International Journal of Science and Research (IJSR) ISSN: 2319-7064

SJIF (2022): 7.942

Unusual Case of Cerebral Venous Thrombosis in a Young Girl with Acute Mastoiditis: A Case Report

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Abstract: <u>Background</u>: Cerebrovascular diseases are a significant factor contributing to children's morbidity and death globally. Cerebral venous thrombosis (CVT) is defined as a disruption of cerebral circulation caused by the development of a blood clot in the brain's venous system and blockage of the dural sinus, one or more brain vessels, or both. Thrombosis of the intracranial venous system represents an uncommon complication of mastoiditis, which can cause fatal neurological sequelae. However, there is an ongoing debate on the diagnostics and treatment of complicated mastoiditis in children. Early detection of cerebral venous sinus disorders and effective therapy relies on accurate brain imaging. Currently, anticoagulation therapy along with intravenous antibiotics are considered the mainstay of treatment. <u>Case presentation</u>: We describe the infrequent case of a 2 - year - old white girl who was admitted to the hospital for respiratory insufficiency. The patient's overall status deteriorated within a few minutes for which she was intubated and connected to artificial ventilation. CT scan of the brain showed massive thrombosis of the intracranial venous system and signs of mastoiditis. A course of anticoagulation therapy with low - molecular - weight heparin (LMWH) was immediately initiated. Furosemide, clindamycin and dexamethasone were added to the therapeutic regimen. <u>Conclusion</u>: The symptomatology of cerebral venous thrombosis in pediatric patients is highly variable and can range from signs of elevated intracranial pressure to focal neurologic deficits, altered level of consciousness, headache or seizures. Therefore, early detection and intervention are essential to avert complications and reduce permanent brain damage.

Keywords: Atrioventricular block, Mastoiditis, Intracranial thrombosis, Klebsiella pneumoniae, Pseudomonas aeruginosa

1. Introduction

One of the most prevalent infectious disorders affecting children of all ages is acute otitis media (AOM). Some of the most typical microorganisms associated with AOM in the pediatric population include Streptococcus pneumoniae, streptococcus, Haemophilus influenzae, Group A Moraxella Staphylococcus aureus and catarrhalis. Exceptionally, Pseudomonas aeruginosa and Klebsiella pneumoniae, can also lead to AOM in children. This contrasts significantly with the general misconception in the previous literature, according to which both these microbes are exclusively associated with chronic otitis media. The most frequent AOM complication is acute mastoiditis (AM); more severe complications such as facial paralysis, meningitis, subperiosteal, epidural, or intracerebral are still possible albeit rare [1]. Mastoiditis in children can lead to an uncommon but deadly condition called cerebral venous sinus thrombosis. Cerebral venous sinus thrombosis may often lead to elevated intracranial pressure (ICP) that is life threatening or may precede neurodevelopmental sequelae

The origin and pathophysiology of CVT in the pediatric population is still poorly understood, mainly because of its low incidence, which is estimated at 0.67 per 100 000 children [3]. Nevertheless, the prevalence of these complications has dramatically decreased over the past decades due to several contributing factors. Subsequent introduction of antibacterial agents has greatly decreased the incidence of complicated mastoiditis and has improved the overall outcomes [4].

2. Case Presentation

A previously healthy 2 - year - old white girl was admitted

to the Emergency Department for acute respiratory insufficiency. The patient was unconscious (GCS=6) and pulseless. During the physical examination, the child was hypothermic and unresponsive to both, physical and auditory stimuli. Following this, the toddler rapidly developed a lack of spontaneous breathing with an unusual agonal breathing pattern. Subsequently, CPR was immediately started, the child was intubated epinephrine was administered. Indeed, aminophylline and dexamethasone plus dextrose were added to the infusion. Continuous monitoring of vital functions was crucial in the management. Surprisingly, large amounts of yellow - to - green drainage contents were obtained from the nasal cavity following nasogastric tube insertion. During lung auscultation, decreased breath sounds on the left side along with bilateral murmurs and wheezing were detected. The child was ventilated with a bag valve mask (BVM) and connected to positive pressure ventilation (PEEP=7.25). Arterial blood gas test (ABG) showed acidosis (pH=7.209) with markedly decreased bicarbonate levels (HCO3=14.40). Therefore, the patient was transferred to our center for further therapeutic guidance.

At the time of admission, the child was in a comatose state (GCS=8) and presented with a course of green stool and repetitive episodes of vomiting over the previous 2 days. The patient was intubated and connected to artificial ventilation. During ECG examination, a 2nd - degree atrioventricular (AV) block, Mobitz type I was detected. ABG test revealed metabolic acidosis with severe hyponatremia (Na=117) and hypokalemia (K=1.44). Therefore, the patient was administered potassium parenterally. CT scan of the brain revealed massive thrombosis of the left transverse sinus, the confluence of sinuses and the sigmoid sinus up to the proximal part of the internal jugular vein (IJV).

