CASE REPORT

Chiari malformation and tuberculous meningitis: aetiology and management

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SUMMARY

This is the first reported case of a Chiari 1 malformation in association with tuberculous (TB) meningitis. We present a case of a 23-year-old woman with a 2-week history nocturnal fever, vertigo, headache and projectile vomiting. She had nystagmus, scanning speech, bilateral papilloedema and ataxia. Cranial imaging showed a 10 mm tonsillar herniation. Posterior fossa decompression was done. Because the patient's gamut of symptoms was highly suspicious for a central nervous system infection, a lumbar tap was done which revealed TB meningitis. Four years later, after anti-TB medications and rehabilitation, all her symptoms except gait instability resolved.

BACKGROUND

Chiari 1 malformation (CIM) is defined radiologically as tonsillar herniation of 3–5 mm below the foramen magnum.¹ The disease usually presents in adolescence and early adulthood as progressive occipital headache accompanied by cerebellar signs and symptoms.¹ Tuberculous (TB) meningitis, is a central nervous system infection from *Mycobacterium tuberculosis*.² Without ample treatment patients with TB meningitis have a mortality rate of 69% in underdeveloped areas.² The Philippine General Hospital census reveals around 250 cases of central nervous system infection each year, of these, 75%–90% is due to TB meningitis.³

No case report was found in the literature to present the two conditions in one patient. The prevailing notion that a Chiari malformation is an exclusively congenital condition has been challenged by reports of an acquired form of Chiari malformation, associated with pseudotumour cerebri and paediatric patients who have undergone lumbar shunting procedures.^{4–5} In addition, the definitive surgical management of CIM with hydrocephalus is also up for the debate.⁶ The presence of concomitant TB meningitis further complicates the surgical decision-making.

CASE PRESENTATION

A 23-year-old Filipina data encoder from Manila was admitted at the emergency department for vomiting and severe headache. She had a 2-week history of gradually progressive frontal headache, radiating to the occiput associated with nocturnal cough, vertigo, projectile vomiting, unsteady gait, upward deviation of primary gaze and nocturnal fever.

At the emergency department, she was afebrile, drowsy, arousable to name calling, oriented and still following simple verbal commands. Her visual acuity was limited to counting fingers at 14 inches. Funduscopy revealed bilateral grade 3 papilloedema. Her pupils were dilated at 5 mm but still briskly reactive to light. Though primary gaze was deviated upward, she still had full extraocular muscle movements. Cerebellar signs of dysmetria on all extremities and bidirectional nystagmus on all directions of gaze were found. Motor and sensory testing elicited no lateralising signs.

A cranial MRI revealed communicating hydrocephalus and a Chiari malformation characterised by tonsillar herniation of 10 mm (figure 1A,B). A fibrous dysplasia in the left aspect of the frontal bone extending to the left frontal sinus and to the underlying extra-axial region of the left frontal convexity was also found (figure 1C,D).

The preoperative axial MRI demonstrating communicating hydrocephalus is illustrated further in figure 2A–C. Posterior fossa decompression (PFD) by suboccipital craniectomy with C1 laminectomy and duroplasty was done on the sixth hospital day to address Chiari malformation and the patient's persistent symptoms. A day after the operation, her headache had completely disappeared and her gaze shifted to the midline. Serial postoperative cranial CT scans showed decrease in hydrocephalus (figure 2D–I). Lumbar tap was done and revealed elevated protein and white blood cell count (table 1).

Treatment with isoniazid, rifampicin, pyrazinamide and ethambutol was started after the lumbar tap yielded a positive TB PCR test. She was discharged on the 21st hospital day headache free but still with dysmetria, titubation and a wide-based stance.

OUTCOME AND FOLLOW-UP

Four years after discharge, she was seen in the outpatient department still headache free. She finished her prescribed rehabilitation and medical regimen of 9 months of treatment with anti-TB medications. Visual acuity had improved to 20/20. Papilloedema and titubation had resolved. Dysmetria and nystagmus were much less than her neurological physical examination prior to discharge. She was able to ambulate with assistance but her gait was still moderately ataxic (video 1). Despite these limitations, she has managed to maintain a

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Figure 1 (A) MRI T1 sagittal view showing a 10 mm tonsillar herniation. (B) MRI T2 showing dilated temporal horns representative of hydrocephalus. (C) MRI T1 with contrast axial cut showing a non-enhancing basal portion of the brain as well as a heterogeneously enhancing osseous lesion of the left frontal bone. (D) Bone window of the paranasal sinus CT scan showing the mass located on MRI to be consistent with fibrous dysplasia.

cheerful disposition and to make a living, managing her own online store.

DISCUSSION

CIM is considered a congenital condition, but new literature has described an acquired type of CIM. A case report describes a CIM that is postulated to be due to pseudotumour cerebri in

Figure 2 (A–C) Axial MRI T2 images demonstrate ventriculomegaly, compatible with hydrocephalus. (D–F) Axial non-contrast cranial CT on postoperative day 3 demonstrates improvement of ventriculomegaly. (G–I) Axial non-contrast cranial CT on postoperative day 10 shows continued improvement and near resolution of ventriculomegaly.

