Neurosciences Quiz

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An infant with an unusual cause of lethargy

Case Presentation

A 4-month-old boy, known case of meningocele not repaired, presented to the Pediatric Emergency Department with a 3-day history of fever, lethargy, and decreased oral intake. There was no history of vomiting or seizure, and no history of apnea or cyanosis. On examination, he was febrile with a 39°C temperature, tachycardic (170/min), tachypnic (60/min), and dry oral mucosa. Neurologically, he was semiconscious on presentation with a Glasgow Coma Score of 13 out of 15, anterior fontanel opened not bulging, pupils 3 mm bilateral reactive to light, and absent reflexes on the lower limbs. After rehydration and antibiotics, a CT of the brain was performed (**Figure 1**).



Figure 1 - Brain CT without contrast.

Questions

- 1. What are the abnormalities noted on his brain CT in Figure 1?
- 2. What is the most likely underlying diagnosis, and what are the possible predisposing risk factors?
 - 3. What are the treatment modalities of this disorder?
 - 4. How common is the recurrence of this disorder?

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Answers & Discussion

- 1. *Brain CT abnormalities.* A CT of the brain without contrast shows focal hyperdensities within the course of both lateral transverse sinuses (arrows in Figure 1), which is suggestive of bilateral transverse sinus thrombosis.
- 2. *The most likely diagnosis.* Bilateral cerebral transverse sinuses thrombosis. The possible predisposing risks factors are infection (meningitis), and severe dehydration.
- 3. *Treatment.* 1) The most basic measures are necessary: supportive treatment of the seizures; treat predisposing factors. 2) Anticoagulation, if appropriate with subcutaneous heparin, or intravenous heparin. 3) Thrombolysis, thrombectomy, and surgical decompression may be considered, however, it is still controversial.
- 4. *Risk of recurrence of this disorder.* Recurrence of venous sinus thrombosis is less common than that of arterial stroke in this age group; younger patients are less likely to recur, as with patients in whom venous sinuses are recanalized.

Cerebral sinovenous thrombosis (CVST) occurs following the occlusion of one of the major cerebral veins resulting in outflow obstruction and venous congestion.¹ The estimated incidence in children including neonates is between one to 12 per 100,000/year, males are known to be more at risk, and presentation is often very nonspecific, but apnea and generalized seizures were most common in neonates.² Decreased level of consciousness, focal and diffuse neurologic signs like headache, nausea, and emesis are more common in older infants and children.³ Risk factors for CVST are present in the vast majority; in neonates these are usually adverse perinatal factors or systemic illness, including sepsis or dehydration, and in older children; neck or head infections, cardiac disease, sickle cell disease, lupus, nephritic syndrome and dehydration.³

The most important tool for diagnosis is a high index of suspicion, as well as basic brain CT, and brain MRI venography. Recurrence of venous sinus thrombosis is less common than that of arterial stroke, although it does happen with a mortality rate of 19%.² Younger patients are less likely to recur, as the venous sinuses are recanalized, in patients with relapsed risk factors, such as nephritic syndrome or inflammatory bowel disease, anticoagulation may be warranted to prevent recurrence. In management, supportive treatment of the seizures, anticoagulation, if appropriate, thrombectomy and surgical decompression may be considered, however, it is still controversial.

References

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