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Acute cerebellitis with hydrocephalus

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CASE REPORT

Acute Cerebellitis with Hydrocephalus

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ABSTRACT

We report a case of an 8 years old child who presented with sudden onset of headache and vomiting. He had broad-based gait and intention tremors on admission. MRI brain revealed isointense signals on T1-weighted imaging and hyperintense signals on T2-weighted imaging. Cerebellar swelling was also identified with significant mass effect obliterating the fourth ventricle. CT head showed prominent third and lateral ventricles. He was treated with high dose corticosteroids and required an external ventricular drain (EVD) insertion. He made an uneventful recovery and suffered no neurologic deficit. The clinical and radiologic findings in this boy were consistent with cerebellitis complicated by hydrocephalus.

Key Words: *Acute cerebellitis. Headache. Hydrocephalus.*

INTRODUCTION

Acute cerebellitis is a rare inflammatory disorder, observed both as a sequel to acute viral infection or vaccination. It has gained much recognition in the recent years, probably due to widespread use of magnetic resonance imaging (MRI).[1] The course is variable, from benign and self-limiting to life-threatening. Acute cerebellitis with cerebellar swelling, hydrocephalus and brainstem compression is a rare but life-threatening condition.[2] Various causative pathogens, such as *Varicella-zoster*, EBV, *Coxsackie* virus, *Echo* virus, mumps, *Rubella*, *S. pneumonia* and *Mycoplasma* have been identified in children.[3-5]

We report a case of an 8 years old child who presented with acute cerebellitis complicated by hydrocephalus.

CASE REPORT

An 8 years old boy presented with a one week history of headache and vomiting, that have been progressive in severity and frequency. He also had a seizure episode before arriving at the emergency room that was described as being focal, confined to the left upper limb, lasting 3 minutes and self-aborted. He had no history of fever, fall or head trauma. There was no recent history of viral infection or immunization. His past medical history was unremarkable and he was doing well in mainstream school.

At his arrival, he was well oriented and vitally stable. On examination, he was found to have brisk reflexes and anisocoric pupils (right pupil dilated, sluggish papillary reflex). He had history of eye trauma about 3 months back. Fundoscopy revealed bilateral papilloedema. Visual acuity was 6/6 in both eyes and he had no complaints of diplopia or blurring of vision. The remaining physical examination was unremarkable. An initial assessment of migraine headache was made in the ER and he was put on pain medications. His initial laboratory workup and EEG were reported normal. Neurological examination showed intention tremors, difficulty in performing finger-nose test and a wide-based gait.

An MRI brain done at some other hospital revealed abnormal signals in the cerebellar hemispheres bilaterally. These signals appeared isointense on T1 and hyperintense on T2 weighted images. Cerebellar swelling was also identified with significant mass effect obliterating the fourth ventricle. The findings were consistent with cerebellitis.

The child continued to have persistent headache and vomiting so pulse treatment with high dose corticosteroids was started. Acyclovir and appropriate antibiotic cover was also given in view of possible meningoencephalitis.

He experienced a sudden episode of apnoea and bradycardia (H/R 44/minute) followed by unresponsiveness. He was intubated, shifted to PICU and a CT head was done. The CT scan showed prominent ventricles especially the third ventricle and temporal horns of the lateral ventricles. External ventricular drain (EVD) was immediately placed and an opening pressure of 20 cm H_2O was recorded. Examination of the CSF revealed 10 white cells/mm^3, 1743 red cells/mm^3, protein 9 mg/dl and glucose 110 mg/dl. CSF PCR analysis for herpes simplex virus (HSV) and acid-fast bacilli (AFB) were negative. No virus was isolated from CSF or blood culture. C-reactive protein was < 0.3 mg/dl. ANA done in view of an autoimmune etiology was also negative. An MRI brain was repeated after EVD insertion that showed persistence of hyperintense signals on T2-weighted images in bilateral cerebellar hemispheres. However,

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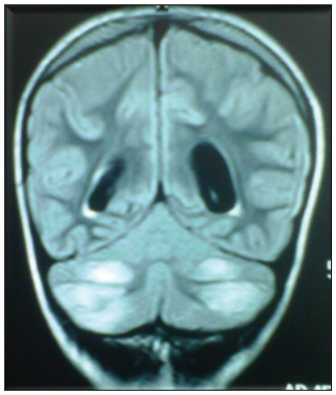

Figure 1: Hyperintense signals seen in bilateral cerebellar hemispheres on T2-weighted images.

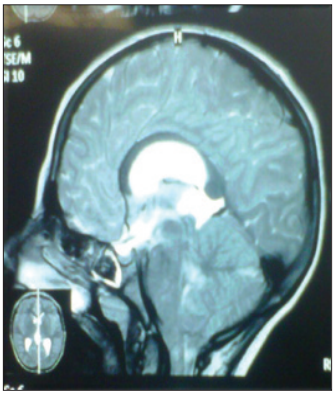

Figure 2: Cerebellar swelling resulting in significant mass effect obliterating the fourth ventricle.

the cerebellar swelling had reduced with no significant mass effect on the posterior fossa structures.

DISCUSSION

Owing to the widespread use of MRI, acute cerebellitis in children is now being reported more frequently.[1] Over the past few years, cases of cerebellitis presenting as cerebellar swelling with hydrocephalus have been seen in children and documented.[6,7] Cerebellitis is an inflammatory disorder characterized by headache, vomiting, ataxia, tremors, seizures and altered sensorium.[3,5-8] The etiology is thought to be post-infectious (EBV, HSV, Varicella, Mumps, Measles, *Mycoplasma pneumonia* etc.) post-vaccination or autoimmune in some cases.[3-5] Acute cerebellitis usually occurs bilaterally and symmetrically however, Shkalim *et al*. have mentioned quite a few case reports of hemicerebellitis in children aged 4 - 15 years.[5]

MRI is the imaging modality of choice.[5,9] Typically, MRI reveals T2-weighted cerebellar hemispheric hyperintensities with cerebellar swelling.[1,4,6,10] Hydrocephalus with posterior fossa compression is a recognized grave complication of cerebellitis. It is usually associated with obstruction at the level of the fourth ventricle.[2,4-7] Ribaupierre and colleagues have reported two cases where resection of cerebellar tonsils was performed to avoid secondary brainstem lesions. Both the children made full recovery.[4] Both medical and surgical therapeutic options are available for cerebellitis. Medical management includes treatment with high dose corticosteroids whereas surgical options comprise of external ventricular drainage, posterior fossa decompression, or both.[4,6,8]

This child had bilateral cerebellar swelling with hydrocephalus. He responded to the combined treatment with steroids and EVD placement. He did not require posterior fossa decompression. However, surgical decompression should not be delayed if the child does not respond immediately after EVD insertion. It is recommended that surgery should be performed early to avoid brainstem compression and irreversible damage.[1,4] With timely intervention, the prognosis is generally good and recovery is almost complete,[1,4,5] though deaths have also been reported in the literature where cerebellitis complicated tonsillar or brainstem herniation.[2,4,8] Cerebellar mutism is a rarely reported complication of cerebellitis, causing a complete but transient loss of speech. Fantacci described a 6 years old girl with cerebellar mutism. She presented with dysarthria that persisted for several months on follow up.[9] The reported patient had no dysarthria or mutism. He responded remarkably after CSF drainage and suffered no neurologic deficit.

Therefore, we conclude that cerebellitis complicated by hydrocephalus and posterior fossa compression may manifest a fulminant course and warrants timely and appropriate intervention. MRI is the imaging modality of choice and prompt surgical management can be life saving in case of failure of medical treatment.[4,5,8]

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