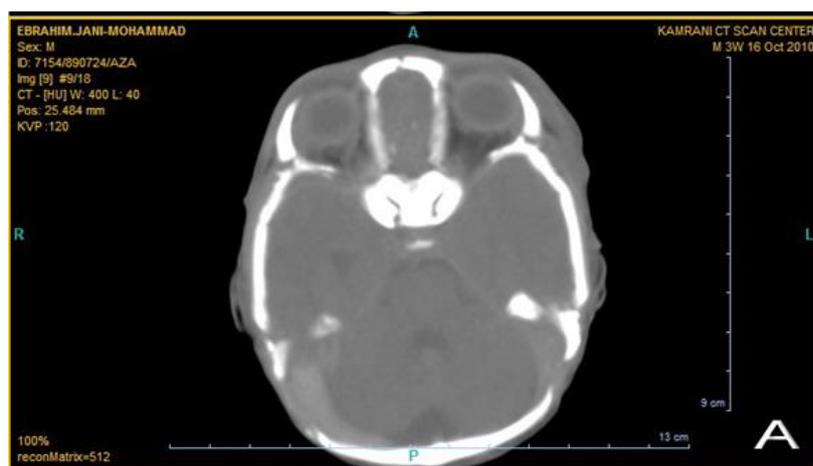


Figure 2. Unenhanced CT scan of the brain shows a high density in and both transverse and sigmoid sinus (a) superior sagittal (b)

Case 2

A fourteen days girl who was the first child of the family and born with natural delivery.

She was admitted with the history of three days of fever, poor feeding, decreased urination, and defecation. The family history of disease was not remarkable. During the visit in the emergency room, she was pale. Physical examination revealed tachycardia, fever, severe dehydration and respiratory distress and her reflexes was decreased. This symptom was followed by generalized tonic-clonic seizure. The neonate with diagnosis of septic shock was treated with antibiotics. The the patient

was intubated because of repeated apnea. Laboratory studies showed hypernatremia, hyperglycemia, thrombocytopenia (Platelets<16000), and rise in Cr and BUN. Lumbar puncture was unremarkable, and arterial blood gas showed metabolic and respiratory acidosis. Non-contrast CT scan of the brain uncovered high density in superior sagittal sinus suggestive of acute thrombosis (Figure 3) and wedge shape hypodensity in right temporal lobe put forward hypoxic ischemic infarction (Figure 4). Sonography demonstrated mild hydrocephaly and superior sagittal sinus thrombosis. Hypercoagulability study carried out in the emergency

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department, which was normal. She was treated with amikacin, vancomycin, meropenem, dexamethasone, platelets, fresh frozen plasma (FFP), insulin, phenytoin, and phenobarbital. Due to a pneumothorax happened on the second day of hospitalization a chest tube inserted for the patient and taken out four days later? The patient

was admitted for 18 days in neonatal ICU. Fortunately, she had a good recovery and was discharged. At 1 and 2 months follow-up, there were no clinically detectable neurological or functional deficits, and all the neurological and systemic examinations were normal.

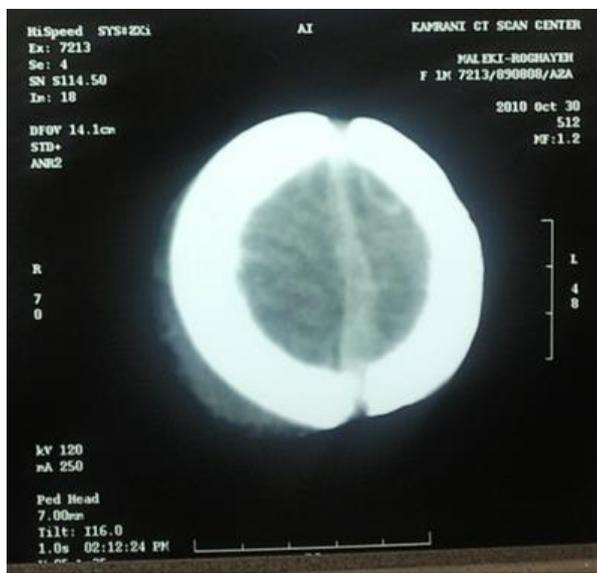


Figure 3. Unenhanced CT scan of the brain shows a high density in superior sagittal

