Neurol Med Chir (Tokyo) 53, 000～000, 2013

Bilateral Retrocerebellar Arachnoid Cysts Exerting Mass Effect and Associated With Cerebellar Tonsillar Ectopia in an Otherwise Healthy Adult

—Case Report—

Tim KILLEEN,[1] Chani TROMOP-VAN-DALEN,[1]
Hamish ALEXANDER,[1] and Agadha WICKREMESEKERA[1]

[1]Department of Neurosurgery, Wellington Regional Hospital, Wellington, New Zealand

Abstract

Rarely, midline or unilateral posterior fossa arachnoid cysts (ACs) exert local mass effect resulting in the symptoms and signs of cerebellar and brainstem dysfunction. These cysts are sometimes seen in conjunction with cerebellar tonsillar ectopia (TE), although the relationship between these two entities is unclear. Bilateral ACs in the posterior fossa are virtually unprecedented. We describe the case of a 33-year-old man with a history of multiple minor head injuries observed to harbour asymptomatic, bilateral cerebrospinal fluid-density collections over the cerebellar hemispheres. Six years later, he presented with headaches, limb paraesthesias, and drop attacks. Computed tomography, magnetic resonance imaging, and operative findings during burrhole drainage of the lesions showed bilateral posterior fossa ACs, with associated cerebellar TE of 11 mm. The cysts partially recurred, necessitating reopening of the burrholes, after which the patient's symptoms resolved entirely. We then discuss the challenges in diagnosing this unusual case, the relationship between AC and TE, and the role of minor head injury in the symptomatic progression of AC.

Key words: arachnoid cyst, posterior fossa, tonsillar ectopia, head injury, retrocerebellar cyst

Introduction

Midline or unilateral posterior fossa arachnoid cysts (ACs) are rare causes of local mass effect resulting in the symptoms and signs of cerebellar and brainstem dysfunction.[4,5,10,16] These cysts are sometimes seen in conjunction with cerebellar tonsillar ectopia (TE), although the relationship between these two entities is unclear.[5,11] Bilateral ACs in the posterior fossa are virtually unprecedented. We describe the case of a 33-year-old man with a history of multiple minor head injuries found to harbour asymptomatic, bilateral ACs over the cerebellar hemispheres. We then discuss the challenges in diagnosing this unusual case, the relationship between AC and TE, and the role of minor head injury in the symptomatic progression of AC.

Case Report

A 33-year-old male presented with headache, dizziness, fatigue, intermittent dysarthria, bilateral upper and lower limb paraesthesias, and drop attacks persisting for several weeks. Outpatient workup revealed no cardiovascular abnormality. He also related a 5-year history of occasional bilateral hand tremors and dizziness, gradually increasing in frequency. He was a keen rugby player and worked as a plasterer, and reported many prior instances of falls and minor head injuries but denied any recent trauma. Six years previously he had presented to the emergency department after striking his occiput in a 2-metre fall from scaffolding. Computed tomography (CT) performed 6 hours after this injury was normal except for symmetrically located, bilateral posterior fossa subdural fluid collections of cerebrospinal fluid (CSF) density, considered at the time to be incidental variants not warranting follow up.

Repeat CT demonstrated the previously imaged bilateral, biconvex subdural collections exerting mass effect on the cerebellum, causing right-sided effacement of the fourth ventricle (Fig. 1A). Magnetic resonance (MR) imaging confirmed these lesions contained fluid consistent with CSF, appearing as hypointense and homogeneous on T_1-weighted imaging (Fig. 2A) and hyperintense on T_2-weighted imaging (Fig. 2B), with no blood products apparent on gradient echo sequence. Cerebellar TE of 11 mm was also demonstrated (Fig. 2C). There was no syrinx or hydrocephalus, and the corpus callosum, cerebellar vermis, and fourth ventricle were normal. No other intracranial abnormalities were present.

Bilateral, suboccipital burrhole drainage was performed using image guidance. On opening the dura via the left

Received March 1, 2012; Accepted May 1, 2012

burrhole, a delicate, apparently arachnoid membrane was incised and clear, colourless fluid expelled under high pressure, confirming these collections to be ACs. The same procedure was then performed on the right, which was also under considerable pressure despite decompression of the contralateral compartment, with no evidence of communication between the cysts. The cerebellum was observed to expand bilaterally, occupying the drained spaces. Fluid protein and glucose levels were 0.27 g/l and 3.2 mmol/l, respectively, with 5×10^6 white cells/l and 91×10^6 red cells/l. The patient's symptoms resolved entirely and he was discharged on day 3. CT showed good decompression of the left cyst, with partial decompression of the right cyst and resolution of the mass effect (Fig. 1B). Histological analysis of the membrane showed a delicate, fibrous structure with no epithelial lining, arachnoid granulations, or inflammatory changes consistent with arachnoid rather than subdural membrane.[15]

