



Case Report

# Multiple etiologies of secondary headaches associated with arachnoid cyst, cerebrospinal fluid hypovolemia, and nontraumatic chronic subdural hematoma in an adolescent: A case report

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## ABSTRACT

**Background:** Diagnosing the cause of headaches can be challenging. Even if intracranial lesions are found in a patient, careful assessment is essential for diagnosis, and treatment strategies will differ for each etiology.

**Case Description:** A 16-year-old boy presented with sudden-onset headache which had lasted for 2 days. His headache was aggravated in the orthostatic position. The patient denied recent head trauma. He had been diagnosed with an arachnoid cyst (AC) in his right middle cranial fossa. Computed tomography (CT) revealed bilateral subdural effusions and slit-like lateral ventricles with no significant changes to the AC. After intravenous hydration followed by 2 days bed rest, his symptoms abated. He was diagnosed as having suffered spontaneous cerebrospinal fluid (CSF) hypovolemia. One month later, the patient experienced recurrent gradual onset headache and vomiting. CT revealed chronic right side subdural hematoma (SDH) with intracystic hemorrhage, which resulted in the elevation of intracranial pressure. An urgent hematoma evacuation was performed. He became symptom-free immediately after surgery. Postoperative follow-up CT showed no change in the AC and no recurrence of SDH. The lateral ventricles and subdural space were normal in size.

**Conclusion:** We report a case presenting multiple types of secondary headaches, which were caused by intracranial hypotension or hypertension, with different etiologies. These were spontaneous CSF hypovolemia, nontraumatic intracystic hemorrhage form of AC, and nontraumatic chronic SDH. Although lesions seen at the time of initial diagnosis did not need surgical treatment, careful observation and repetitive imaging assessments might be useful for discovering unsuspected additional etiologies requiring surgical intervention.

**Keywords:** Cerebrospinal fluid hypotension, Children, Clinical presentation, Diagnosis, International Classification of Headache Disorders 3<sup>rd</sup>

## INTRODUCTION

Diagnosing the cause of headaches can be challenging. Even if intracranial lesions are found in a patient, careful assessment is essential for diagnosis, and treatment strategies will differ for each etiology. Here, we report a patient with multiple etiologies of secondary headaches associated with multiple intracranial conditions.

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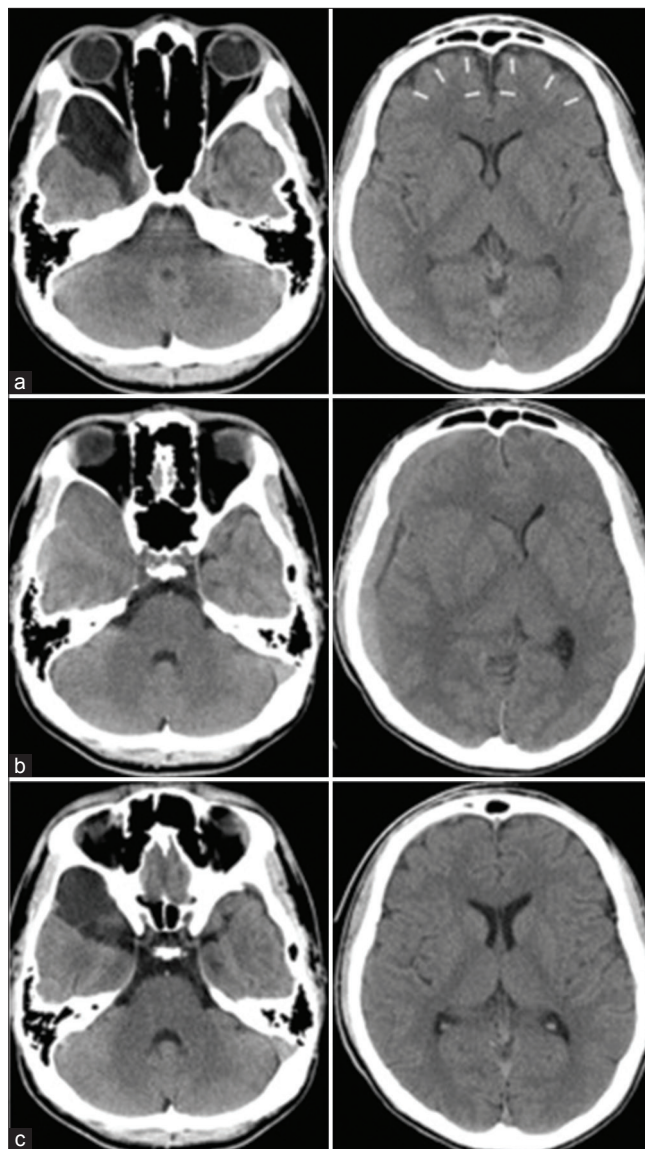
## CASE DESCRIPTION

