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Chiari I malformation with underlying pseudotumor cerebri: Poor symptom relief following posterior decompression surgery

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ABSTRACT

INTRODUCTION: ' Pseudotumor cerebri (PTC) patients exhibit clear clinical signs and symptoms of higher intracranial pressure (ICP) without ventricular enlargement or mass lesions. The clinical picture of patients with PTC can sometimes be similar to that of Chiari Malformation type I (CMI). There is some evidence that Chiari I malformation and PTC may coexist, which raises the question of whether PTC is an idiopathic disease or a complication of posterior decompression surgery—treatment of choice for Chiari I malformation.

PRESENTATION OF CASES: A retrospective review of electronic medical records of patients diagnosed with PTC at the University of Toledo Medical Center (UTMC) was performed. The objective was to determine whether PTC patients had a concurrent diagnosis of Chiari I malformation and whether the diagnosis of PTC occurred before or after posterior decompression surgery. Out of the 8 eligible patient medical records reviewed, 5 patients diagnosed with PTC had undergone posterior decompression surgery for Chiari I malformation at anywhere from several days to three years prior to being diagnosed with PTC. The diagnosis of PTC was based on temporary symptomatic relief following lumbar puncture which also showed elevated CSF opening pressures. Finally, a VP shunt was placed in each of the 5 patients to relieve the elevated intracranial pressure which resulted in the complete resolution of the patients' symptoms.

DISCUSSION: Our study focuses on patients who were diagnosed with and treated for CMI then reported back to the clinic within several days to three years complaining of symptoms of headache. Upon representing to the clinic, a CSF flow study was performed which showed normal flow of CSF. Then, these patients underwent a lumbar puncture which demonstrated an elevated opening pressure (and ICP) and a temporary relief of the headache with lumbar drainage. A VP shunt was placed for each patient to treat for PTC, and the patients' headaches were relieved.

CONCLUSION: This study suggests that the presence of Chiari I malformation in a patient conceals the symptoms of PTC which may become apparent following posterior decompression surgery. Other possibilities could be that the patients are misdiagnosed for Chiari I malformation when they are in fact suffering from PTC, or that PTC is a complication of surgery.

1. Introduction

Pseudotumor cerebri (PTC) patients exhibit clear clinical signs and symptoms of higher intracranial pressure (ICP) without ventricular enlargement or mass lesions [1]. The clinical picture of patients with PTC can be similar to that of Chiari Malformation type I (CMI) (Fig. 1) some instances. There is evidence that Chiari I Malformation and PTC may coexist in the same patient. In fact, in a small population of patients, symptoms of PTC may be confused for CMI. *Johnston* et al. found that 6% of adults diagnosed with PTC also have CMI [2]. This is eight times higher than the incidence of CMI in the general population. *Banik* et al. retrospectively deduced that up to 24% of PTC patients may demonstrate radiographic evidence of cerebellar tonsillar ectopia similar to that present in CMI patients [3]. Reasons for this overlap in symptoms between CMI and PTC have occasionally been suggested, however a definitive mechanism has yet to be affirmed.

CMI is classified as a disorder of the paraxial mesoderm with hindbrain mal-development and a small, underdeveloped posterior

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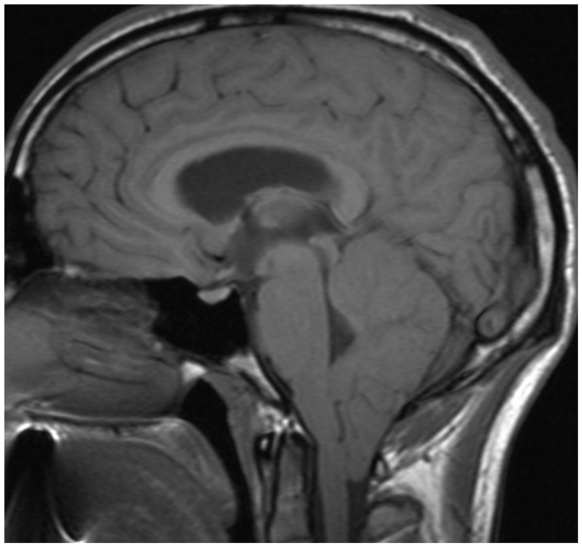

Fig. 1. Sagittal T1 weighted image demonstrates herniation of the cerebellar tonsils 2.2 cm below the foramen magnum with downward pointing.