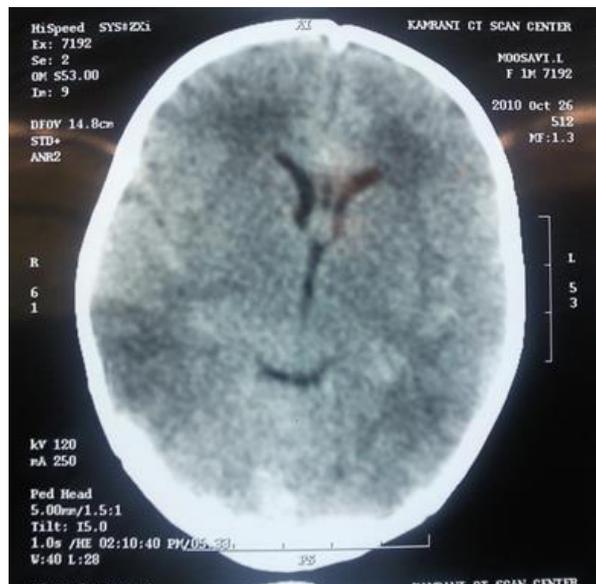


Figure 4. CT scan of the brain shows a hypodensity in right temporal lobe is suspicious of hypoxic ischemic infarct

Discussion

The majority of neonates with significant CVT (81%) present on the day of birth or within the first week of life (3). The incidence of CVT varies between 0.4 and 0.7 per 100,000 children per year, and neonates

are the most commonly affected age group, and it is reported to be as high as 2.6 per 100,000 per year (4,5). Conditions associated with neonatal cerebral sinovenous thrombosis are different from those reported in adults and include maternal conditions, chorioamnionitis, hypertension, perinatal conditions, meconium aspiration,

Apgar score <7 at 5 min, intubated at birth, neonatal infection, polycythemia, severe dehydration, congenital heart disease and disseminated intravascular coagulation (5). The perinatal complications included hypoxia at birth, premature rupture of membranes, maternal infection, placental abruption, and gestational diabetes are also reported (4).

Neurologic manifestations of CVT such as seizures and focal or diffuse neurologic signs vary with age. Seizures are more common and in neonates than in non-neonates (4). In adults, CVT presents with a wide spectrum of signs and symptoms and the clinical manifestation varies with the location and extent of the sinuses involved. Several patterns of clinical presentation have been defined, and the most common signs include diffuse headache, vomiting, papilledema, focal motor or sensory deficits, seizures, and increased intracranial pressure (6,7). Although symptoms and signs are often subtle and nonspecific, moderate to severe neurological sequelae are also reported (8). The primary neurologic manifestation in the neonates are seizures and diffuse neurologic signs; other symptoms include lethargy, irritability, poor feeding, apnea or changes in muscle tone. Seizures of various subtypes accompanied by focal or diffuse neurologic signs are the most common presentation in neonates (3,4), which should arouse clinicians' suspicion for CVT in neonates.

The seizure activity in two cases and the focal ischemic area in the second case were most likely caused by the retrograde extension of the dural sinus thrombus into the cortical veins with right temporal lobe infarction occurring in the second case, and IVH in the first case. Also, perhaps their thrombocytopenia is caused by CVT. In the first case, severe dehydration was the important risk factor for CVT. His symptoms progressed over one week from lethargy and poor feeding to tonic-clonic seizure. He was suffered from sagittal and both transverse and sigmoid sinus thrombosis with IVH. The patient was managed successfully. He recovered in terms of neurologic examination and development. The girl in the second case presented with symptoms resembling septic shock and severe dehydration which both are important risk factors for CVT. Her symptoms progressed over three days from fever and poor feeding to respiratory distress and grandmal seizure. She was experienced sagittal sinus thrombosis with evidence of non-hemorrhagic right temporal infarct. The patient was managed successfully and recovered to normal neurologic examination and development.

CVT should be considered in the differential

diagnosis for neonates with recent and unusual seizure or neurologic symptoms. Diagnosis of this condition is very difficult and necessitates neuroimaging studies. Transfontanel doppler ultra-sonography, although not sensitive enough to exclude cerebral venous thrombosis, is a powerful tool for the noninvasive diagnosis and monitoring of neonatal CVT (9,10). The CT scans with and without contrast should be carried out first in the emergency department in order to rule out any other causes.

Supportive care is important for all types of perinatal stroke. Early intervention can stop the progression of CVT (7). The most important point in the management of CVT is the active treatment of any underlying cause that may have predisposed the patient to thrombus formation, such as sepsis, meningitis, hypoxic-ischemic encephalopathy, dehydration, and hemodynamically significant congenital cardiac disease. Although the treatment of CVT is controversial, it is proposed that anticoagulant therapy is started (11,12). Increasing the risk of further bleeding into an already hemorrhagic infarct, heparin is supposed to prevent the progression and extension of thrombus into adjacent veins (7). No major complications occurred in preliminary studies of unfractionated heparin (UFH) or low-molecular-weight heparin (LMWH) in neonates with CVT (13), but it is not clear whether anticoagulation is beneficial in these neonates.

Prognosis for recovery of function is favorable with early therapeutic intervention. Most patients recover fully. The long-term neurologic outcome of CVT in children remained to be clarified.

These cases show the importance of recognizing neurologic symptoms as potential indicators of CVT in neonates. Because the clinical presentation, onset, and severity of CVT are relatively variable, the misdiagnosis can occur.

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