




Aseptic Meningitis Due to Spontaneous Rupture of a Multicystic Craniopharyngioma with an Ommaya Catheter: A Case Report

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Abstract

Craniopharyngiomas (CPs) are benign tumors that are believed to arise from embryonic remnants of the Rathke pouch epithelium. Herein, we report a case of aseptic meningitis due to spontaneous rupture of multicystic CP, which contained an Ommaya catheter. A 19-year-old boy was admitted to the hospital with a 4-day history of acute severe headache after strenuous physical exercise followed by altered sensorium, fever, and neck stiffness. Cerebrospinal fluid (CSF) analysis revealed marked pleocytosis and elevated protein levels. CSF culture was otherwise negative. Cyst reduction on subsequent imaging confirmed the diagnosis. The patient received intravenous steroid therapy and was discharged asymptomatic. This is a rare evolution of a multicystic CP, which was previously treated with intracystic therapy and had an Ommaya catheter. Clinicians should be aware of spontaneous CP rupture and look actively for the occurrence of cholesterol crystals or elevated CSF levels of cholesterol as well as prompt follow-up imaging.

Keywords

- ▶ aseptic meningitis
- ▶ craniopharyngioma
- ▶ spontaneous rupture
- ▶ Ommaya catheter

Key Messages

Facing a patient with a cystic craniopharyngioma with primary or recurrent meningeal signs, clinicians should be aware of spontaneous craniopharyngioma rupture and look actively for the occurrence of elevated cerebrospinal fluid levels of cholesterol, as well as prompt follow-up imaging of the sellar region.

Introduction

Craniopharyngiomas (CPs) are histologically benign tumors, which are believed to arise from embryonic remnants of the Rathke pouch epithelium.¹ CPs are relatively rare tumors

with the overall incidence of 0.13 per 100,000 person years² corresponding to 11% of all pediatric central nervous system tumors.³ CPs have a bimodal age distribution occurring from 5 to 14 years and then a second peak between 50 and 74 years.² Headaches, visual disturbances, hypopituitarism, and cognitive decline comprise major observations at clinical presentation regardless of age.^{4–6} Typical imaging reveals a sellar and parasellar solid and/or cystic lesion with variable degree of calcification and a lobular shape of approximately 20 to 40 mm.^{6,7}

Spontaneous rupture of a cystic CP is an extremely rare event, which was first described by Worster-Drought et al in 1927 in a 19-year-old girl with recurrent episodes of aseptic

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meningitis.⁸ To date, less than 35 cases of spontaneous CP rupture have been reported in the English literature.^{5,8-31} Herein, we present a report of an adolescent with a multicystic CP who experienced an episode of aseptic meningitis due to spontaneous rupture of the ventricular cyst, which contained an Ommaya catheter.

Case Report

A 19-year-old boy who was affected by a multicystic sellar and suprasellar CP was admitted to our hospital with a 4-day history of acute severe headache after strenuous physical exercise. Headache was followed by altered sensorium, fever, and neck stiffness. The patient initially presented at the age of 15 years and 4 months, when he came to emergency comatose after a long history of growth defect and headache. Cranial computed tomography (CT) revealed a giant multicystic mass with gross calcifications (►Fig. 1). He underwent cystic aspiration in an emergent fashion when we collected 95 mL of a motor oil brownish fluid.

The patient was submitted to a pterional craniotomy with subtotal resection of the mass and wide fenestration of the cysts because of the potential hypothalamic involvement (Puget Grade 2).³² Also, fragments of capsule were left adherent to right carotid artery bifurcation, anterior communicating and anterior cerebral arteries, and pituitary stalk. He did a fast recovery postoperatively and was discharged completely alert, with visual disturbances and signs of panhypopituitarism.

Three months after resection, follow-up magnetic resonance imaging (MRI) disclosed the cysts had enlarged with three major components, namely right ventricular, right ventricular/basal ganglia, and intrasellar (►Fig. 2). Reoperation was performed for placement of an Ommaya reservoir under neuronavigation guidance at the most superficial ventricular cyst. The patient was otherwise clinically stable; therefore, he was scheduled for intratumoral therapy with α interferon, according to the protocol of Dastoli et al, which comprises the application of 3,000,000 IU three times a week for 4 weeks totalizing 36,000,000 IU.³³ The first follow-up MRI occurred 1 month after the first cycle disclosing a 47% reduction in the ventricular cyst volume, but an increasing intrasellar cyst (►Fig. 2). In parallel, he complained of pro-

gressive visual loss and later underwent endoscopic endonasal transsphenoidal partial CP excision 1 month before current admission. This surgery was particularly uneventful with no intraoperative cerebrospinal fluid (CSF) leak. Immediate postoperative CT revealed regular postoperative findings and regrowth of the ventricular cysts (►Fig. 3A).

