CASE REPORT

Complicated acute cerebellitis with obstructive hydrocephalus and tonsillar herniation in a child

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SUMMARY

Acute cerebellitis (AC) is a rare inflammatory syndrome presenting as cerebellar dysfunction, seen more frequently in children. AC can have a variable course with features of cerebellar dysfunction, raised intracranial pressure and neurological deficits, and can sometimes even be potentially fatal due to complications such as obstructive hydrocephalus and brainstem compression, warranting surgical intervention. We report a case of a 12-year-old boy who presented with raised intracranial pressure and ataxia. Imaging with CT and MRI showed AC with obstructive hydrocephalus and tonsillar herniation. He was managed with medications for raised intracranial pressure and with ventriculoperitoneal shunt, and he recovered completely over a period of 2 weeks. Imaging has an important role in the diagnosis of AC and in differentiating it from acute cerebellar ataxia, which has a more benign course. It is crucial to diagnose and promptly manage the rarely occurring but life-threatening complications of AC.

weight less than third centile for age. Gait ataxia was present and deep tendon reflexes were exaggerated. Fundus examination revealed bilateral papilloedema. There were no other neurological deficits nor signs of meningeal irritation.

INVESTIGATIONS

Routine blood investigations showed neutrophil-predominant leucocytosis. After placing the ventriculoperitoneal (VP) shunt to decompress the ventricles, lumbar puncture was done and cerebrospinal fluid (CSF) was obtained. CSF analysis was unremarkable. Serum viral markers and work-up for tuberculosis were negative. Plain CT of the brain (figure 1) showed swollen hypodense cerebellar hemispheres compressing the fourth ventricle and brainstem with obstructive hydrocephalus. Contrast MRI of the brain (figure 2) showed oedematous cerebellar hemispheres with bilaterally symmetrical hyperintense signals in T2/fluid-attenuated inversion recovery sequences without restricted diffusion or contrast enhancement, compressing the fourth ventricle and brainstem and

BACKGROUND

The clinical presentation of acute cerebellitis (AC) can be very vague, with features of cerebellar dysfunction such as ataxia and nystagmus, or rarely with features of raised intracranial pressure. It may be difficult to make a confident diagnosis based on clinical features alone as many other clinical conditions can present similarly. Our patient presented with obstructive hydrocephalus and tonsillar herniation, both rarely reported and potentially fatal complications of AC. This case is reported because prompt diagnosis with the help of imaging, appropriate medical management and timely surgical intervention helped the patient recover completely. We would like to stress the role of MRI to confidently diagnose AC and its complications early in the course of the disease when clinical features and laboratory findings are equivocal. We would also like to mention the importance of surgical ventricular decompression in selected cases, which proved life-saving in our case.

CASE PRESENTATION

A 12-year-old boy presented with a 7-day history of low-grade intermittent fever, headache, disturbed sleep, projectile vomiting and gait abnormalities. There was no recent history of vaccination. On examination he was conscious and oriented but irritable. He was emaciated with both height and

Figure 1 (A–D) Plain CT images of the brain on axial plane showing swollen hypodense cerebellar hemispheres (arrows) with obstructive hydrocephalus.

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Figure 2 (A–D) Post-VP shunt MRI images. Fluid-attenuated inversion recovery image on axial plane (A) and T2-weighted image on coronal plane (C) showing swollen cerebellar hemispheres with diffuse hyperintense signals (asterisk) and compression of fourth ventricle and brainstem. VP shunt tube is seen in situ within the right lateral ventricle (arrow) (B). T1-weighted image on sagittal plane (D) showing 5 mm tonsillar descent below the foramen magnum (arrow). VP, ventriculoperitoneal.

5 mm tonsillar descent below the foramen magnum. Diagnosis of AC was made based on imaging findings.

DIFFERENTIAL DIAGNOSIS

In a setting of 7-day history with low-grade intermittent fever, features of raised intracranial pressure and emaciation, *Mycobacterium tuberculosis* infection of the central nervous system was considered to be a likely aetiology. Other differential diagnoses included intracranial space-occupying lesion, acute disseminated encephalomyelitis (ADEM) and viral encephalitis.