A 16-year-old boy presented at our hospital with sudden-onset general headache and vomiting which had lasted for 2 days. His headache was aggravated in the orthostatic position and relieved in the recumbent position. Neurological examination was uninformative. The patient stated that he had had no recent head trauma. He had been diagnosed with an arachnoid cyst (AC) in his right middle cranial fossa at 6 years of age, without any comorbidities. Computed tomography (CT) revealed bilateral subdural effusions and slit-like lateral ventricles with no significant changes to the AC [Figure 1a]. In addition to the CT findings, MRI showed obliteration of the prepontine cistern with flattening of the pons against the clivus and descent of the cerebellar tonsils, suggesting brain sagging. After intravenous hydration followed by 2 days' bed rest, his symptoms abated. Based on the clinical course, he was retrospectively diagnosed as having suffered spontaneous cerebrospinal fluid (CSF) hypovolemia. One month later, the patient again experienced recurrent headache and vomiting. This time, the headache was significantly on the right and had been gradual in onset. The neurological examination was still unremarkable but repeat CT after another month revealed a hyperdense right subdural hematoma (SDH) and a hyperdense change in the AC with obvious compression of the right lateral ventricle and evident midline shift to the left [Figure 1b]. MRI showed the presence of the outer membrane of the AC between the AC and the SDH. The size of the AC was unchanged. His blood count and coagulation markers were normal. He was diagnosed as having chronic right side SDH with intracystic hemorrhage, which resulted in the elevation of intracranial pressure (ICP). An urgent hematoma evacuation was performed under local anesthesia by burr-hole irrigation surgery. Postoperative CT confirmed complete evacuation of the SDH and the patient became symptom-free immediately after surgery. Postoperative follow-up CT 3 months after surgery showed no change in the volume of the AC and no recurrence of SDH. The lateral ventricles and subdural space were again normal in size [Figure 1c].

## DISCUSSION

### Diagnoses of headache

In general, spontaneous CSF hypovolemia is diagnosed according to the criteria of the International Classification of Headache Disorders 3<sup>rd</sup> edition (ICHD-3).<sup>[6]</sup> The diagnostic criteria are as follows: (a) headache fulfilling criteria for 7.2 headache attributed to low CSF pressure and criterion C; (b) absence of a procedure or trauma known to be able to cause CSF leakage; (c) headache has developed in temporal relation to occurrence of low CSF pressure or CSF leakage, or has led to its discovery; and (d) not better accounted for by another ICHD-3 diagnosis. Additional criteria for headache



**Figure 1:** Brain computed tomography (CT) of the patient. CT obtained 2 days after the onset of the first headache showing bilateral subdural effusions (arrows) and slit-like lateral ventricles with an arachnoid cyst in the right temporal fossa (a). CT obtained at recurrence of headache showing a hyperdense right subdural hematoma and a hyperdense change in the arachnoid cyst with obvious compression of the right lateral ventricle and evident midline shift to the left (b). CT obtained 3 months after surgery showing no change in the volume of the arachnoid cyst and no recurrence of subdural hematoma with the lateral ventricles and subdural space normal in size (c).

attributed to low CSF pressure coded as 7.2 in the ICHD-3 includes either or both of the following: (1) Low CSF pressure and (2) evidence of CSF leakage on imaging. The presentation of the initial symptoms and imaging in our case fulfilled the criteria for spontaneous CSF hypovolemia.

The exacerbated right-sided headache and vomiting in the patient were accompanied by right AC hemorrhage

and right chronic SDH (CSDH). With the brain images showing midline shift of the brain and the symptoms having disappeared following hematoma evacuation, ICP elevation was considered a possible cause of headache and vomiting. This headache was coded as 6.2 headache attributed to nontraumatic intracranial hemorrhage in ICHD-3.