The current admission occurred in the early postoperative phase. Headache and fever (38.0°C), together with overt meningeal signs (neck rigidity, positive Kernig and Brudzinski signs), indicated the diagnosis of meningitis. A lumbar puncture was performed revealing a clear-water CSF with pleocytosis (713 cells/mm³, neutrophils 89%), elevated protein level (124 mg/dL), a glucose count of 49 mg/dL, and no cholesterol crystals. White cell count was 7,100 cells/mm³ with a normal differential count. Ceftriaxone and vancomycin were started on the assumption of postoperative meningitis. However, on the third admission day, the patient was still obtunded with meningeal signs, even though afebrile. CSF culture was otherwise negative. Subsequent MRI disclosed a significant reduction in the superficial ventricular cyst indicating the diagnosis of cystic spontaneous rupture ultimately causing aseptic meningitis (►Fig. 3B). Steroids were immediately given, which was followed by a substantial improvement of neurological symptoms. The patient was asymptomatic at the second day of treatment. A second lumbar puncture yielded 106 cells/mm³ (lymphocytes 60%), the protein was 61 mg/dL, and the glucose 55 mg/dL. The patient was discharged asymptomatic and then scheduled for radiation therapy. One year after rupture, ventricular and sellar cysts showed almost complete remission (►Fig. 4). The patient provided informed consent for the publication of this manuscript.

Discussion

Aseptic meningitis due to rupture of an intracranial tumor is a relatively well-known event,³⁴ but the spontaneous rupture of CPs causing aseptic meningitis is even more rare with 20 reported patients in the English literature (►Supplementary Table S1 [online only]).^{5,8-10,12-24} Such clinical scenario comprises the most frequent clinical presentation of spontaneous CP rupture, which also includes improvement of neurological

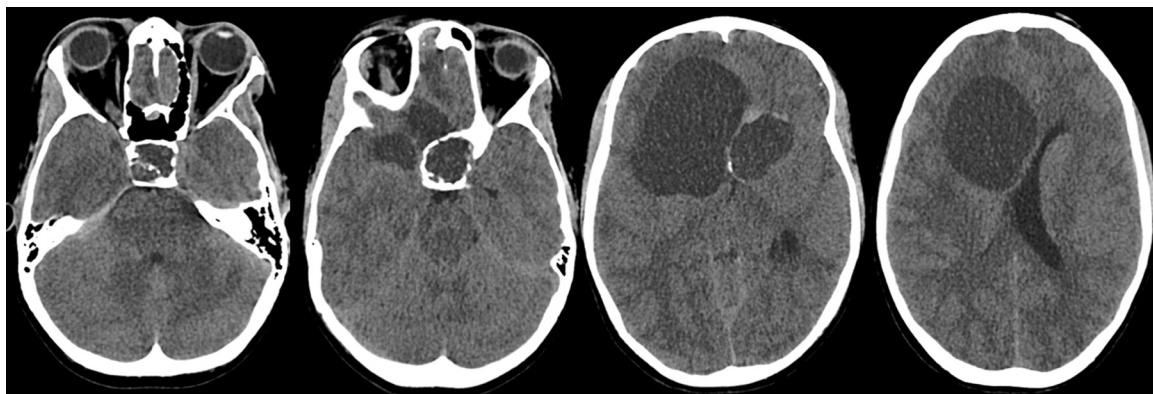


Fig. 1 Sequential cranial computed tomographic images without contrast enhancement at the time of diagnosis showing the giant multicystic mass with gross calcification pattern on cyst walls.

