THE AGA KHAN UNIVERSITY

eCommons@AKU

Section of Neurosurgery

Department of Surgery

5-10-2016

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

Muhammad Waqas

Yousaf Bashir Hadi

Sadaf Sheikh

Muhammad Shahzad Shamim

Follow this and additional works at: https://ecommons.aku.edu/pakistan_fhs_mc_surg_neurosurg

 Part of the Neurology Commons, Neurosurgery Commons, and the Surgery Commons

BMJ Case Reports

Acute Cerebellitis Successfully Managed with Temporary Cerebrospinal Fluid Diversion Using Long Tunnel Extra Ventricular Drain: A Long Term Radiological Follow Up of Two Cases

Journal:	*BMJ Case Reports*
Manuscript ID	bcr-2016-215631
Manuscript Type:	Novel treatment (new drug/intervention; established drug/procedure in new situation)
Date Submitted by the Author:	31-Mar-2016
Complete List of Authors:	Waqas, Muhammad; The Aga Khan University, Surgery; Hadi, Yousaf; Aga Khan University, Medical College Sheikh, Sadaf; Aga Khan University Shamim, Shahzad; Aga Khan University Hospital, Surgery
Keywords:	Neurosurgery < Surgery, Surgery

SCHOLARONE™
Manuscripts

BMJ Case Reports

TITLE OF CASE ***Do not include "a case report"***

Acute Cerebellitis Successfully Managed with Temporary Cerebrospinal Fluid Diversion Using Long Tunnel Extra Ventricular Drain: A Long Term Radiological Follow Up of Two Cases

SUMMARY ***Up to 150 words summarising the case presentation and outcome (this will be freely available online)***

Acute cerebellitis (AC) is a rare inflammatory childhood disorder. Although there is no consensus on standard treatment for cerebellitis, its outcome is usually favorable. Here, we report two cases of AC in pediatric age group successfully managed with long tunnel EVD. First patient was an 8 year old boy with history of fever and headache. Sequential MRI showed diffuse cerebellar swelling with tonsillar herniation and resulting hydrocephalus. The second patient was a six months old boy who presented with high-grade fever associated with chills. CT scan of head showed triventricular hydrocephalus with obliteration of CSF 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, and the patients show no neurological deficits at a follow up of 2 years and 1 year respectively.

BACKGROUND ***Why you think this case is important – why did you write it up?***

Acute cerebellitis (AC) is a rare inflammatory childhood disorder. Depending on the extent of cerebellar edema, 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 amongst available surgical options with no consensus or guidelines.[2] The neuroimaging of cerebellitis raises challenging differential diagnosis. Data on the disease is limited therefore we lack a 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 favorable as long as brainstem compression and hydrocephalus are treated. Herein we report two cases where we successfully managed patients with EVD insertion and present their long term clinical and radiological follow up.

CASE PRESENTATION FOR CASE 1 ***Presenting features, medical/social/family history***

An eight years boy presented to our emergency room with a five-day history of fever and headache. The child was reported to be fine before these symptoms with no history of any flu-like illness or myalgia. The fever was low-grade and the headaches were severe and diffuse. Two days after the onset of fever, he developed projectile vomiting with worsening headache.
On examination the child appeared slow and lethargic. His hemodynamic parameters were normal but neurological examination revealed bilateral signs of cerebellar dysfunction including dysdiadokokinesia, 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. There were no signs of meningeal irritation or lymphadenopathy.

BMJ Case Reports

INVESTIGATIONS ***If relevant***

Laboratory investigations revealed a total leukocyte count of 16000 per mm^3 with 81% neutrophils, hemoglobin of 12.4 g/dl, and platelets count of 314,000 per mm^3. Plasma ammonia was 98 ug/dl and erythrocyte sedimentation rate was 5 mm/hr. The 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 edema. There was minimal, diffuse hemispheric contrast enhancement. The cerebellar swelling caused significant tonsillar herniation through foramen magnum, compression of the brainstem and was also obliterating the fourth ventricle, resulting in moderate to severe hydrocephalus. [Fig 1]

DIFFERENTIAL DIAGNOSIS ***If relevant***

TREATMENT ***If relevant***

The child remained stable for a few hours but suddenly became apneic and bradycardic. He was intubated and rushed to the pediatric intensive care unit (PICU). Methylprednisolone, acyclovir, ceftriaxone and vancomycin were started and an emergency right-sided long tunnel EVD was placed. Intra-ventricular pressure was measured to be 25 cmH_2O and the CSF was clear. The child remained in the ICU and continued to improve. CSF study showed protein of 9 mg/dl, leukocyte count was 10/mm^3 with 30% polymorphs and 70% lymphocytes, glucose was 110 mg/dl, RBCs were 1728/mm^3, and no microorganisms 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 cmH_2O. It was removed after challenge on day 9. A repeat MRI showed markedly decreased cerebellar edema. The child regained a GCS of 15/15 and was discharged on the 11^{th} day of admission with no neurological deficits. The patient remained asymptomatic at a 2-year follow up and an MRI at 2-year follow up was also normal.

