

Case Report

Case Report Presentation: Measles as the Cause of Cerebral Venous Sinus Thrombosis in Childhood

Vaka Sunumu: Çocukluk Çağında Serebral Sinus Ven Trombozu Sebebi Olarak Kızamık

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ABSTRACT

The major cause of stroke in children is cerebral venous sinus thrombosis (CVST), which may cause neurological impairment and even death. The most frequent causes of CVST in kids are infections, followed by anemia, and then dehydration. Despite the existence of a safe and reliable vaccination, measles is an infectious viral infection that continues to be a major cause of death in young children worldwide. With a high morbidity and death rate, measles may result in neurologic consequences, such as acute demyelinating encephalomyelitis (ADEM), coma, seizures, or obtundation. In this case, a 4-year-old Syrian girl with status epilepticus and an altered sensorium was brought to the emergency room and was later identified as having CVST. There have never been any reports of CVST with measles.

Keywords: Measles, status epilepticus, cerebral venous sinus thrombosis, childhood

ÖZET

Çocuklarda inmenin başlıca nedeni, nörolojik bozukluğa ve hatta ölüme neden olabilen serebral venöz sinüs trombozudur (SSVT). Çocuklarda SSVT'nin en sık nedenleri enfeksiyonlar, ardından anemi ve dehidrasyondur. Güvenli ve güvenilir bir aşının varlığına rağmen, kızamık, dünyada küçük çocuklarda başlıca ölüm nedeni olmaya devam eden bulaşıcı bir viral enfeksiyondur. Yüksek morbidite ve ölüm oranının yanında, akut demiyelinizan ensefalomyelit (ADEM), koma, nöbetler veya şuur bozuklukları gibi nörolojik sonuçlara neden olabilir. Bu vaka çalışmasında, status epileptikuslu ve şuuru değişmiş 4 yaşındaki Suriyeli bir kız çocuğu acil servise getirilmiş ve daha sonra SVST olduğu tespit edilmiştir. Daha önce kızamık ile SVST olgu sunumu bildirilmemiştir.

Keywords: Kızamık, status epileptikus, serebral venöz sinüs trombozu, çocukluk çağı

INTRODUCTION

Cerebral venous sinus thrombosis (CVST) is a leading cause of stroke in children, resulting in neurological morbidity and possibly death (1). The most frequent

causes of CVST in children are infections, followed by anemia and dehydration. Measles is a contagious viral infection that continues to be a significant cause of de-

Received: 07.07.2022 · Accepted: 04.09.2022 · Published: 29.09.2022

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Cite this article as: Yimenicioglu S, Aynaci AM. Case Report Presentation: Measles as the Cause of Cerebral Venous Sinus Thrombosis in Childhood. *Pediatr Acad Case Rep.* 2022;1(1):25-8.

ath in young children worldwide, despite the availability of a safe and effective vaccine (2). Measles may cause neurologic complications, such as acute demyelinating encephalomyelitis (ADEM), coma, seizures, or obtundation, with morbidity and mortality (3-5). Here we present a 4-year-old Syrian girl brought to the emergency department with status epilepticus and altered consciousness and was diagnosed with CVST. CVST has not been reported before with measles.

Case Report

A 4-year-old Syrian girl was brought to the emergency department with status epilepticus and altered consciousness. It was preceded by a disturbance in walking for the last two days. She developed an exanthem rash typical of measles following two days of fever seven days ago. She had not been vaccinated before. There was no history of prior seizures. At the time of admission, she was unconscious and lethargic with a Glasgow coma scale of 8. She was intubated. She had abduction limitation in the right eye during outward gaze when examining for doll's eye movements and intact pupillary reflexes. Fundi examination was normal. There was nystagmus in both eyes. Spasticity was noticed in the left upper and lower limbs, and there were no meninge-

al signs. Deep tendon reflexes were hyperactive, with the Babinski sign present on the left. Examination revealed left hemiparesis.

Her blood counts and metabolic parameters, including sugar, urea, creatinine, electrolytes, calcium, and liver function tests, were normal: Homocysteine 18.5 (0-12 $\mu\text{mol/L}$), Protein C antigen 77.6 (72-160%), Protein S activity 64 (55-160%), Protein C activity 70 (70-140%), Factor 5 level 29 (50-150%), Dimer 3603 (0-243 ng/ml), CRP 2.69 mg/dL (normal range 0-0.35 mg/dL). The other test results were unremarkable.

Axial unenhanced brain computerized tomography (CT) showed acute hyperdense thrombosis in the superior sagittal sinus and edema with a small hemorrhage in the right frontal lobe. T2 weighted magnetic resonance imaging (MRI) showed right frontal lobe parenchymal edema and contrast-enhanced T1 weighted MRI image showed a thrombosis defect in the sagittal sinus called the empty delta sign. Enhanced venous MR angiography revealed a sinus thrombosis defect (Figure 1). Transthoracic echocardiography was normal except for minimal pericardial effusion. She had peribronchial infiltrations on her chest X-ray (Figure 1).