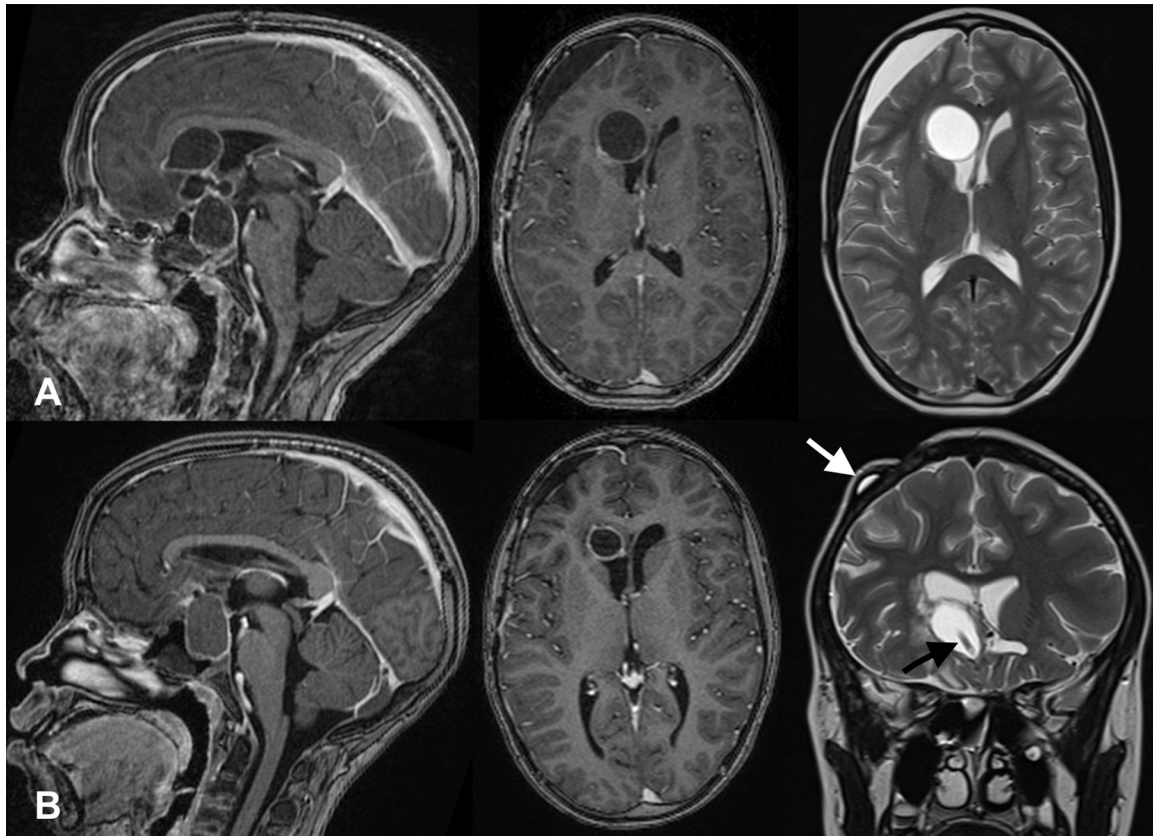


Fig. 2 T1-weighted gadolinium enhanced and T2-weighted magnetic resonance images obtained 3 months after the initial resection of the tumor (A) and after 1 month of intracystic therapy administered through the Ommaya reservoir (arrows) (B).

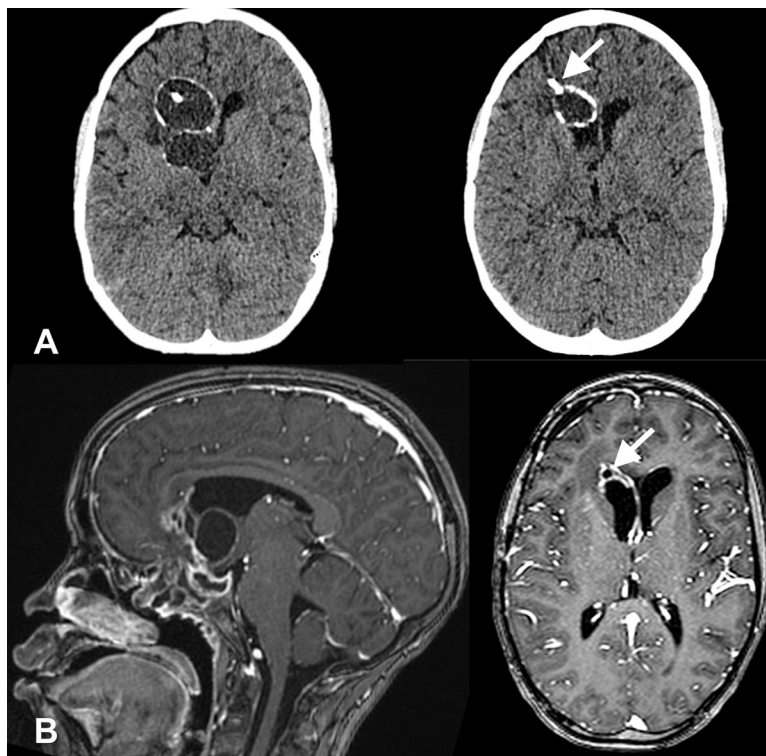


Fig. 3 Immediate postoperative cranial computed tomographic without contrast enhancement obtained at the time of endoscopic endonasal transsphenoidal surgery (A). T1-weighted gadolinium-enhanced magnetic resonance images done in the current admission (1 month postoperatively), which is found lining the Ommaya catheter (arrows) (B).