After 2 months, the patient remained asymptomatic and had returned to work. MR imaging showed reduction in the size of the cyst and resolution of the previous mass effect on the cerebellum. TE was unchanged at 11 mm. Nine months postoperatively, the patient returned with renewed headache, mild ataxia, and right paraesthesia but no further syncope. MR imaging showed increased convexity of the right cyst and mild effacement of the fourth ventricle. The patient was admitted for reopening of the burrholes. Clear fluid was once more expelled under high pressure from the right burrhole, which was associated with a small, pressurised, extracranial pseudomeningocele. A negligible amount of fluid was encountered on the left.

Three months following his revision operation, the patient was completely asymptomatic and CT showed marked reduction in size of both cysts and resolution of the mass effect (Fig. 1C). He remained entirely well 18 months postoperatively.

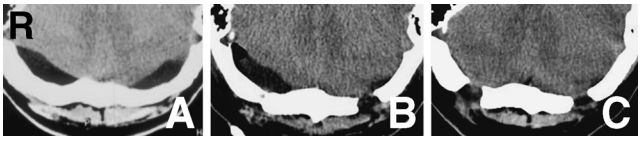

Fig. 1 A: Preoperative axial head computed tomography (CT) scan demonstrating cerebrospinal fluid collections causing cerebellar mass effect. B: Axial head CT scan showing partially resolved collections at 1 day after first burrhole drainage. C: Axial head CT scan showing almost complete resolution of the cysts at 3 months after second burrhole drainage.

Discussion

The differential diagnosis of chronic extra-axial CSF-density lesions in the posterior fossa is not always straightforward and includes subdural hematoma (SDH), subdural effusion (SE), cystic tumour, and the various cystic malformations of the Dandy-Walker/megacisterna magna complex as well as AC. Up to 1.75% cases of acute SDH occur in the posterior fossa,[12,13] usually secondary to trauma or anticoagulation therapy. Some cases may develop into chronic SDHs, an occurrence which seems commoner at the extremes of age.[14] MR imaging usually shows established chronic SDH as hyperintense with respect to brain tissue on T_1- and T_2-weighted imaging, and thus easy to differentiate from lesions containing CSF appearing as hypointense on T_1- and hyperintense on T_2-weighted imaging.[7] Although acute rebleeding into a chronic SDH can occasionally mimic CSF collections on MR imaging, gradient echo sequences reveal the presence of blood products. Infratentorial SE, thought to arise following disruption of the arachnoid/dura interface or due to local CSF flow anomalies, have been reported following foramen magnum decompression and trauma.[1] The natural history of these lesions seems to be gradual resolution or transformation into chronic SDH,[9] although they may exert mass effect on the cerebellum and/or brainstem and require evacuation.

In the present case, the nature of the lesions was initially unclear. MR imaging excluded chronic SDHs, so it was at first felt that the lesions were SEs arising from arachnoid tears secondary to occipital head trauma. Several factors led us to reject this diagnosis in favour of retrocerebellar ACs, despite their bilateral, symmetrical configuration being most unusual for ACs. The chronicity of the cysts, first recorded 6 years prior to this presentation, is something never before documented with SE/hygroma. The operative findings of arachnoid membranes further supported this diagnosis.

ACs are benign, congenital CSF collections which are thought to arise through aberrant duplication of the arachnoid during development.[2,8] The contents of ACs have been shown to be similar, but not identical, to CSF,

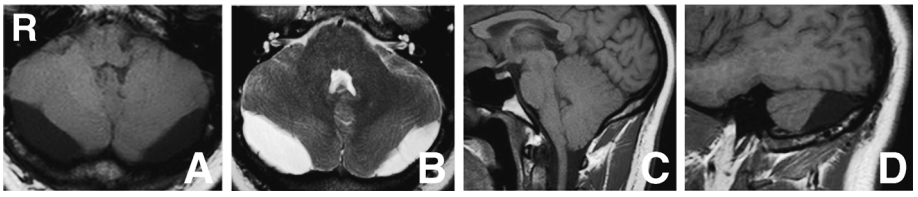

Fig. 2 A: Axial T_1-weighted magnetic resonance (MR) image showing bilateral hypointense retrocerebellar subdural collections. B: Axial T_2-weighted MR image confirming fluid of cerebrospinal fluid density, and mass effect on the right cerebellum and fourth ventricle. C: Mid-sagittal T_1-weighted MR image showing cerebellar tonsillar ectopia of 11 mm. D: Left paramedian sagittal T_1-weighted MR image showing the relationship of the left retrocerebellar arachnoid cyst and the cisterna magna.