Table 1 Cerebrospinal Fluid Profiles		
Date	7th day of admission	12th day of admission
Qualitative	Clear, colourless	Xanthochromic, clear
Opening pressure	15	5
Closing pressure	7	2
Sugar	46%	70%
Protein (mg/dL)	69	277
Red cell count	40	270
White cell count	70	82
Neutrophils/Lymphocytes	51/47	24/76
Cultures	No organisms	No organisms
Other tests	Bactigen negative	TB PCR positive

CSF, cerebrospinal fluid; TB, tuberculosis.

a patient presenting with a prolonged history of headache.⁴ A case series of 10 children with lumboperitoneal shunts exhibited the development of CIM in seven of these patients.⁷ With these studies in mind, acquired CIM seems to stem from an increased craniospinal pressure gradient—either from an increased intracranial push or a decreased spinal canal resistance. The hydrodynamic pulsion theory, one of the theories for congenital CIM, points to progressive fetal hydrocephalus that leads to tonsillar herniation is in line with the postulated concept of acquired Chiari malformation.⁸ The patient’s tonsillar herniation may due to a congenital CIM aggravated by concomitant TB meningitis. On the other hand, she may have had an acquired form of Chiari from increased intracranial pressure from the TB meningitis.

After an exhaustive search of databases, the best evidence for the surgical management of patients with CIM stems only from observational studies—no trials have been attempted on the condition. The surgical decision-making involved in the management of CIM takes into account a myriad of factors, the most important of which are patient symptoms and findings on cranial imaging.⁶ Of patients with CIM, 6.8%–10% present with hydrocephalus.¹ For symptomatic patients with CIM and hydrocephalus, the basic principle of addressing the hydrocephalus first through ventriculo-peritoneal shunting (VPS) prior to PFD is an option that most authors recommend.^{6,9} The intracranial pressure of patients with CIM and ventricular dilation is usually normal. PFD with duraplasty has been proposed to be an effective and safe treatment for CIM with ventricular dilation provided that there is no persistent headache, vomiting and papilloedema.¹⁰ The main dilemma with our case is that the patient had all these three signs and symptoms. In addition, TB

Video 1 The patient ambulating using the clinic walker at 4 years after hospital discharge. Despite improvement in her headache and visual symptoms she remained moderately ataxic. CIM, Chiari 1 malformation.

meningitis, for which VPS is the surgical standard of treatment, was also being considered even prior to the surgery because of the prominent symptoms of nocturnal fever and cough and the high endemicity of TB in the Philippines.¹¹

The decision to proceed with PFD despite symptoms of increased intracranial pressure was based on the prominent and disabling posterior fossa symptoms and cranial imaging that showed moderate hydrocephalus with minimal transependymal effusion but with a striking tonsillar herniation of 10 mm with flattening of the medulla and compression of the cervical cord. The preponderance of radiological and clinical findings relating to the posterior fossa led to PFD.

Immediately after PFD, there was resolution of the headache and of the gaze deviation. CSF diversion was deemed unnecessary; subsequently, the patient was saved from a permanent VPS. Despite the resolution of these symptoms with the surgery, workup for TB meningitis was still pursued because of the aforementioned symptoms nocturnal fever and cough that could not be explained by CIM only.

Patient's perspective

- At first, I remember feeling really dizzy. My headache was so bad that I could no longer sleep. I don't remember much of the events leading up to the surgery. After I went home from the hospital, I could no longer do all the tasks that I used to be able to do—including my work as a data encoder. It was a very big adjustment for my family because they had to help me care for myself at first.
- But now I am better. I am able to ambulate with the use of my walker at home. I contribute to the family income by doing online merchandising. My boyfriend takes care of acquisition of goods and deliveries while I do marketing through my Facebook page.

Learning points

- The association of tuberculous (TB) meningitis and Chiari 1 malformation (CIM) reported in this case could be causative, in setting of an acquired CIM secondary to TB meningitis, or coincidental, in the setting of a congenital CIM.
- Posterior fossa decompression with suboccipital craniectomy is a viable option for the treatment of patients with CIM and TB meningitis with associated hydrocephalus.
- Despite prevailing current neurosurgical practices and recommendations the treatment of CIM should be individualised—taking into account the entire clinical picture and coexisting comorbidities.

This is the first reported case of CIM in association with TB meningitis. PFD with suboccipital craniectomy combined with an anti-TB drug treatment was effective in the treatment of patient with a symptomatic CIM and hydrocephalus and TB meningitis. The success of the procedure for CIM emphasises a point made clear by a review on the surgical management of CIM that indeed, there is no consensus among the specialists regarding the aetiology of the disease or how to approach, treat, and follow-up the CIM cases. The individualisation of treatment in patients with CIM cannot be overemphasised.⁶ In addition, the importance of history taking is highlighted by the patient's course. The lumbar tap that led to a diagnosis of TB meningitis would not have been done had it not been for the elicited symptoms that were not consistent with CIM. The results of the lumbar tap and neuroimaging corroborate for the consideration of both TB meningitis and CIM. Lastly, the case report also opens the possibility of an acquired form of CIM from TB meningitis—a condition that has not been reported elsewhere.

Contributors All authors JDBD, JECB, ATO, JGM, and PAR all contributed to the medical and surgical management of the case. JDBD was mainly responsible for the acquisition of data. All authors were involved in the conception of the paper, interpretation of data and writing the case report.

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