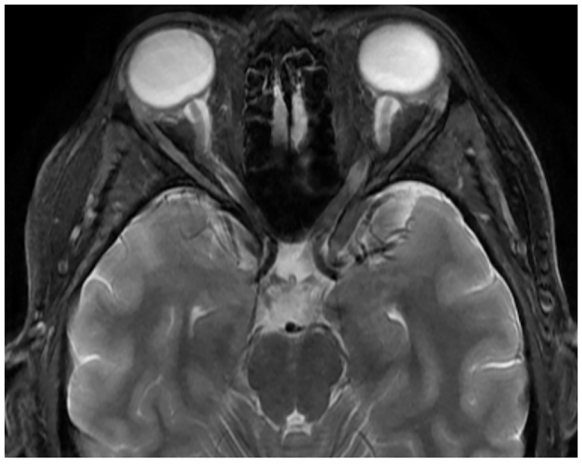

Fig. 2. Axial T2 fat saturated image demonstrates bulging of the optic disc with flattening of the posterior aspect of the globe. There is also prominence of the CSF space along the heads of the optic nerve sheath complexes. The optic nerves are tortuous in course.

fossa [3]. More specifically, CMI is a caudal descent of the cerebellar tonsils through the foramen magnum into the upper cervical spinal canal, resulting in compression of the caudal brainstem and upper cervical spinal cord as well as obstruction of normal CSF flow through the fourth ventricle, which can result in elevated ICP [3]. Radiographically, CMI is defined as inferior tonsillar displacement (ITD) of at least 5 mm below the foramen magnum, although several studies have used 3 mm as a threshold for the diagnosis of CMI [3,4].

On the other hand, PTC (Fig. 2) has evidence of higher edema and an altered CSF absorption, primarily due to abnormal intracranial compliance [2,4]. It is presumed that patients with coexistence of PTC and CMI may only have temporary relief after posterior decompression for CMI. This decompression may slightly change intracranial compliance and exacerbate the symptoms of potentially previously existing PTC. In the context of CMI with underlying PTC, it is possible for symptoms to reoccur at an average of 5.6 months following posterior decompression surgery if the PTC is not treated appropriately via a CSF shunting procedure [4]. In this study, we present a series of five patients with comorbid PTC and CMI and discuss the etiology of each disease independently and as they relate to one another.

2. Material and methods

A retrospective review of electronic medical records at the University of Toledo Medical Center was performed to identify patients who underwent a posterior decompression surgery followed by a shunting procedure. Five patients who were diagnosed and surgically treated for CMI (via posterior decompression surgery) and successively diagnosed and surgically treated for PTC (ventriculoperitoneal shunting) were identified. The PROCESS guidelines for reporting clinical case series were followed [5].

3. Results (Table 1)

3.1. Case 1

A 36-year-old female first presented to our clinic after initially being diagnosed and treated for Chiari malformation type I with 13 mm tonsillar ectopia. She had undergone a posterior decompression procedure at an outside hospital two years prior. She presented with intractable occipital headaches that radiated to her orbit. MRI was conclusive of Chiari malformation, and a posterior decompression revision was successfully completed at our institution. Several weeks later, she returned with new onset frontal headaches and diminished vision. A lumbar drain was inserted to relieve intracranial pressure. The patient's symptoms were relieved following VP shunting.

3.2. Case 2

A 45-year-old female initially presented with occipital headache, nausea, and neck pain. MRI showed Chiari malformation with 6 mm tonsillar ectopia with a secondary reduction of CSF flow around the foramen magnum. Posterior decompression with C1 laminectomy was performed. Improvement was initially noted postoperatively, but the headache and nausea returned several months later. Upon the patient's return to clinic, a CSF flow study revealed normal CSF motion at the level of the foramen magnum. A lumbar puncture showed elevated CSF opening pressure and resulted in marked improvement in the patient's headaches. The patient was then admitted for a lumbar drain trial to assess for the appropriateness of placing a VP shunt. Based on the patient's improvement during the lumbar drain trial, a decision was made to proceed with installing a VP shunt. The patient's headache symptoms were relieved with no additional complaints.