with reduced protein content (0.30 g/l vs 0.41 g/l in one study) cited as evidence for secretion or active transport as the mechanism underlying cyst expansion.[3] As our patient was treated with minimally invasive burrhole drainage, no CSF was available to provide comparison, but the protein level of the cyst contents was 0.27 g/l, at the low end of the normal range for CSF, and consistent with AC. The small number of red cells present was likely related to surgery.

ACs demonstrate a strong predilection for the middle fossa, where, despite its relatively small contribution to overall intracranial volume, two thirds of ACs are found.[18] Thirteen to thirty percent of ACs occur in the posterior fossa[6,18] and form part of an interesting spectrum of disorders including the Dandy-Walker/megacisterna magna complex, in which ACs are defined as discrete CSF collections within a normal-sized posterior fossa, separate from the fourth ventricle and associated with an intact vermis and a normally positioned torcula.[2,8] Posterior fossa ACs are retrocerebellar in up to 50% of cases, with others found in the cerebellopontine angle and quadrigeminal regions.[6,10]

More than 95% of ACs are unilateral or occupy the anatomical midline, with bilateral or multiple ACs almost exclusively restricted to the middle fossa, usually associated with other congenital malformations.[18] There was nothing in our patient's history or examination to suggest any such syndrome. While undoubtably rare, bilateral retrocerebellar ACs are possibly not unprecedented. Vaquero et al. report a case of a woman with ataxia, dysphagia, and nystagmus, who subsequently underwent suboccipital craniectomy revealing paramedian ACs overlying the cerebellar hemispheres. These cysts were drained and found not to be in communication with one another.[16] No imaging was performed and the case report was brief.

The present case was associated with cerebellar TE of 11 mm, although this was neither documented prior to presentation nor shown to resolve on drainage of the cysts. There is no consensus regarding the existence and mechanism of causation when these malformations coexist. The theory that retrocerebellar ACs exert a mass effect on the tonsils, chronically inducing herniation, is supported by some cases of unilateral or midline cerebellar hemispheric AC in which TE and/or syringomyelia was reversed through foramen magnum decompression and excision or drainage of the AC.[5,11]

It is tempting to infer that this patient's cysts, presumably harboured asymptomatically for several decades, gradually became symptomatic as a result of the significant blow to the occiput received when falling 6 years previously. ACs have been observed to expand (and in one case disappear[19]) following trauma, often with a significant delay between injury and symptom onset.[17] The mechanism accounting for this observation is unclear but bleeding or effusion from the cyst walls or the traumatic development of a one-way arachnoid flap-valve have been suggested.[17] In this case, the successful resolution of the cysts following simple drainage supports the former hypothesis over the latter.

While many surgeons would advocate endoscopic/open membranectomy or shunting for symptomatic posterior fossa ACs,[4,5] the present case was treated, perhaps controversially, with simple burrhole drainage. At first presentation and with considerable preoperative uncertainty regarding the nature of the collections and the contribution of TE to the patient's symptoms, this minimally invasive approach was chosen over a more extensive craniotomy +/− foramen magnum decompression. All options were revisited when the cysts recurred and the patient's preference for a minimally invasive procedure played a significant role in the decision to repeat burrhole drainage in the first instance. This approach provided complete resolution of the symptoms and a highly satisfactory result clinically and on final follow-up imaging.

This case highlights the difficulty sometimes encountered in diagnosing chronic CSF collections of the posterior fossa. The eventual diagnosis of bilateral retrocerebellar ACs is almost unprecedented, with one possible case previously reported. The association between retrocerebellar cysts and TE has previously been proposed to be causative and reversible, although while this patient's symptoms resolved with decompression of the ACs, stable TE of 11 mm remained on follow-up imaging. Mild to moderate head injury can precipitate asymptomatic ACs to expand and cause mass effect. We propose that occipital trauma triggered such a transformation in this patient causing gradually worsening symptoms over 5 years. This effect is worthy of further investigation.

Conflicts of Interest Disclosure

The authors confirm they are not aware of any conflicts of interest.

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Address reprint requests to: Mr. Tim Killeen MRCS, Sennhofstrasse 8, 8125 Zollikerberg, Switzerland.
e-mail: tim.killeen@doctors.org.uk