



## Chiari Malformation and Hydrocephalus Masking Neurocysticercosis

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### Key words

- Chiari malformation
- Hydrocephalus
- Neurocysticercosis
- Subarachnoid cysts

### Abbreviations and Acronyms

CP: Cerebellopontine  
MRI: Magnetic resonance imaging  
VP: Ventriculoperitoneal

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### BACKGROUND

Neurocysticercosis is the leading neuro-parasitic infection in the United States, affecting approximately 2000 individuals every year.<sup>1</sup> The disease is caused by the ingestion of eggs from the tapeworm *Taenia solium* that are excreted in the stool of people with taeniasis. Neurocysticercosis symptoms range from headaches to seizures but can also present with symptoms of hydrocephalus, which can be caused by chronic arachnoiditis, meningeal fibrosis, or intraventricular cysts.<sup>2</sup> Here we report a unique presentation of neurocysticercosis following surgical intervention for a Chiari I malformation.

### CASE DESCRIPTION

A 55-year-old Hispanic male presented to the clinic complaining of balance issues, diplopia, visual field deficits, and ongoing headaches that had persisted for about a year. Radiographic imaging demonstrated the presence of an Arnold Chiari I malformation (Figure 1). He was taken to the operating room and underwent posterior fossa decompression via a suboccipital craniectomy with C1 laminectomy and

■ **BACKGROUND:** Various diagnostic characteristics associated with neurocysticercosis have been well studied; however, their potential to be implicated in other differential diagnoses has not been well demonstrated.

■ **CASE DESCRIPTION:** We report the case of a 55-year-old Hispanic man who underwent a Chiari decompression surgery, which was complicated with hydrocephalus. Despite a ventriculoperitoneal shunt placement, he continued to have headaches and was soon found to have several skull base subarachnoid lesions, which were later diagnosed as the sequelae of an active neurocysticercosis infection.

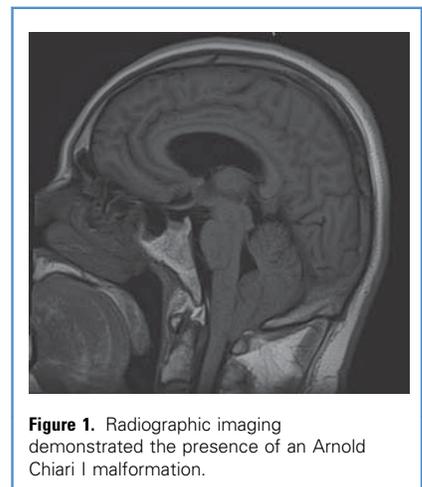
■ **CONCLUSION:** This case report highlights the importance of overlapping symptoms between diseases in a short temporal context.

duraplasty. He had an uneventful hospital course and was discharged home after 3 days. The patient did well for several months but then presented with recurrent headaches about 5 months after his surgery. Brain imaging demonstrated a large 7.4 × 9.6 cm pseudomeningocele within the surgical site and extending into the posterior tissues of the neck (Figure 2A and B).