3.3. Case 3

A 25-year old female was admitted to the Neurology service for severe headache, blurry vision, papilledema, diplopia, bradycardia, and nystagmus. MRI showed CMI with a tonsillar herniation of 12 mm and ventriculomegaly. CSF flow study showed reduction of flow around the foramen magnum. Chiari posterior decompression with C1 laminectomy was performed. Two days following surgery, the headache and double vision persisted, and was accompanied by severe bradycardia. A postoperative MRI showed a radiological findings suggesting the PTC (Figs. 3–6), so a decision was made to proceed with the placement of a VP shunt. The patient's symptoms dramatically improved following shunt placement.

Table 1
Summary of findings in the five presented cases.

Case	Age	Sex	Symptoms	Tonsillar herniation	1st procedure	Returning symptoms	2nd procedure	Symptoms Resolved
1	36	F	Occipital headache radiating to eyes	13 mm	CD*	Frontal headaches Diminished vision	VP shunt	Y
2	45	F	Occipital headache Nausea Neck pain	6 mm	CD + C1 laminectomy	Headache Nausea	VP shunt	Y
3	25	F	Occipital headache Blurry vision Diplopia Bradycardia Nystagmus Papilledema	12 mm	CD	Headache Blurry vision Papilledema Bradycardia	VP shunt	Y
4	33	F	Headache Suboccipital pain Shoulder pain	2 mm	CD	Frontal headaches	VP shunt	Y
5	45	F	Severe headache Paresthesia of arm Blurry vision	5 mm	CD	Headache Blurry vision Papilledema	Lumbar drain	N

* CD: Chiari decompression.

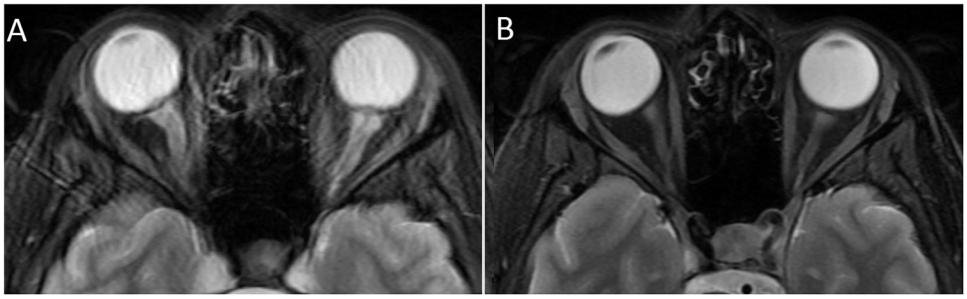

Fig. 3. A) Pre-decompression axial T2 MR image over the orbits demonstrates prominence of the optic nerve sheath complex near the heads of the optic nerves. B) Post-ventricular shunting axial T2 MR image over the orbits demonstrates a return to normal with the optic nerve sheath complex now uniform in diameter along its length with significantly less T2 hyperintense cerebral spinal fluid tracking along the optic nerves.

