CASE REPORT

Acute cerebellitis successfully managed with temporary cerebrospinal fluid diversion using a long tunnel external ventricular drain: a long-term radiological follow-up of two cases

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SUMMARY

Acute cerebellitis (AC) is a rare inflammatory childhood disorder. Although there is no consensus on standard treatment for cerebellitis, its outcome is usually favourable. We report two cases of AC in the paediatric age group, successfully managed with long tunnel external ventricular drain (EVD). The first patient was an 8-year-old boy with a history of fever and headache. Sequential MRI showed diffuse cerebellar swelling with tonsillar herniation and resulting hydrocephalus. The second patient was a 6-month-old boy who presented with high-grade fever associated with chills. CT scan of the head showed triventricular hydrocephalus with obliteration of cerebrospinal fluid spaces and cisterns. Both patients underwent immediate emergency right-sided long tunnel EVD insertion. The EVD was removed on the 9th day in the first patient and the 10th day in the second patient; the patients showed no neurological deficits at a follow-up of 2 years and 1 year, respectively.

BACKGROUND

Acute cerebellitis (AC) is a rare inflammatory childhood disorder. Depending on the extent of cerebellar oedema, AC may cause varying degrees of brainstem compression and hydrocephalus.¹ Both of these conditions are potentially life-threatening. Cerebrospinal fluid (CSF) diversion and posterior fossa decompressions are among the available surgical options, with no consensus on guidelines.² The neuroimaging of cerebellitis raises challenging differential diagnoses. Data on the disease are limited, therefore we lack detailed knowledge and understanding of the natural history, clinical and radiological features, and management of the disease. Although there is no consensus on standard treatment for cerebellitis, its outcome is usually favourable as long as the brainstem compression and hydrocephalus are treated. We report two cases where we successfully managed patients with external ventricular drain (EVD) insertion and present their long-term clinical and radiological follow-up.

CASE PRESENTATION FOR CASE 1

An 8-year-old boy presented to our emergency room, with a 5-day history of fever and headache. The child was reported to be fine before these symptoms with no history of flu-like illness or myalgia.

The fever was low-grade and the headaches were severe and diffuse. Two days after the onset of fever, the child developed projectile vomiting with worsening headache.

On examination, the child appeared slow and lethargic. His haemodynamic parameters were normal but neurological examination revealed bilateral signs of cerebellar dysfunction including dysdiadochokinesia, incoordination in movements, dysmetria, dysarthric speech, intention tremors and a broad-based gait. The child also had decreased power (MRC score 4/5) in all four limbs and had brisk deep tendon reflexes. Neither meningeal irritation nor lymphadenopathy was present.

INVESTIGATIONS

Laboratory investigations revealed a total leucocyte count of 16 000/mm³ with 81% neutrophils, haemoglobin of 12.4 g/dL and platelet count of 314 000/mm³. Plasma ammonia was 98 µg/dL and erythrocyte sedimentation rate was 5 mm/h. Bleeding profile and serum electrolytes were within normal limits. Sequential MRI showed diffuse cerebellar swelling with signal changes involving bilateral cerebellar hemispheres, low on T1-weighted images, and high on T2-weighted images, likely to be oedema. There was minimal, diffuse hemispheric contrast enhancement. The cerebellar swelling caused significant tonsillar herniation through the foramen magnum and compression of the brainstem, and was also obliterating the fourth ventricle, resulting in moderate to severe hydrocephalus (figure 1).

TREATMENT

The child remained stable for a few hours but suddenly became apnoeic and bradycardic. He was intubated and rushed to the paediatric intensive care unit (PICU). Methylprednisolone, acyclovir, ceftriaxone and vancomycin were started and an emergency right-sided long tunnel EVD was placed. Intraventricular pressure was measured to be 25 cm H₂O and the CSF was clear. The child remained in the ICU and continued to improve. CSF study showed protein of 9 mg/dL, leucocyte count was 10/mm³ with 30% polymorphs and 70% lymphocytes, glucose was 110 mg/dL, red blood cells were 1728/mm³ and no microorganisms

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Figure 1 (A and B) CT brain scan of the child at presentation. There is downward herniation of cerebellar tonsils and effacement of prepontine space and dilation of lateral and third ventricles. (C and D) MRI scan 1 week after presentation. Tonsillar herniation is still prominent and there are bilateral hyperintense signals in both cerebellar hemispheres. (E and F) Complete resolution of cerebellar oedema and hydrocephalus. Previously effaced fourth ventricle has normal appearance on 2-year follow-up scan.

were found on culture. Acyclovir was discontinued after herpes PCR was found to be negative.

OUTCOME AND FOLLOW-UP

The child was extubated on day 3 but the EVD was retained and kept at 10 cm H₂O. It was removed after challenge on day 9. A repeat MRI showed markedly decreased cerebellar oedema. The child regained a Glasgow Coma Scale score of 15/15 and was discharged on the 11th day of admission, with no neurological deficits. He remained asymptomatic at 2-year follow-up, at which time an MRI was also found to be normal.

CASE PRESENTATION FOR CASE 2

A 6-month-old boy presented to our emergency room with fever and vomiting. One week before presentation, the child had developed high-grade fever associated with chills. He had developed non-projectile vomiting the same day. There was no history of preceding sore throat or vaccination. Earlier on the day of presentation, the child had had a tonic-clonic seizure followed by a post-ictal phase, which prompted the parents to rush him to the tertiary care facility.