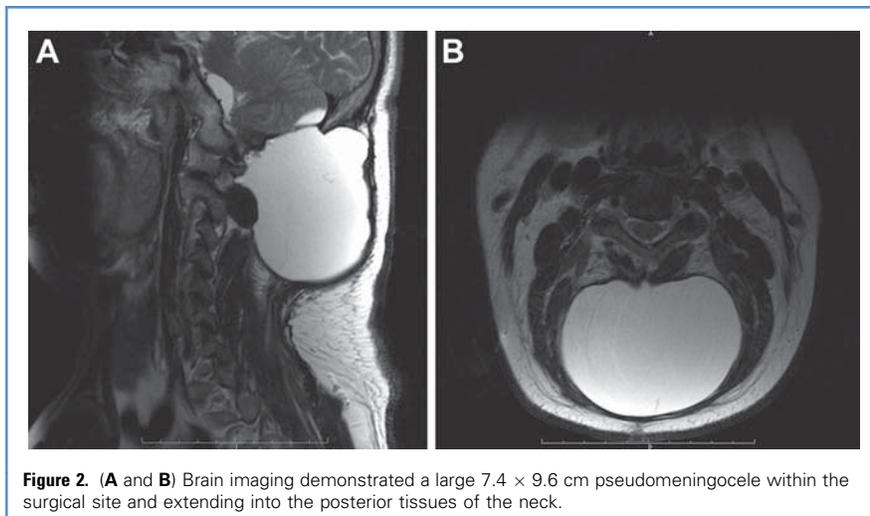
The patient was taken to the operating room for exploration and revision of the suboccipital pseudomeningocele and placement of a lumbar drain. At the time of surgery, a small pinhole was visualized at the edge of the duraplasty graft and revised with a 6-0 nonabsorbable polypropylene suture, fat graft, and dural sealant. He was hospitalized with continuous lumbar drainage and discharged home on the eighth day reporting an improvement in his headaches. Approximately 4 weeks later, the patient returned to the emergency department complaining of worsening bitemporal headaches, nausea, and vomiting. A magnetic resonance imaging (MRI) scan indicated that he had developed hydrocephalus with reaccumulation of fluid in the subarachnoid space consistent with a pseudomeningocele measuring 5.6 × 4.8 cm (Figure 3A and B). Due to the recurring nature of hydrocephalus, the patient was taken to the operating room and underwent a right frontal ventriculoperitoneal (VP) shunt placement (Figure 4). He did well and was discharged home the next day.

After approximately 8 months, the patient was seen in the emergency department again for a fever, headache, balance problems, and myalgias. MRI of the brain showed a large arachnoid cyst in the left cerebellopontine (CP) angle measuring 3.1 × 1.7 cm and another arachnoid cyst in the right ambient cistern measuring 2.8 × 1.7 cm (Figure 5A and B). The larger cyst in the CP angle was causing compression on the brainstem. The VP shunt valve was readjusted in the clinic over 2 separate visits in an attempt to increase the flow rate with close monitoring of the skull-based cysts for any size changes.

Five months later, the patient continued to complain of headaches, dizziness, loss



**Figure 1.** Radiographic imaging demonstrated the presence of an Arnold Chiari I malformation.

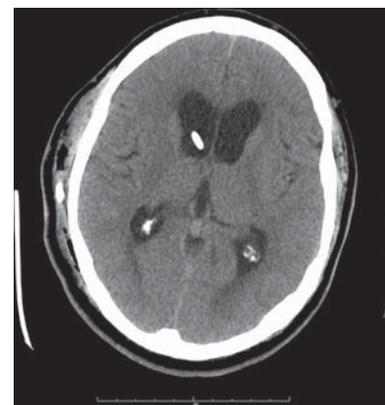


**Figure 2.** (A and B) Brain imaging demonstrated a large 7.4 × 9.6 cm pseudomeningocele within the surgical site and extending into the posterior tissues of the neck.

of lateral vision, and left sided face and tongue numbness. The cyst in the left CP angle had increased to 3.8 × 2.6 cm although the cyst in the right ambient cistern decreased in size to 1.8 × 1.4 cm. The decision was made to proceed with surgical exploration and cyst fenestration because they were becoming more symptomatic and not responding to his VP shunt valve adjustments. The patient subsequently underwent a retrosigmoid craniotomy for fenestration of the left CP angle cyst. During his surgery, a small firm lesion was unexpectedly noted within

the cyst fluid and sent to Pathology. **Figure 5C** His postoperative MRI, demonstrating an improvement in his compressive cyst.