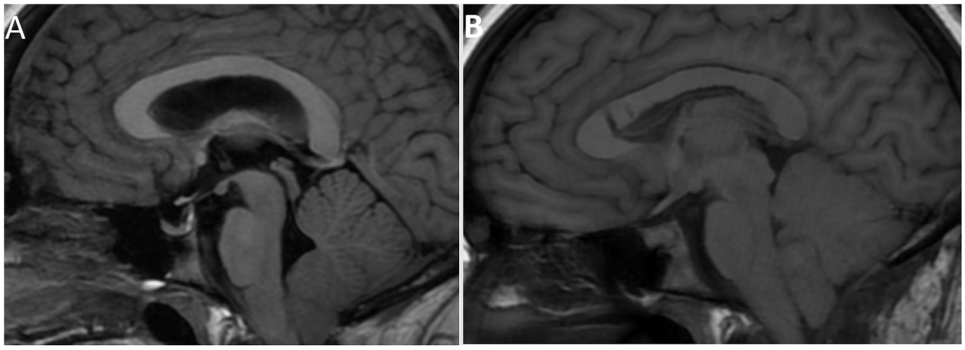

Fig. 4. A) Pre-decompression sagittal T1 MR image of the midline structures demonstrates flattening of the pituitary gland within the sella. This is atypical for a 25-year-old female as she should have a relatively full pituitary gland with a convex upper margin. Also notice that the lateral, third, and fourth ventricles are slightly prominent. Additionally, there is cerebellar tonsillar herniation measuring 12 mm. B) Post-shunting sagittal T1 MR image demonstrates interval expansion of the pituitary gland within the sella. The shunt tract can be seen in the genu of the corpus callosum. The ventricles are now significantly more decompressed as well.

3.4. Case 4

A 33-year-old female presented with headache, suboccipital pain, and shoulder pain. MRI showed CMI with approximately 2 mm cerebellar tonsillar ectopia with decreased posterior CSF flow near the foramen magnum. Chiari posterior decompression was performed, but the patient returned 3 years later complaining of a persistent frontal headache. CSF flow study demonstrated adequate flow both anteriorly and posteriorly around the foramen magnum. A lumbar puncture showed an elevated opening pressure of 32 mm Hg with headache relief upon drainage. A VP shunt was placed, and resulted in complete relief of the patient's headache symptoms.

3.5. Case 5

A 45-year-old female presented with severe headache, numbness of her arm, and blurry vision. MRI showed 5 mm inferior displacement of the cerebellar tonsils, and a CSF flow study revealed limited flow in the posterior region of the brainstem around the foramen magnum. She was diagnosed with CMI and posterior decompression was performed. Initially, the decompression resulted in symptom relief, but her headache and blurry vision returned one month later. At that time, the CSF flow study showed adequate flow anteriorly and posteriorly around the foramen magnum. Neurological examination was remarkable for papilledema.

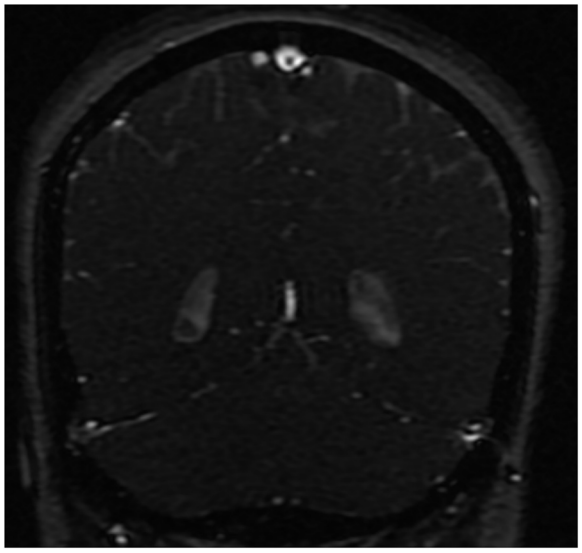

Fig. 5. Pre-decompression coronal 2D time of flight MR image showing diminished size of the transvers sinuses bilaterally. This is a finding frequently associated with elevated intracranial pressures.

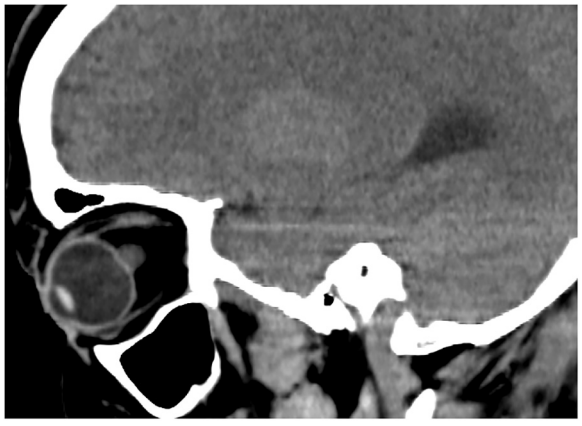

Fig. 6. Post sub-occipital decompression, pre-ventricular shunting sagittal non-contrast CT image of the orbit demonstrate flattening of the posterior globe and some bulging of the optic disc into the back of the orbit. This suggests that the elevated intracranial pressure has persisted after sub-occipital decompression for the Chiari I malformation.