### **Etiological relationships between AC, CSF hypovolemia, and nontraumatic CSDH**

#### ***CSF hypovolemia and nontraumatic CSDH***

Numerous reports have shown that CSDH is one of the comorbidities of CSF hypovolemia.<sup>[4,13]</sup> It is currently deemed likely that the formation of CSDH is caused by tearing of the bridging veins caused by downward displacement of the brain as a result of reduced hydrostatic force.<sup>[4]</sup>

#### ***CSF hypovolemia and AC***

In the published literature, we found only two cases of CSF hypovolemia thought to be related to AC.<sup>[5,9]</sup> Han *et al.* emphasized that the structural defects of the meninges might cause both the AC and dural defects. The hypothesis was that these two conditions did not occur simultaneously coincidentally but that they might be related to each other.<sup>[5]</sup>

#### ***Nontraumatic CSDH and AC***

From the literature, it may be postulated that nontraumatic CSDHs in young patients could be associated with spontaneous AC-related hemorrhage.<sup>[1,2,7,8,10,11,14,15]</sup> The etiologies of hemorrhage in subdural and/or intracystic spaces include: (1) rupture of bridging veins by tearing of the outer wall of the AC;<sup>[7,14]</sup> (2) rupture of unsupported blood vessels around the cyst wall;<sup>[7,8]</sup> and (3) rupture of leptomeningeal vessels at the base of the cyst.<sup>[7,8]</sup> To the best of our knowledge, there are no reported cases of nontraumatic SDH followed by AC hemorrhage.

#### ***Treatments***

Spontaneous CSF hypovolemia is most commonly treated by bed rest and hydration, but is successful in only 28% of patients.<sup>[3,4]</sup> If targeted treatment is needed, in the case of treatment failure, spinal investigation is required.<sup>[3]</sup> However, identifying the location of the CSF leakage by radiological investigations is challenging in a high proportion of patients who have a clinical history convincingly indicating CSF hypovolemia.<sup>[3]</sup> Therefore, it would be reasonable to suggest further investigation as an elective option. If conservative treatment is ineffective and when the site of the CSF leakage can be determined, application of an epidural blood patch (EBP) can be attempted.<sup>[3,4]</sup>

For CSDH with CSF hypovolemia, if CSF leakage was diagnosed and persisted despite conservative treatment, hematoma evacuation and EBP might be a possibility<sup>[3,4]</sup> as described above. Otherwise, CSDH evacuation alone might be the best option.<sup>[8]</sup>

Several authors have pointed out that AC hemorrhage might need surgery to prevent recurrent hemorrhage.<sup>[8]</sup> However, the optimal treatment strategy for CSDH with AC hemorrhage is still controversial.<sup>[12]</sup> Reported successful initial treatments are CSDH drainage by burr-hole surgery,<sup>[10,12]</sup> CSDH evacuation and AC membrane removal,<sup>[12,15]</sup> and CSDH evacuation and AC membrane fenestration.<sup>[4,12,15]</sup>

#### ***Diagnoses, etiologies, and treatments in our patient***

The AC existed before CSF hypovolemia in our case. Because the symptoms caused by CSF hypovolemia were resolved without recurrence on conservative treatment, spinal investigation was not performed. Fragile dural structures might have been present elsewhere.

Concurrent hemorrhage in the AC and the occurrence of CSDH were identified after recovery from CSF hypovolemia in our case. It was not clear which hemorrhage had preceded the other. One hypothesis would be that the AC hemorrhage occurred before the appearance of CSDH. In that case, the change in CSF pressure, hypotension, and restoration would have caused hemorrhage in the fragile structures of the AC. The AC outer membrane might have ruptured and leaked blood into the subdural space, resulting in CSDH after a few weeks. A second hypothesis is that the CSDH occurred before AC bleeding. In that case, CSF hypovolemia might have caused CSDH as a previously described common comorbidity. The CSDH was identified only in the right convexity with an atrophic right temporal lobe because of coexisting AC. Elevated subdural pressure might then have caused rupture of the AC. If this hypothesis is true, this is the first case where CSDH caused rupture of the adjacent AC.

Because elevated ICP was caused by CSDH, our case was treated with minimally invasive burr-hole hematoma evacuation surgery alone. Fortunately, no recurrence of hemorrhagic events or any comorbidities occurred after this palliative surgery, as described in earlier literature.

Our study has some limitations: (1) we could not explore the cause of CSF leakage, because the CSF hypovolemia disappeared on conservative treatment and (2) we have not operated on the AC because AC hemorrhage did not recur. Therefore, the cause of CSF hypovolemia and the exact location of the AC rupture have not been identified. Our study has not proved any mechanisms of CSF hypovolemia, CSDH, or AC formation. Because AC hemorrhage, CSDH, and/or CSF hypovolemia could recur at any time in the future, careful observation of this patient is clearly necessary.