CASE PRESENTATION FOR CASE 2 ***Presenting features, medical/social/family history***

A 6 months 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 developed non-projectile vomiting the same day. There was no history of preceding sore throat or vaccination. On the day of presentation, the child had a tonic-clonic seizure followed by post ictal phase, which prompted the parents to rush to the tertiary care facility.

BMJ Case Reports

On examination, the infant was irritable, tachypneic, tachycardic, and responded only to painful stimuli. The infant 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 ***If relevant***

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

DIFFERENTIAL DIAGNOSIS ***If relevant***

TREATMENT ***If relevant***

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 intra-ventricular pressure was more than 25 cm H_2O and CSF appeared clear. The EVD was set at 10 cm H_2O. 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 foramen magnum was also seen. [Fig. 2] A diagnosis of acute cerebellitis was made and once CSF analysis was negative for any infection, antibiotics were discontinued though patient continued to receive steroids.

OUTCOME AND FOLLOW-UP

The infant's condition gradually improved and he started tolerating breast feed. 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 with normal growth and development.

DISCUSSION ***Include a very brief review of similar published cases***

The differential diagnosis of acute cerebellar dysfunction includes inflammatory or infective processes, tumors, trauma, stroke or demylineating disease. AC is a rare childhood disorder with viral or autoimmune etiologies. In many cases infectious agent cannot be identified.[3] We have presented two children belonging to different age groups, both presenting with gradually worsening symptoms over one week. Both children had hydrocephalus at presentation and

BMJ Case Reports

underwent emergency EVD. In the first of these cases there was history of fever preceding the signs of raised intra-cranial pressure however in the second case there was no history of recent illness. In both these cases etiological agent could not be isolated.

There was marked vasogenic cerebellar edema apparent in the form of prominent, bilateral hypo-intensities on T1-weighted images and hyper-intensities 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 literature reveals largely similar radiological features for most reported cases.[4] Both cases were managed with temporary CSF diversion procedure. 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 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 majority of cases even with a posterior fossa mass (tumors, infarcts, and even hemorrhages), it has been shown to be safe.[5] We were cognizant 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 high dependency unit for possible deterioration and need of posterior fossa decompression. In both cases an excellent recovery was seen with no deficit or radiological sequel on follow up MRI scans. Mahesh Kamate et al [3] reported a case of fulminant acute cerebellitis that led to refractory raised 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 Fantacci et al, [4] only one case required EVD.[6] It also reported vermian atrophy as a long-term sequel to be 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, edema and tonsillar herniation with no evidence of vermian atrophy.

Acute cerebellitis sometimes also present with radiological features of tumor leading to surgical interventions such as biopsy.[7,8] Given 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 minimize 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 have reported two cases of AC in pediatric age group successfully managed with long tunnel EVD. Long term radiological follow up showed no evidence of cerebellar atrophy.

LEARNING POINTS/TAKE HOME MESSAGES ***3 to 5 bullet points – this is a required field***

- Acute Cerebellitis is a rare, but potentially fatal disease presenting in the pediatric population.
- We report two cases of Acute Cerebellitis in pediatric age group successfully managed with long tunnel EVD.

BMJ Case Reports

- Long term radiological follow up in these patients showed no evidence of cerebellar atrophy.

BMJ Case Reports

REFERENCES ***Vancouver style (Was the patient involved in a clinical trial? Please reference related articles)***

[1] Pruitt AA. Infections of the cerebellum. Neurologic clinics. 2014 Nov 30;32(4):1117-31.

[2] De Ribaupierre S, Meagher-Villemure K, Villemure JG, Cotting J, Jeannet PY, Porchet F, Roulet E, Bloch J. The role of posterior fossa decompression in acute cerebellitis. Child's Nervous System. 2005 Nov 1;21(11):970-4.

[3] Kamate M, Chetal V, Hattiholi V. Fulminant cerebellitis: a fatal, clinically isolated syndrome. Pediatric neurology. 2009 Sep 30;41(3):220-2.

