

Case report

Middle cranial fossa arachnoid cysts: not always a benign entity

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Abstract. Arachnoid cysts are often discovered as incidental findings on cranial imaging. A rare manifestation is described in a child presenting acutely with symptoms and signs of raised intracranial pressure.

Arachnoid cysts are often discovered as incidental findings on cranial imaging. However, they may occasionally become symptomatic because of their pressure effects on surrounding structures. Symptoms include headaches, seizures, mental retardation, cognitive impairment and, rarely, focal neurological deficit [1, 2]. Associations with both intracystic and subdural haemorrhages, due to rupture of bridging veins or vessels in the cyst wall are also well known [3]. We report a rare presentation of an arachnoid cyst as a rapidly expanding mass lesion with severe papilloedema.

Case report

An 11-year-old girl presented with a 3 week history of early morning vomiting and a 2 day history of headache and progressive visual loss. The only previous medical history of note was mild developmental delay.

On examination, the patient was distressed with a severe headache. Fundoscopy demonstrated marked bilateral papilloedema and retinal haemorrhages. Both pupils were dilated. The right pupil reacted sluggishly to light and the left pupil was fixed. Visual acuity was severely impaired and the patient could only perceive hand movements. No other neurological deficit was demonstrated.

Cranial magnetic resonance imaging showed a large left sided arachnoid cyst occupying most of the middle cranial fossa, with an associated mass effect manifested by ventricular compression and midbrain distortion (Figure 1). As a result of the significantly elevated intracranial pressure, there was dilatation of the subarachnoid space around

the optic nerves with elevation of the optic papilla (Figure 2).

Emergency craniotomy was performed with fenestration of the arachnoid cyst into the interpeduncular cistern. The headache was much improved post-operatively but her visual acuity remained poor. Cranial CT demonstrated a marked reduction in cyst size, re-expansion of a significant proportion of the previously compressed brain, less ventricular compression and resolution of the midbrain distortion.

Discussion

Arachnoid cysts comprise 1% of non-traumatic intracranial masses [1]. They consist of clear fluid enclosed between reduplicated layers of arachnoid membrane and give identical signal to cerebrospinal fluid (CSF) on MRI. Their origin and natural history have been widely debated. Primary localized cerebral agenesis has been proposed, but this theory neither explains why the brain re-expands following surgery [4] nor the spontaneous disappearance of arachnoid cysts which is often associated with rupture into the subdural space following trauma [5, 6]. Arachnoid cysts probably arise from maldevelopment of the leptomeninges [7] either at birth or soon after [8]. Following their initial growth early in life, most subarachnoid cysts remain stable over many years and are discovered as incidental findings on cranial imaging or at post-mortem. Some disappear spontaneously [9] and some continue to grow slowly [7, 10]. It is unclear why some cysts remain stable and asymptomatic and why others enlarge and become symptomatic.

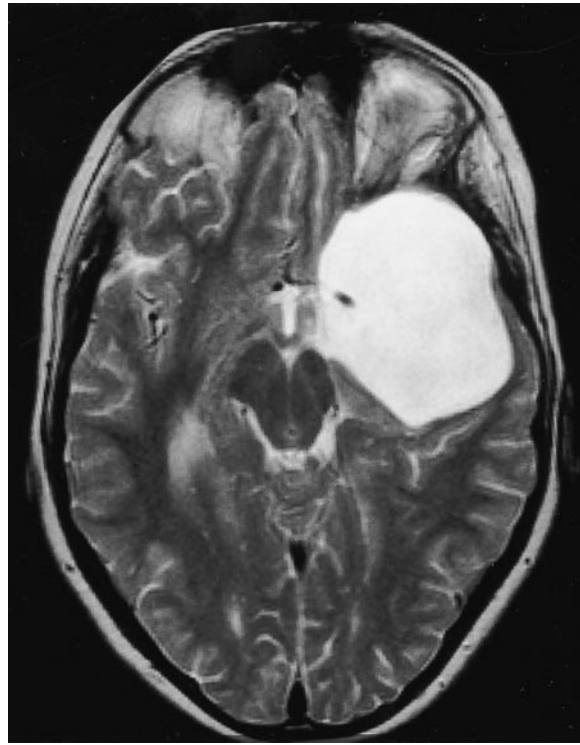
By far the commonest location is the anterior portion of the middle cranial fossa [7], with left-sided predominance [1]. Cysts in this location have been divided into three basic types [4] which increase in size and mass effect. Type 1, the mildest