Volume 12 Issue 7, July 2023

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Paper ID: SR23722013749 DOI: 10.21275/SR23722013749 1584

International Journal of Science and Research (IJSR)

ISSN: 2319-7064 SJIF (2022): 7.942

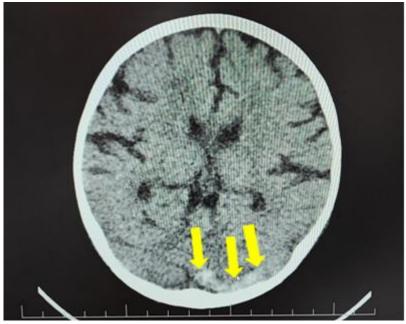


Figure 1: Non - contrast axial CT of the brain. Large thrombus within the confluence of sinuses that extends into the left transverse sinus

After consultation with the hematologist, the patient initiated a course of anticoagulation therapy with low - molecular - weight heparin (LMWH) for the management of intracranial thrombosis. Antiedematous therapy with furosemide and parenteral clindamycin was administered. Chest radiographic findings showed left perihilar and paracardial region infiltrates along with throat infiltrates. Culture of the sputum and bronchoalveolar lavage revealed Extended - spectrum - beta - lactamase (ESBL) producing Klebsiella pneumoniae and Pseudomonas aeruginosa, evaluated as a

nosocomial infection. Ventilation tubes were placed following the recommendations of the otorhinolaryngologist. The next day, the child was extubated after recovery of spontaneous breathing. The child's overall condition improved and recovered consciousness. However, generalized hypotonia along with severe quadriparesis and paraplegia persisted. Finally, the child was transferred to the Department of Paediatric Neurology for further investigation.

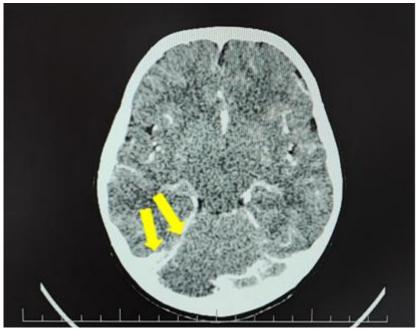


Figure 2: Contrast axial CT of the brain. Thrombosis of the sigmoid sinus and proximal part of the IJV

3. Discussion

CVST associated with mastoiditis is more commonly seen in children due to certain anatomical factors. For instance, the mastoid air cells, which are present in the mastoid bone, are not fully developed in children. This can facilitate the spread of infection to nearby structures and increase the risk of complications such as CVT. Instead, AM represents a severe complication of an acute (sometimes chronic) otitis media, favored by several factors (anatomic condition of the

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Paper ID: SR23722013749 DOI: 10.21275/SR23722013749 1585

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SJIF (2022): 7.942

temporal bone, age, bacterial flora, immunological defects, etc) [5]. Pediatric CVST occurs due to both intravascular and vascular factors, and dehydration is a common risk factor for all ages [6]. Risk factors contributing to pediatric CVST include infections, trauma, recent intracranial surgery and prothrombotic disorders [7]. The symptomatology is highly variable in children, which makes this condition a difficult diagnosis that requires a certain degree of expertise in the clinical setting. Children may present with symptoms such as progressive headache, papilledema, diplopia secondary to sixth cranial nerve palsy, progressive loss of vision, or with acute focal deficits. Seizures, lethargy, and confusion are common [8].

Neuroimaging is the principal basis for the diagnosis of CVST [9]. To demonstrate filling deficiencies in the cerebral venous system, a preliminary computed tomographic (CT) scan or magnetic resonance (MR) venography is required. The primary sign of acute CVT on a non - contrast CT is hyperdensity of a cortical vein or dural sinus [10]. The primary finding suggestive of CVST in the traditional magnetic resonance imaging (MRI) procedure is the absence of a flow - void signal in the conventional T1 and T2 sequences. The diagnosis of sinus thrombosis is based on the presence of a hyperintense signal on T1 and hyperintense or isointense signal on T2 and lack of flow signal (filling defect) in the MRV [11]. The prevalence of other significant findings in emergency MRI (15%) is only slightly higher than that previously reported for nonenhanced CT (11%) [12].

Management for CVST remains controversial. Supportive treatment is more or less uniform for all pediatric age groups and includes rehydration, antibiotics for suspected sepsis, antiepileptic drugs for seizure control and measures to reduce intracranial hypertension [13]. Studies have shown that anticoagulant therapy can be safely applied to children, particularly when coagulation is monitored [14, 15, 16]. However, further studies with a broader spectrum of patients are required to increase survival rates and monitor possible adverse effects. Indeed, we believe that case studies such as ours will both shed light on this unusual neurologic complication and also promote the development of unique diagnostic and therapeutic approaches to ensure a favorable outcome in the pediatric population.

4. Conclusion

CVT is a rare but serious complication that can occur as a result of mastoiditis in children. The highly unpredictable symptomatology and a long list of differentials make this condition a complicated diagnosis for most healthcare providers. Despite the large literature regarding paediatric mastoiditis, only 14 case studies have associated this condition with CVT. However, prompt recognition and treatment are crucial to prevent potential complications and minimize long - term neurological damage. As indicated parenteral earlier, treatment typically involves administration of antibiotics and, in severe cases of CVT, anticoagulation therapy may be necessary to dissolve or prevent the formation of blood clots. Surgical intervention, such as drainage of abscesses or mastoidectomy, may also be required in some cases. Despite the low incidence of this cerebrovascular complication, we still need to be aware of its existence because of the possibly fatal implications it may entail.

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Volume 12 Issue 7, July 2023

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Paper ID: SR23722013749 DOI: 10.21275/SR23722013749 1586

International Journal of Science and Research (IJSR) ISSN: 2319-7064

ISSN: 2319-7064 SJIF (2022): 7.942

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Paper ID: SR23722013749 DOI: 10.21275/SR23722013749 1587