[4] Montenegro MA, Santos SL, Min Li L, Cendes F. Neuroimaging of acute cerebellitis. Journal of Neuroimaging. 2002 Jan 1;12(1):72-4.

[5] Cuneo RA, Caronna JJ, Pitts L, Townsend J, Winestock DP. Upward transtentorial herniation: seven cases and a literature review. Archives of neurology. 1979 Oct 1;36(10):618-23.

[6] Chiaretti A, Fantacci C, Bersani G, Valentini P, Pierri F, Mariottti P, Serranti D. Acute pediatric cerebellitis and mutism. Case report and review of the literature. Signa Vitae. 2011 Oct 1;6(2):78-81.

[7] Amador N, Scheithauer BW, Giannini C, Raffel C. Acute cerebellitis presenting as tumor. Journal of Neurosurgery: Pediatrics. 2007

[8] Jabbour P, Samaha E, Lahoud GA, Koussa S, Abadjian G, Nohra G, Rizk T, Moussa R, Okais N. Hemicerebellitis mimicking a tumour on MRI. Child's Nervous System. 2003 Feb 1;19(2):122-5.

FIGURE/VIDEO CAPTIONS ***figures should NOT be embedded in this document***

Caption figure 1:

F1 a and 1b show the CT scan brain of the child at presentation. There is downward herniation of cerebellar tonsils and effacement of prepontine space and dilation of lateral and third ventricles. In Fig 1c and 1d is MRI scan one week after presentation. Tonsillar herniation is still prominent and there is bilateral hyperintense signals in both cerebellar hemispheres. Fig 1e and 1f demonstrate complete resolution of cerebellar edema and hydrocephalus. Previously effaced fourth ventricle has normal appearance on two year follow scan.

Caption of Figure 2:

Fig 2a shows an axial non contrast enhanced CT scan of brain. Posterior fossa appears tight secondary to swelling of cerebellum. Fourth ventricle is effaced and pre-pontine space is obliterated with severely dilated right temporal horn. Fig 2b shows sagittal T2 weighted image after 1 week of presentation. There is diffuse cerebellar edema with tonsillar herniation. Basilar artery is compressed with effacement of pre-pontine space. Fig 2c At 4 month follow up scan the swelling has completely resolved. Basilar compression is relieved and cisterns appear normal.

PATIENT'S PERSPECTIVE ***Optional but encouraged***

Copyright Statement

BMJ Case Reports

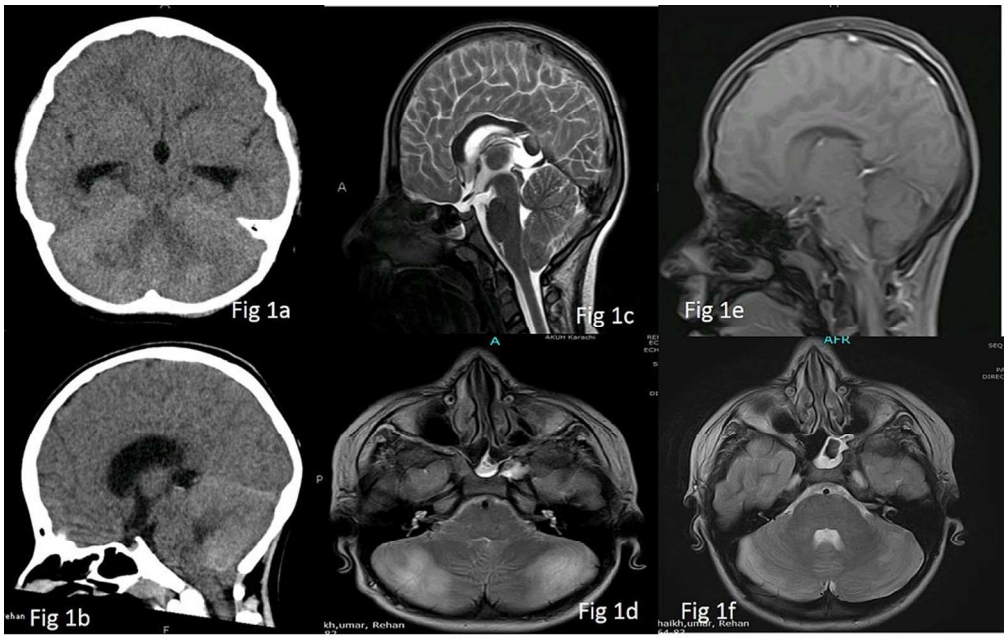
I, *Muhammad Waqas* The Corresponding Author, has the right to assign on behalf of all authors and does assign on behalf of all authors, a full assignment of all intellectual property rights for all content within the submitted case report (other than as agreed with the BMJ Publishing Group Ltd) ("BMJ")) in any media known now or created in the future, and permits this case report (if accepted) to be published on BMJ Case Reports and to be fully exploited within the remit of the assignment as set out in the assignment which has been read. http://casereports.bmj.com/site/misc/copyright.pdf.