On examination, the infant was irritable, tachypnoeic and tachycardic, and responded only to painful stimuli. The infant

Figure 2 (A) Axial non-contrast enhanced CT brain scan. Posterior fossa appears tight secondary to swelling of the cerebellum. The fourth ventricle is effaced and prepontine space is obliterated with severely dilated right temporal horn. (B) Sagittal T2-weighted image after 1 week of presentation. There is diffuse cerebellar oedema with tonsillar herniation. Basilar artery is compressed with effacement of prepontine space. (C) At 4-month follow-up scan the swelling has completely resolved. Basilar compression is relieved and cisterns appear normal.

had up-rolled eyes with a fixed gaze, bilateral pupillary constriction and a bulging anterior fontanelle. He had increased tone in all limbs and deep tendon reflexes were brisk.

INVESTIGATIONS

Laboratory investigations revealed a deranged coagulation profile with International Normalised Ratio of 1.9. Serum electrolytes and complete blood count were normal. CT scan of the head showed triventricular hydrocephalus and a cramped posterior fossa, with obliteration of CSF spaces and cisterns.

TREATMENT

The child received vitamin K and was started on ceftriaxone, vancomycin and acyclovir in meningitic doses with intravenous dexamethasone. He also underwent immediate emergency right-sided long tunnel EVD insertion. The intraventricular pressure was more than 25 cm H₂O and CSF appeared clear. The EVD was set at 10 cm H₂O. MRI scan after EVD insertion showed diffuse swelling of the cerebellum, involving bilateral hemispheres, with prominent cerebellar folia causing obstruction of the fourth ventricle, resulting in non-communicating hydrocephalus. The signal changes were low on T1-weighted images and high on T2-weighted images, with minimal post-contrast enhancement. Significant tonsillar herniation through the foramen magnum was also seen (figure 2). A diagnosis of acute cerebellitis was made and once CSF analysis was negative for any infection, antibiotics were discontinued though the patient continued to receive steroids.

OUTCOME AND FOLLOW-UP

The infant's condition gradually improved and he started tolerating breast feed. The EVD was removed after 10 days and a repeat MRI with and without contrast, and MR venogram, showed no venous involvement and resolution of all pathological findings, including the cerebellar swelling, tonsillar herniation and hydrocephalus. The patient was discharged on oral dexamethasone tapered over several weeks. At 1-year follow-up, the patient showed no neurological deficits, and displayed normal growth and development.

DISCUSSION

The differential diagnosis of acute cerebellar dysfunction includes inflammatory or infective processes, tumours, trauma, stroke and demyelinating disease. AC is a rare childhood disorder with either viral or autoimmune aetiologies. In many cases, the infectious agent cannot be identified.³ We described cases of two children belonging to different age groups, both presenting with gradually worsening symptoms over 1 week. Both children had hydrocephalus at presentation and underwent emergency EVD. In the first of these cases, there was a history of fever preceding the signs of raised intracranial pressure, however, in the second case, there was no history of recent illness. In neither of these cases could the aetiological agent be isolated.

There was marked vasogenic cerebellar oedema apparent in the form of prominent, bilateral hypointensities on T1-weighted images and hyperintensities on T2-weighted images, involving both cerebellar hemispheres. There was minimal enhancement on contrast administration, significant pressure on the brainstem and fourth ventricle, and tonsillar herniation in both cases. A review of the literature revealed largely similar radiological features for most reported cases.⁴ Both cases were managed with temporary CSF diversion procedures. Since we were not sure of the diagnosis and speed of recovery, we chose to insert a long

tunnel EVD, which allowed us to manage these children with observation alone, until the cerebellar swelling resolved and CSF pressures returned to normal, thereby avoiding permanent CSF diversion. In acute cerebellar swelling and hydrocephalus, placing an EVD has a potential risk of upward herniation,⁵ although for the majority of cases even with a posterior fossa mass (tumours, infarcts and even haemorrhages), it has been shown to be safe.⁵ We were cognisant of the fact and avoided rapid drainage of large volume of CSF at the time of EVD, to avoid this complication.

Both children in our series were managed postoperatively in a high-dependency unit, for possible deterioration and need of posterior fossa decompression. In both cases an excellent recovery was seen with no deficit and no radiological sequel on follow-up MRI scans. Kamate *et al*³ reported a case of fulminant acute cerebellitis that led to refractory raised intracranial pressure (ICP) and death despite insertion of EVD.³ Other authors, however, have reported AC as a temporary and self-limiting phenomenon. In a review of 17 cases by Chiaretti *et al*⁴ only one case required EVD.⁶ The authors also reported vermian atrophy as a long-term sequel being present in 94% of the cases.⁶ However, in our cases, there was complete resolution of all the radiological abnormalities including decrease in areas of signal alteration, oedema and tonsillar herniation, with no evidence of vermian atrophy. AC is sometimes also present with radiological features of a tumour leading to surgical interventions such as biopsy.^{7 8} Given the subtle enhancement of cerebellar hemispheres on contrast, the possibility of a neoplastic process was raised, which we ruled out with follow-up imaging. For management of patients with AC and associated brainstem compression with hydrocephalus, we would strongly recommend immediate placement of a long tunnel EVD, with controlled CSF drainage, to minimise the risk of upward herniation. Considering the possibility of a fulminant course, we would also recommend managing these patients in high-dependency units, and continuous monitoring for changes in ICP. The EVD can be removed when the cerebellar swelling resolves and hydrocephalus subsides, which can be assessed through repeat imaging and bedside challenge of EVD, although a possibility of the patient requiring permanent CSF diversion cannot be ruled out.

Conclusion

We reported two cases of AC in the paediatric age group, successfully managed with long tunnel EVD. Long-term radiological follow-up showed no evidence of cerebellar atrophy.

Learning points

- ▶ Acute cerebellitis is a rare but potentially fatal disease presenting in the paediatric population.
- ▶ We report two cases of acute cerebellitis in the paediatric age group, successfully managed with long tunnel external ventricular drain.
- ▶ Long-term radiological follow-up in these patients showed no evidence of cerebellar atrophy.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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