Histologic analysis (**Figure 6A–C**) of the firm lesion, measuring 1.4 × 1.1 cm, showed the classic 3-layer distinctive wall of pleomorphic neurocysticercosis with microtriches/microvilli, an outer eosinophilic cuticle layer, a cellular layer with uniform cellular dark nuclei, and an inner reticular layer containing loose fibrils. In addition, intense lymphoplasmacytic inflammation and dystrophic calcifications



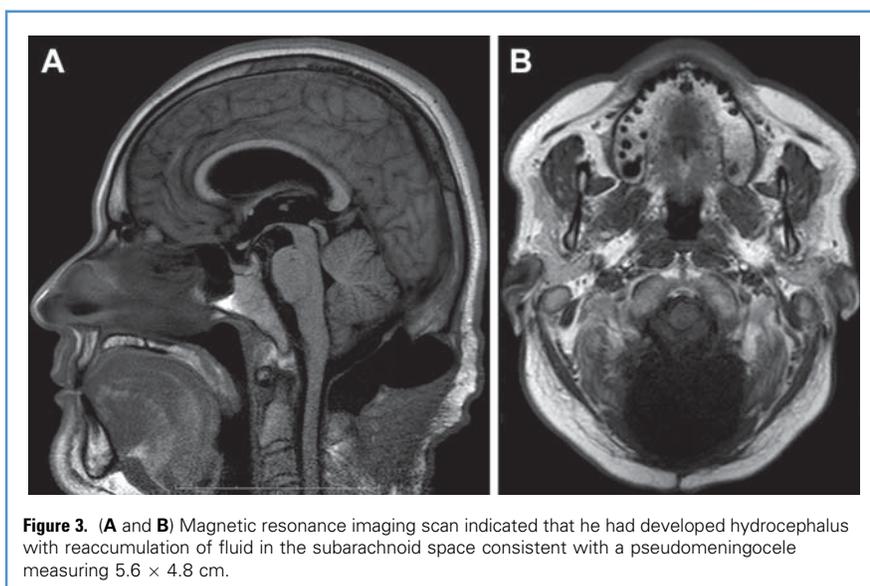
**Figure 4.** Due to the recurring nature of hydrocephalus, the patient was taken to the operating room and underwent a right frontal ventriculoperitoneal shunt placement.

but no scolex were observed. Pathologic and histologic evidence was sufficient to confirm the diagnosis of neurocysticercosis.

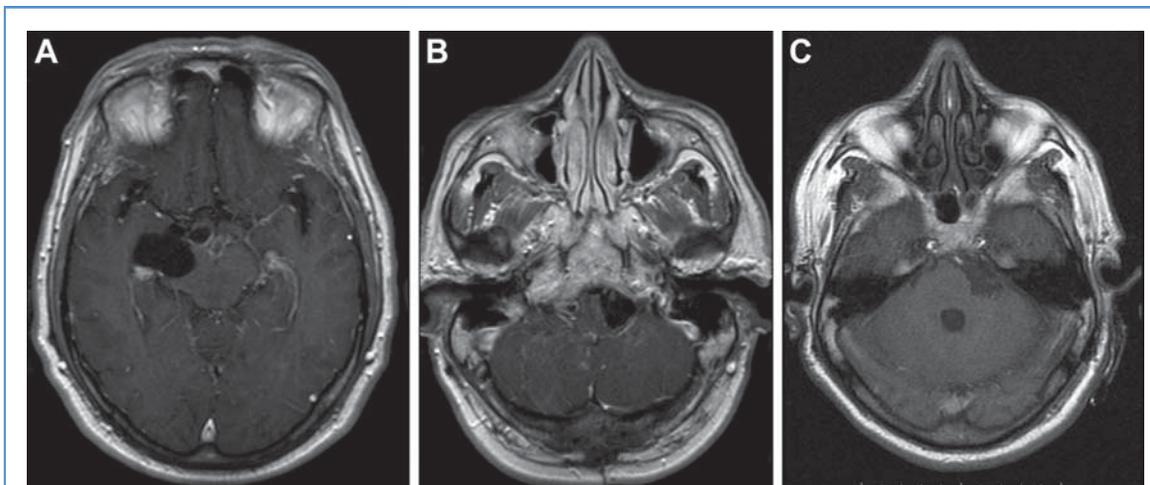
The patient was seen by an infectious disease expert and placed on close observation for further possible infection and symptoms related to neurocysticercosis. In this case, anthelmintic drugs were not prescribed because it was felt the patient had undergone definitive surgical treatment of his neurocysticercosis. The patient recovered without any postoperative complications or remaining symptoms other than slight balance impairment. His follow-up imaging at 4 months (**Figure 7A and B**) has demonstrated complete resolution of the arachnoidal cyst in the left CP angle and near-complete resolution of the right ambient cistern cyst measuring 1.2 × 0.4 cm.