Date: 3/29/2016

PLEASE SAVE YOUR TEMPLATE WITH THE FOLLOWING FORMAT:

Corresponding author's last name and date of submission, eg,

Smith_October_2013.doc



F1 a and 1b show the CT scan brain of the child at presentation. There is downward herniation of cerebellar tonsils and effacement of prepontine space and dilation of lateral and third ventricles. In Fig 1c and 1d is MRI scan one week after presentation. Tonsillar herniation is still prominent and there is bilateral hyperintense signals in both cerebellar hemispheres. Fig 1e and 1f demonstrate complete resolution of cerebellar edema and hydrocephalus. Previously effaced fourth ventricle has normal appearance on two year follow scan.
90x57mm (300 x 300 DPI)

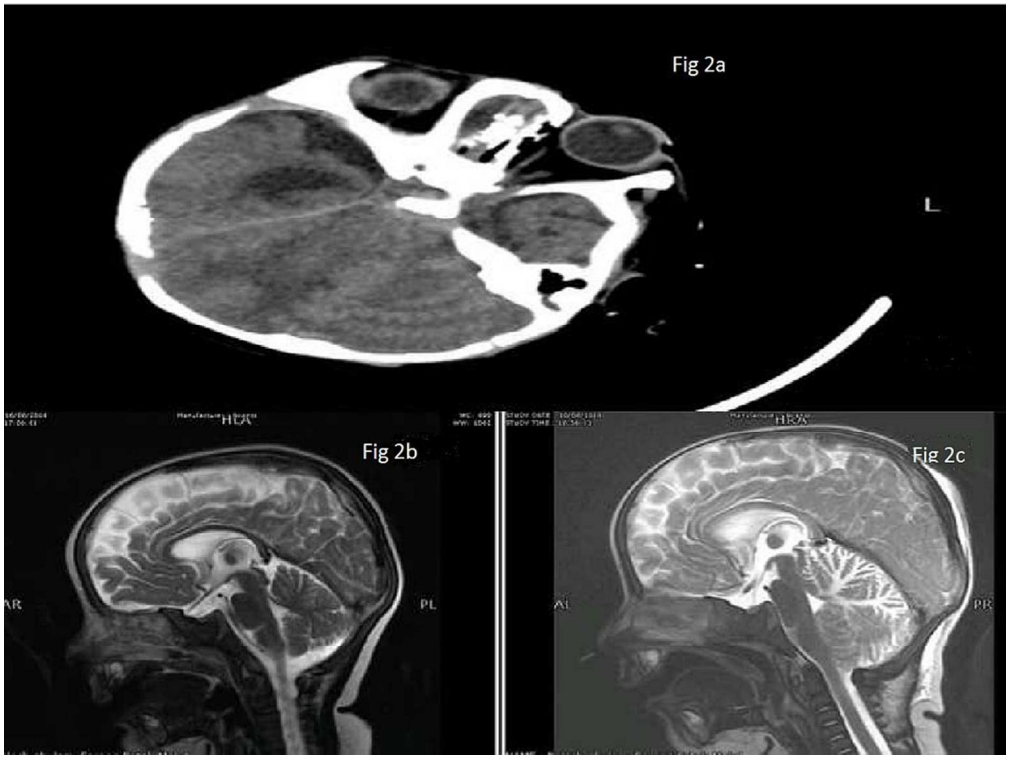


Fig 2a shows an axial non contrast enhanced CT scan of brain. Posterior fossa appears tight secondary to swelling of cerebellum. Fourth ventricle is effaced and pre-pontine space is obliterated with severely dilated right temporal horn. Fig 2b shows sagittal T2 weighted image after 1 week of presentation. There is diffuse cerebellar edema with tonsillar herniation. Basilar artery is compressed with effacement of pre-pontine space. Fig 2c At 4 month follow up scan the swelling has completely resolved. Basilar compression is relieved and cisterns appear normal.
90x67mm (300 x 300 DPI)