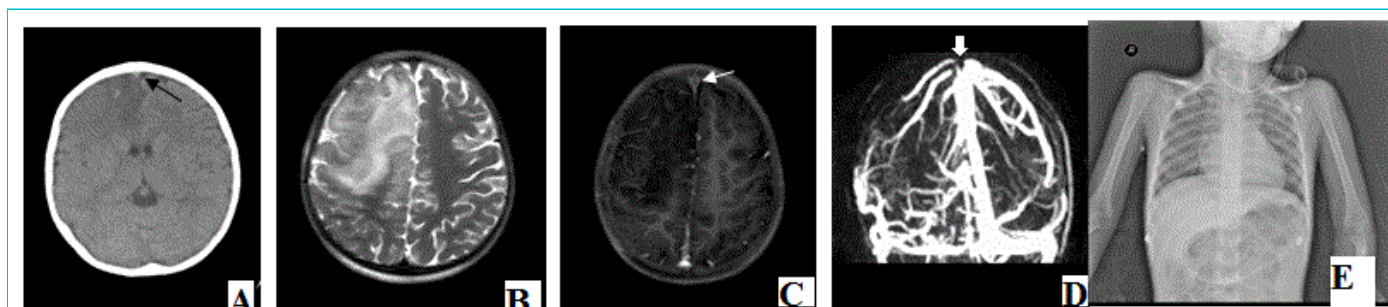


Figure 1. (A) Axial unenhanced brain CT shows acute hyperdense thrombosis in the superior sagittal sinus (black arrow) and edema with a small hemorrhage in the right frontal lobe. (B) An axial T2 weighted MR image presents right frontal lobe parenchymal edema. (C) An axial contrast-enhanced T1-weighted MRI of the sagittal sinus shows a thrombosis defect (white arrow) known as the empty delta sign. (D) Coronal enhanced venous magnetic resonance venography reveals a sinus thrombosis defect (thick white arrow). On a chest X-ray, there is peribronchial infiltration (E).

The patient had generalized tonic-clonic seizures. She was treated with levetiracetam and then sodium valproate for intractable seizures. Then a midazolam infusion was started. Dexamethasone was added at the dose of cerebral edema. Levetiracetam and sodium valproate were continued. She was extubated on the tenth day of internalization. The patient did not have bleeding and received subcutaneous injections of LMWH (two times a day, 180 U/kg BID) for two weeks,

and then we switched to acetylsalicylic acid at discharge. At the time of discharge, she had left hemiparesis and could walk without support. Her speech was not impaired. She was advised to continue anticonvulsants and acetylsalicylic acid for the next three months. The consent of the patient's parents was obtained for this study.

DISCUSSION

Measles is spread by droplets using the respiratory route and has an incubation period of 8 to 12 days. This is followed by 2-4 days in which a fever sets in, followed by the characteristic trio of cough, coryza, and conjunctivitis (6,7).

The classic rash begins on the face and moves down the body, eventually forming a confluence. Patients are contagious for 1-2 days prior to the beginning of symptoms and for four days after the rash first appears (3,4). Sometimes children may lack these features at the beginning. Acutely, complications of measles can include pneumonia, superinfection of cutaneous lesions, and most concern to neurologists, acute encephalitis. The most frequent manifestations are convulsions, lethargy, coma, and irritability (7). Our patient had intractable seizures with status epilepticus. Headaches, irregular breathing, exaggerated movements, shock, confusion, tremors, and disorientation are less frequent. Symptom onset occurs within eight days of the onset of measles in almost all cases (7). Our patient had a fever following a rash seven days before admission. She did not have pneumonia but had bronchitis.

Most cases of primary measles encephalitis actually represent a post-infectious demyelinating syndrome, acute post-measles encephalitis (7,8). Our case did not have meningeal irritation signs or radiological evidence of encephalitis. Most deaths due to measles are caused by pneumonia, diarrhea, or neurological complications in young children, severely malnourished or immunocompromised individuals, and pregnant women (9).

Cerebral venous sinus thrombosis (CVST) has not been reported before with measles. CVST clinical signs may range from asymptomatic to neurological symptoms such as headache, vomiting, lethargy, 6th cranial nerve palsy, and newborn seizures (1,10).

CVST is diagnosed solely with MRI, especially magnetic resonance venography (MRV), or CT venography, due to vague clinical characteristics. Infections of the head and neck, either contiguous (such as mastoiditis) or systemic (such as nephritis or nephrotic syndrome), tumors, and head injury with skull fractures, are the most common causes of CVST. To date, there have been few reports of this condition among children (10).

Early diagnosis and aggressive treatment have resulted in a low mortality trend in CVST (1). Anticoagulant therapy, in conjunction with adequate hydration,

antibiotics, and anemia treatment, may result in a better outcome (1).

Javed et al. (1) reported the clinical spectrum and outcome of CVST in children. They found that fever was the most frequently encountered symptom, followed by headache and lethargy. All patients that they reported had thrombosis of the superior sagittal sinus, while a quarter had additional thrombosis of the internal cerebral veins. Additionally, ischemic and hemorrhagic infarctions were observed. They reported the CVST-related mortality rate as 6.5%.

Lazzareschi et al. (10) included 24 CVST-affected children. Infection was the most common cause, followed by trauma in 25% of instances, except one patient who had a thrombophilia component. The superficial venous system was the most common site of thrombosis. They had administered anticoagulant medication with low molecular weight heparin (LMWH) to all of the children for an average of 86 days. Only one patient died because of systemic issues. At discharge, none of the survivors complained of any neurological sequela from CVST.

Low molecular weight heparin can be administered subcutaneously twice daily at a dose of 1 mg/kg/dose (equivalent to 100 UI/kg/dose). The dose must then be modified based on the results of the anti-factor Xa assays. Subcutaneous LMWH injections (two times a day, 180 U/kg BID) are safe and effective for two weeks before switching to oral warfarin for three months (10,11).

In conclusion, acute measles infection and its unusual complications are conceivable even if you have a proven immunization history. The necessity of recognizing vaccination failure and isolating patients adequately cannot be overstated.

Conflict of Interest / Çıkar Çatışması

The authors declared no conflicts of interest with respect to authorship and/or publication of the article.

Financial Disclosure / Finansal Destek

The authors received no financial support for the research and/or publication of this article.

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