## DISCUSSION

A host of symptomatic or asymptomatic signs can occur depending on the size, location (parenchymal or extraparenchymal), and number of cysts in patients infected with neurocysticercosis. An extraparenchymal racemose form of neurocysticercosis is observed to compose up to 54% of cases with cysts located in the cisternal/subarachnoid spaces, spinal cord, or ocular bulb.<sup>3</sup> Seizures are the most common clinical presentation of this form, but hydrocephalus may occur due to



**Figure 3.** (A and B) Magnetic resonance imaging scan indicated that he had developed hydrocephalus with reaccumulation of fluid in the subarachnoid space consistent with a pseudomeningocele measuring 5.6 × 4.8 cm.



**Figure 5.** (A–C) Magnetic resonance imaging of the brain showed a large arachnoid cyst in the left cerebellopontine angle measuring  $3.1 \times 1.7$  cm and another arachnoid cyst in the right ambient cistern measuring  $2.8 \times 1.7$  cm.

mechanical obstruction or inflammatory reaction and has been associated with higher mortality.<sup>4</sup>

Although signs of hydrocephalus can be easily recognized on imaging studies, uninfamed extraparenchymal cysticerci may be difficult to identify on computed tomography or MRI scans due to the thin membrane and the cyst being isodense to cerebrospinal fluid.<sup>5</sup> This diagnostic aspect can often be overlooked when diagnosing neurocysticercosis because hydrocephalus without accompanying radiologic evidence of cysts is typically attributed to and treated as something else. To make things even more complicated, arachnoid cysts may develop as a complication of overdrainage.<sup>6</sup> This had happened in our case when the patient developed

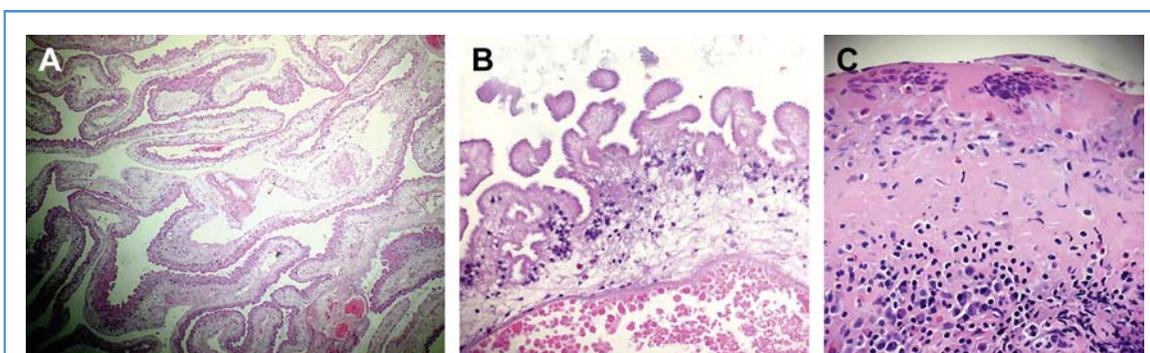
hydrocephalus, which is also a common comorbidity of a Chiari malformation and later develop arachnoid cysts.