A lumbar puncture showed increased CSF opening pressure. The patient was then admitted for a lumbar drain trial, which resulted in improvement in the patient's headache symptoms. Following the placement of a VP shunt, the patient's headaches and papilledema improved.

4. Discussion

4.1. Chiari I malformation

A conservative estimate suggests that approximately 1 in 1000 people or 0.1% of the population have Chiari Malformation [4]. CMI typically presents in adulthood; although, manifestations can appear in younger populations [6]. Clinically, 70–86% of CMI patients present with suboccipital headaches that radiate inferiorly to the neck and shoulders or rostrally to the retro-orbital region. These headaches are intensified by coughing, sneezing, or any maneuver that results in increased ICP [6–8]. Other symptoms including dysphagia, hoarseness, sleep apnea, facial pain or numbness, gait imbalance, and blurred or double vision may also be present [6–8]. However, *Fagan* et al. described cases where up to 30% of Chiari I patients remained asymptomatic or presented with general complaints that lead to an initial misdiagnosis of migraine, chronic fatigue syndrome, multiple sclerosis, or even PTC [9].

4.2. Pseudotumor cerebri

PTC is a disorder characterized by signs and symptoms of increased intracranial pressure in an awake and alert patient in the absence of mass-occupying lesions or obstruction of the ventricular system [10]. Idiopathic intracranial hypertension (IIH) is the primary form of PTC, whereas pseudotumor cerebri syndrome (PTCS) refers to a condition in which pseudotumor cerebri is secondary to another pathological condition [3,10]. IIH commonly affects women (8:1 female to male ratio) [11], especially women of childbearing age and those who are overweight. A study in 1988 found that IIH had an incidence of 0.9 per 100,000 in the general population. The incidence increased to 19.3 per 100,000 for women between the ages of 20 and 44 who were 20% or greater over their ideal weight [11]. The incidence of PTCS is estimated to be 1.6 per 100,000. Sources also suggest that, similar to IIH, PTCS also has an elevated incidence in women of childbearing age (11.9 per 100,000) [10].

The mechanism of PTC remains unclear, but the literature has directed its focus to the interaction between three parts of the brain, namely the vascular beds, parenchyma, and celebro-fluid space [3,4]. Pseudotumor cerebri is hypothesized to be associated with an imbalance between the production of CSF and its impaired reabsorption in the superior sagittal sinus. *McGeeney* et al. suggests that while arachnoid granulations play a crucial role in the reabsorption of CSF from the subarachnoid space, there are other, less understood mechanisms that contribute significantly to this process [10]. Potential causes of CSF pressure disruption include venous sinus thrombosis, low levels of cerebral edema, altered levels of aldosterone or arginine vasopressin, and venous hypertension [3,4,10]. Several studies demonstrated that the total water content of the brain was increased in these patients, thus associating this condition with engorged brains [4]. Brain engorgement leads to an increase in the volume of intracranial contents within a static cranium resulting in intracranial hypertension. Patients experience symptoms consistent with those of increased ICP, including headache, visual impairment secondary to papilledema, photophobia, motor weakness, sensory complications, paresthesia, pulsatile tinnitus, and fatigue[3,10,12–14].