## CONCLUSION

We report a case presenting multiple types of secondary headaches over a short period of time, which were caused by intracranial hypotension or hypertension, with different etiologies. These were spontaneous CSF hypovolemia, nontraumatic intracystic hemorrhage form of AC, and nontraumatic CSDH. As this novel case highlighted, multiple etiologies can coexist, explaining different types of headaches in the same patient. Although lesions seen at the time of initial diagnosis did not need surgical treatment, careful observation of such patients for symptoms, as well as carrying out repetitive imaging assessments, might be useful for discovering unsuspected additional etiologies requiring surgical intervention at the optimal time point. Furthermore, these assessments might be helpful for treating any recurrent conditions in this patient or similar conditions in other patients, in the future.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

## REFERENCES

- Balestrino A, Piatelli G, Consales A, Cama A, Rossi A, Pacetti M, *et al.* Spontaneous rupture of middle fossa arachnoid cysts: Surgical series from a single center pediatric hospital and literature review. *Childs Nerv Syst* 2020;36:2789-99.
- Bora A, Yokuş A, Batur A, Bulut MD, Yavuz A, Gülşen İ, *et al.* Spontaneous rupture of the middle fossa arachnoid cyst into the subdural space: Case report. *Pol J Radiol* 2015;80:324-7.
- D'Antona L, Merchan MA, Vassiliou A, Watkins LD, Davagnanam I, Toma AK, *et al.* Clinical presentation, investigation findings, and treatment outcomes of spontaneous intracranial hypotension syndrome: A systematic review and meta-analysis. *JAMA Neurol* 2021;78:329-37.
- Ferrante E, Savino A, Sances G, Nappi G. Spontaneous intracranial hypotension syndrome: Report of twelve cases. *Headache* 2004;44:615-22.
- Han S, Choi SW, Park BS, Lim JW, Kim SH, Youm JY. Cervical cerebrospinal fluid leakage concomitant with a thoracic spinal intradural arachnoid cyst. *Korean J Neurotrauma* 2019;15:214-20.
- Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3<sup>rd</sup> edition. *Cephalalgia* 2018;38:1-211.
- Hong JC, Kim MS, Chang CH, Kim SH. Arachnoid cyst with spontaneous intracystic hemorrhage and chronic subdural hematoma. *J Korean Neurosurg Soc* 2008;43:54-6.
- Hou K, Li CG, Zhang Y, Zhu BX. The surgical treatment of three young chronic subdural hematoma patients with different causes. *J Korean Neurosurg Soc* 2014;55:218-21.
- Jones WD, Kulkarni M, Ravishankar K, Borgstein R, Dupont P. Intracranial hypotension secondary to spinal arachnoid cyst rupture presenting with acute severe headache: A case report. *J Med Case Rep* 2010;4:406.
- Kieu HD, Le TD, Tran TQ. Arachnoid cyst with spontaneous intracystic hemorrhage and associated chronic subdural hematoma: A case report. *Int J Surg Case Rep* 2021;81:105789.
- Mori K, Yamamoto T, Horinaka N, Maeda M. Arachnoid cyst is a risk factor for chronic subdural hematoma in juveniles: Twelve cases of chronic subdural hematoma associated with arachnoid cyst. *J Neurotrauma* 2002;19:1017-27.
- Müslüman AM, Özöner B, Kirçelli A, Can SM, Yılmaz A, Kaldirimoğlu A, *et al.* Chronic subdural hematoma associated with arachnoid cyst of the middle fossa: Surgical treatment and mid-term results in fifteen patients. *Turk Neurosurg* 2018; 28:776-782.
- Schievink WI, Maya MM, Moser FG, Tourje J. Spectrum of subdural fluid collections in spontaneous intracranial hypotension. *J Neurosurg* 2005;103:608-13.
- Szczygielski J, München D, Ketter R, Rákász L, Schulz-Schaeffer W, Oertel J. Head injury without head blow? A rare case of subdural hematoma associated with minute arachnoid cyst in a teenage skater. *J Neurol Surg A Cent Eur Neurosurg* 2021;82:604-10.
- Wu X, Li G, Zhao J, Zhu X, Zhang Y, Hou K. Arachnoid cyst-associated chronic subdural hematoma: Report of 14 cases and a systematic literature review. *World Neurosurg* 2018;109:e118-30.

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