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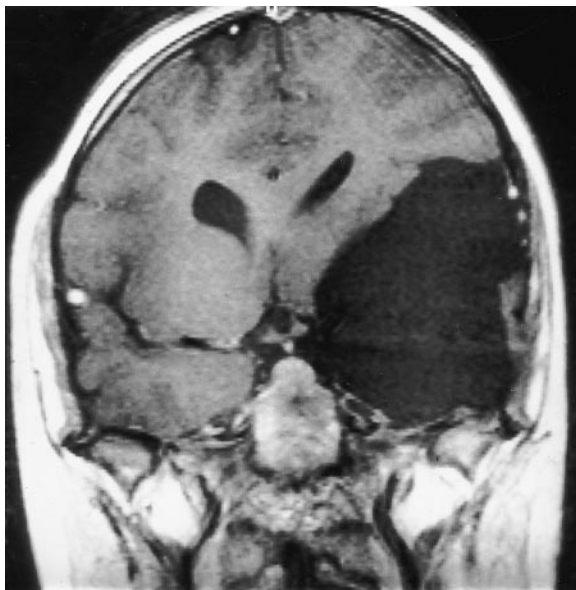
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(a)



(b)



(c)

Figure 1. T_2 weighted axial (a and b) and T_1 weighted coronal (c) MR images demonstrate the typical appearance of a large rounded arachnoid cyst, occupying most of the middle cranial fossa and splitting the opercula of the sylvian fissure (type 3). The left lateral ventricle is compressed and the brain stem is distorted.

form, freely communicates with the subarachnoid space, type 2 slowly communicates and type 3 is the largest and does not communicate. It is suggested that the three types represent subsequent stages in the evolution of the same initial pathological process. Type 1 cysts are arachnoid diverticula which, with progressive obliteration of their free communication with the subarachnoid space, enlarge and evolve into type 3. This patient had a large and rounded type 3 cyst, occupying much of the temporal fossa, splitting the opercula of the sylvian fissure and compressing the temporal and frontal lobes.

Hypotheses to explain progressive enlargement of an arachnoid cyst include the secretion of fluid by the cyst wall, fluid filtration through the cyst wall by osmosis or trapping of fluid within the cyst via a ball-valve mechanism [11]. The first two are obviously applicable to non-communicating cysts and the latter to communicating cysts. The brain usually accommodates this slow increase in size and the relatively rapid enlargement of the cyst as presumably occurred in this patient, with resultant marked elevation of intracranial pressure, is an extremely rare complication. One case of uncal herniation due to an arachnoid cyst has been



Figure 2. An enlarged T_2 weighted axial image of the right orbit demonstrating the MRI features of papilloedema with distension of the subarachnoid space around the optic nerve (arrows) and protrusion of the optic papilla into the posterior aspect of the globe.

previously reported [12], although the cyst enlargement and neurological deficit were apparently slowly progressive.

Arachnoid cysts may be surgically treated by craniotomy and fenestration, cystoperitoneal shunting [13], or endoscopic marsupialization [14].

Pressure changes in the intracranial space are readily transmitted to the optic papilla via the subarachnoid space of the optic nerve sheath complex. Optic papilla protrusion, enlargement of the optic nerve sheath and impairment of visual acuity may occur in patients with elevated intracranial pressure, [15, 16] as in this case. The upper limit of normal of the optic sheath diameter is 5 mm and measurements above this nearly always correspond to a CSF pressure of at least 30 mm Hg [17]. When severe pressure is exerted on the optic papilla or chronic local ischaemia occurs, permanent degeneration of the optic nerve head results as in this patient.

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