4.3. Association between CMI and PTC

In this report, we present a total of 5 patients with coexisting CMI and PTC. All of the five patients presented were initially diagnosed with CMI and underwent suboccipital decompression with adequate radiological evidence of decompression and immediate relief of headache postoperatively. Nevertheless, headache symptoms of different characteristics compared to what was present prior to posterior decompression recurred at anywhere from several days to three years following the initial surgery. Upon the return of these patients to clinic for a post-operative follow-up appointment, they continued to complain of a moderate headache that was characterized as dull in nature. The patients asserted that the headache was different in characteristic than what they had experienced prior to decompression surgery. For this subset of patients, a CSF flow study was ordered to evaluate the flow of CSF through the ventricular system and to possibly correlate the results with the patients' clinical symptomatology. CSF flow studies revealed improved flow following posterior decompthe two clinically. CMI is hypothesized to be the result of a decreased proportion of the cranial vault to the intracranial contents. As the contents of the cranial vault expand

during development of the central nervous system, the cerebellar tonsils herniate below the foramen magnum, which may be asymptomatic or cause headache and neck pain. A similar presentation is possible in patients with engorged brains, like what is seen in PTC. Retention of CSF within the subarachnoid space in PTC patients leads to an increase in cranial vault contents due to brain engorgement. The expansion of CSF volume within the cranial vault may result in cerebellar ectopia, often leading to a similar patient presentation when compared to CMI. It is our opinion that the lack of distinction between PTC and CMI, combined with ill-defined diagnostic guidelines has contributed to the lack of understanding regarding these conditions.

4.4. Are we operating on the wrong population?

Following decompression surgery it is estimated that 50% of patients experience complete alleviation of symptoms, 20–30% improve significantly, and 20% do not improve or have worsening symptoms [4]. According to the literature, female patients who are obese, patients treated with tetracycline, or patients who present with atypical CMI symptoms such as blurred or double vision, papilledema, or tinnitus with normal or small ventricles should be considered for PTC-associated CMI and monitored for recurrence of symptoms post-decompression [10,11,16].

Before subjecting patients who are suspected of having PTC to invasive posterior decompression surgery, other measures could be utilized to reduce the risk of symptom recurrence and revision surgery. Measuring CSF pressure before surgery could be used as a marker for diagnosis of underlying PTC. In the event of poor symptomatic relief following posterior decompression surgery, it is important to determine the etiology of the underlying pathology. In cases where PTC is unlikely to be the cause of poor symptomatic relief following surgery, one must consider other potential causes such as scarring of the dura or intradural tethering (which may lead to hydrocephalus due to obstruction of the outflow of CSF). Cases of hydrocephalus as a result of dural scarring or intradural tethering should be considered prior to performing an LP due to the risk herniation, particularly in the pediatric population [17].

5. Conclusion

The increasing prevalence of PTCS as well as the reoccurrence of symptoms following decompression surgery for CMI raise suspicion that perhaps a coexistence between the two diseases is more common than previously believed. Currently, the diagnosis of CMI is dependent on the extent of tonsillar herniation into the foramen magnum. In patients with borderline tonsillar herniation, great caution must be taken prior to labeling them with the diagnosis of CMI. Increased intracranial pressures in PTC can potentially lead to a similar presentation as CMI, as demonstrated by the relief of symptoms following the placement of a VP shunt in our subset of patients. Further research is crucial for generating a standardized set of data to help medical teams determine a patient's probability for coexistence of PTC and CMI. If standardization were possible it would lend significant credibility to the statistics relating PTC and CMI. Without a stronger understanding of the underlying causes of these disorders, much of the relationship between PTC and CMI will likely remain obscured. This case series underscores the importance of obtaining proper history and imaging in cases with clinical symptoms that could be suggestive of either PTC or CMI.

Conflicts of interest

None.

Funding

None.

Ethical approval

We have registered our research manuscript at Research Registry with the Unique Identifying Number: researchregistry2475.

Consent

As per the Elsevier policy on the use of images or personal information of patients or other individuals: "Formal consents are not required for the use of entirely anonymized images from which the individual cannot be identified."

After numerous attempts, we were unable to reach the patients and obtain consent. Hence, all patient identifiers (including age, gender, etc.) have been removed and all images have been entirely anonymized.

Author contribution

All the authors were involved in the drafting, review and approval of the paper for publication.

Guarantor

Daniel Gaudin.

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