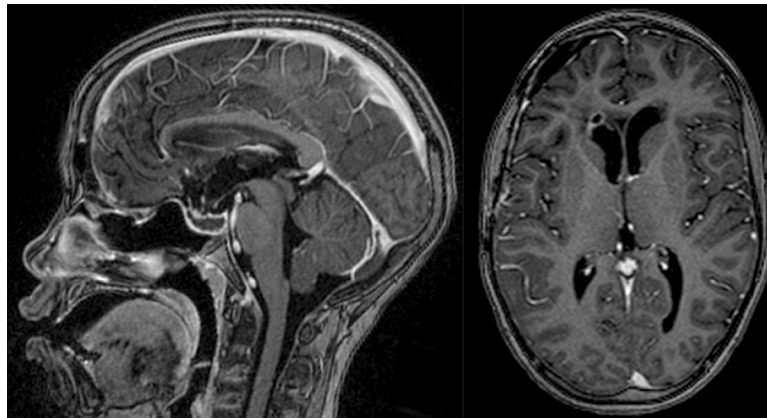


Fig. 4 T1-weighted gadolinium-enhanced magnetic resonance images done 1 year after spontaneous rupture showing no tumor regrowth and complete remission of the right ventricular/basal ganglia cyst.

deficits,^{11,27,30} asymptomatic rupture documented on follow-up imaging,^{25,31} and vasospasm.²⁹ There is a considerable increase in spontaneous CP rupture diagnosis after the 1980s, which is associated with the advent of CT and MRI.²⁰

Aseptic meningitis in the scope of ruptured CPs mainly affects adults with a slight male predilection and papillary histological subtype (► **Supplementary Table S1** [online only]). Few pediatric and adolescent patients have been reported,^{8,16,18,23} however. The pathophysiology of spontaneous CP rupture is yet to be fully elucidated, but it seems to involve weakness of the cystic wall caused by cystic growth.⁴ The formation of large cysts is associated with degenerative changes of the stroma and maturation of the squamous epithelium.⁴ Perhaps, this could explain the higher likelihood of ruptured CPs in adults, which are affected by their tumors for a longer period of time. Our patient is unique since the ruptured ventricular cyst was previously treated with intracystic therapy and had an Ommaya catheter, certainly contributing to further weakening of the cystic wall.

Myriad clinical presentations of patients with aseptic meningitis due to CP rupture are nonspecific and believed to occur by the chemical effect of cholesterol into the subarachnoid space.^{8,15,16} Diagnosis is generally suggested by an altered CSF analysis (pleocytosis and high protein levels) with negative culture.^{14,16–18,20} It is worth mentioning that CSF may be abnormal in as much as 80% of the patients having tumors greater than 3 cm in diameter.⁴

Diagnostic hallmarks comprise the findings of cholesterol crystals^{17,18,20} or elevated cholesterol levels^{13,16–18,20,23,24} on CSF analyses, and/or cyst reduction on subsequent imaging^{11,20,23,24} occurring in 15.8, 36.7, and 21.1% of the reported patients with aseptic meningitis, respectively (► **Supplementary Table S1** [online only]). In addition, cystic content changes to CSF density or intensity on imaging are highly suggestive of a ruptured CP.^{11,13,24} But, a high level of suspicion is the most important for a correct and prompt diagnosis. Treatment is straightforward with intravenous steroid administration^{10,12,13,16–18,20–22,24} and surgery is reserved for maintained mass effect tumors^{10,12–16,18,20,24} or cyst regrowth after rupture.^{11,30} In this regard, the course of ruptured cysts is poorly understood. It seems to be a transi-

tional condition requiring a definite treatment since most of the ruptured cysts generally demonstrate some regrowth on follow-up.^{11,24,30} The patient presented by Shinohara et al and ours had a successful evolution at the late follow-up, but one should consider our patient underwent radiation therapy.²³

Conclusion

We report on a rare case of aseptic meningitis due to spontaneous rupture of a multicystic sellar and suprasellar CP, which contained an Ommaya catheter. Facing a patient with a cystic CP with primary or recurrent meningeal signs or even sudden neurological improvement, clinicians should be aware of spontaneous CP rupture and look actively for the occurrence of cholesterol crystals or elevated CSF levels of cholesterol, as well as prompt follow-up imaging of the sellar region.

Conflict of Interest

None declared.

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