Our case is unique in that the patient initially presented with a Chiari malformation without any clinical evidence of a neurocysticercosis infection. It was only after hydrocephalus was treated with a VP shunt that he developed enlarging subarachnoid cysts, which were later diagnosed to be infectious in etiology; this was also associated with symptoms of fever and myalgias. There was another similar case reported in the literature, but in that case a patient developed an acquired Chiari malformation after VP shunt placement for hydrocephalus associated with neurocysticercosis.<sup>7</sup> It is more likely in our case that Chiari malformation was congenital and did not develop due

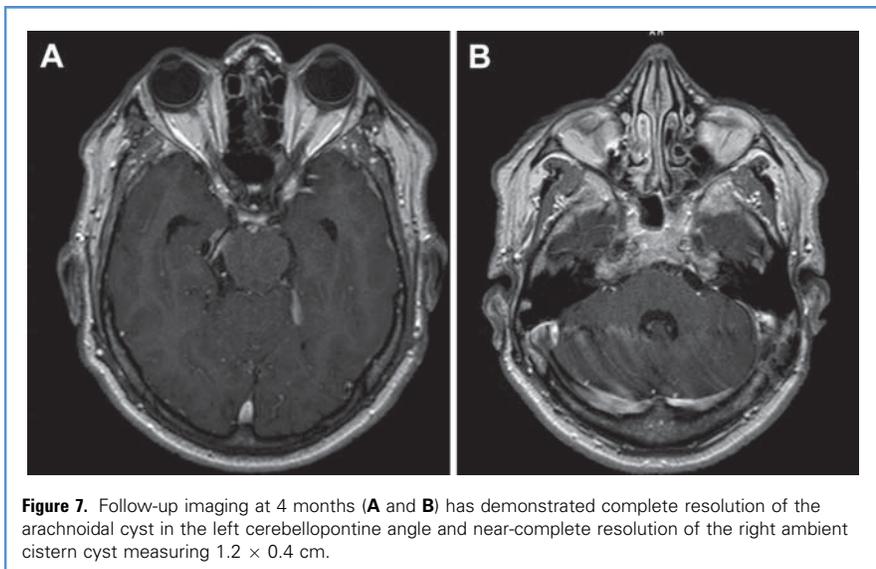
to elevated intracranial pressure from an obstructive process secondary to a neurocysticercosis infection. However, given the close temporal proximity of the Chiari symptoms and the observed cysts, it is also possible that intracranial hypertension from the cysts caused an acquired Chiari malformation or worsened the congenital form and it became symptomatic. Future considerations regarding recurring hydrocephalus should be met with a high level of scrutiny for their possible association with both morphologic and pathophysiologic problems.

## CONCLUSIONS

Management of neurocysticercosis can be contentious due to the wide variety of



**Figure 6.** Histologic analysis (A–C) of the firm lesion, measuring  $1.4 \times 1.1$  cm, showed the classic 3-layer distinctive wall of pleomorphic neurocysticercosis with microtriches/microvilli, an outer eosinophilic cuticle layer, a cellular layer with uniform cellular dark nuclei, and an inner reticular layer containing loose fibrils.



**Figure 7.** Follow-up imaging at 4 months (**A** and **B**) has demonstrated complete resolution of the arachnoidal cyst in the left cerebellopontine angle and near-complete resolution of the right ambient cistern cyst measuring 1.2 × 0.4 cm.

symptoms depending on the location, developmental stage, and number of cysts when presenting. With cases of neurocysticercosis increasing in the United States, it is essential that the various symptoms and accompanying radiographic results are recognized. Our case report is the first in the literature to illustrate a unique manifestation of neurocysticercosis. The potential interactions

among subarachnoid cysts, Chiari malformations, and recurring hydrocephalus are important considerations to study when making differential diagnoses on neurocysticercosis.

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