TREATMENT

Intravenous 3% saline and oral glycerol were given for raised intracranial pressure soon after admission. Antiviral (acyclovir) and antibiotic (ceftriaxone) were started empirically, and antiepileptic (phenytoin) was added prophylactically. As the signs of raised intracranial tension continued to rapidly worsen, ventriculoperitoneal shunt was done. As CT did not show any intracranial space-occupying lesion, *M. tuberculosis* infection was considered a possible aetiology, in which case hydrocephalus is expected to take a fairly long time to resolve after starting antitubercular treatment with steroids. In view of this, VP shunt was chosen over external ventricular drainage to decompress the ventricular system. Phenytoin and acyclovir

were withdrawn after 3 days and antibiotic was stopped after 5 days.

OUTCOME AND FOLLOW-UP

The patient improved symptomatically with the above-mentioned line of management. Recovery was complete without any residual neurological deficit and he was discharged after 12 days of hospital stay.

DISCUSSION

AC and acute cerebellar ataxia represent a spectrum of diseases clinically presenting as cerebellar dysfunction. These are more frequent in early childhood and can be infectious, postinfectious, postvaccination or autoimmune. Commonly described causative agents are varicella, measles, mumps and rubella.¹ Acute cerebellar ataxia of childhood usually has a benign course characterised by cerebellar signs such as truncal and gait ataxia, nystagmus, dysarthria and hypotonia, and responds to medical management. AC represents the other end of the spectrum and presents with clinical features such as altered sensorium and raised intracranial pressure, along with signs of cerebellar dysfunction, and can be potentially fatal in exceptional situations, with complications such as obstructive hydrocephalus, brainstem compression and tonsillar herniation warranting surgical intervention.² Many cases of AC have been reported in literature, but there are only a few reported cases of fulminant AC with potentially fatal complications as in our case. Levy *et al* reported a case of sudden death from acute fulminant cerebellitis in a 13-year-old ballet dancer who had a history of influenza-like illness 10 days prior to the cerebellitis episode. The patient had brainstem compression and obstructive hydrocephalus on imaging, and her condition deteriorated rapidly and progressed to death.³ Another case report by Melaiki *et al* mentions about a case of AC with obstructive hydrocephalus and brainstem compression in an 8-year-old child similar to our case which showed favourable response with medical management with high-dose steroids. Mild residual cerebellar atrophy was seen in follow-up MRI.⁴

Imaging has an important role in the diagnosis of cases presenting with features of cerebellar dysfunction with or without features of raised intracranial tension. CT of the brain in acute phase helps to detect hydrocephalus, oedema of cerebellar hemispheres or brainstem compression, and can give a clue to the diagnosis and impending complications in appropriate clinical settings. Radiologist should have a high degree of suspicion to diagnose AC while reporting CT images of cases that present with vague cerebellar signs/obstructive hydrocephalus without any obvious lesion detectable in CT, especially in paediatric age group. Radiologists can suggest further evaluation with MRI for confirmation of the diagnosis. MRI clearly detects inflammatory changes of the cerebellar hemispheres. Bilateral diffuse cerebellar hemispheric signal intensity changes are the most common imaging findings in a typical case of AC. MRI also helps to exclude other clinical conditions with similar presentation, such as ADEM, Lhermitte-Duclos disease and space-occupying lesions of posterior fossa. The role of diagnostic lumbar puncture in the absence of any contraindications is also significant as it can rule out infective aetiologies such as tuberculous meningitis or viral encephalomyelitis. Management of AC is symptomatic and use of antibiotics/antivirals is advised empirically even though infective aetiology is not clearly established. Administration of steroid is also recommended to reduce brain inflammation.^{5–7} Our case presented as a fulminant type of AC with potentially fatal complications such as obstructive hydrocephalus, raised

intracranial tension and tonsillar herniation. Prompt diagnosis with the help of imaging, appropriate medical management and timely surgical intervention to decompress the ventricles helped the patient recover completely over a period of 2 weeks without any residual neurological deficits.

Learning points

- ▶ Acute cerebellitis (AC) is an acute inflammatory condition but not consistently benign and self-limited as contrasted with acute cerebellar ataxia of childhood, the much more common condition. Initial presentation may be similar.
- ▶ Potentially fatal complications such as obstructive hydrocephalus, brainstem compression and herniation may occur in AC and are emergencies when seen.
- ▶ Early imaging is critical in differentiating AC from the more common acute cerebellar ataxia of childhood, and in detecting serious complications.
- ▶ In cases with obstructive hydrocephalus or brainstem compression, external ventricular drainage may be life-saving.

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