Spontaneous development of a de novo suprasellar arachnoid cyst

Case report

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Arachnoid cysts are commonly thought to arise from either congenital defects or trauma. In this article the authors report the spontaneous development of a suprasellar third ventricular arachnoid cyst whose origin was not clearly congenital or traumatic. At the age of 4 months, the patient presented with hypertonia, and a magnetic resonance (MR) imaging study showed no abnormalities. At the age of 2 years, the boy presented with headaches and projectile emesis, symptoms that prompted further imaging studies. An MR image of the brain revealed a suprasellar cyst and obstructive hydrocephalus. The cyst was endoscopically fenestrated, which led to long-term symptom resolution.

KEY WORDS • obstructive hydrocephalus • suprasellar cyst • cyst formation • pediatric neurosurgery

Intracranial arachnoid cysts are fluid-filled cavities that arise within the arachnoid layer. They occur fairly frequently, but are rarely found in the ventricles. The cysts are often asymptomatic, and can remain undetected for years. Thus, most opportunities to study the growth of arachnoid cysts occur when a patient undergoes an imaging procedure for an unrelated issue. The advent of more advanced imaging techniques has increased the frequency of these findings, but they still remain rare. Furthermore, the causes and dynamics of these cysts are not wholly understood. The majority of arachnoid cysts are thought to be congenital and to result from developmental abnormalities; however, various physical insults have also been proposed as possible instigators of their appearance and growth.1-8,15,18,21 Other reports have indicated that arachnoid cysts can spontaneously grow or resolve with no readily apparent cause.10,13,14,19,20 In this article we describe the spontaneous de novo development of a suprasellar third ventricular arachnoid cyst in a 2-year-old child. This is only the second documented case of an arachnoid cyst whose origin was not clearly congenital or traumatic, and it is the first such case in which the lesion was located in the third ventricle.14 The cyst caused obstructive hydrocephalus and was fenestrated endoscopically. The child has undergone follow-up serial MR imaging for 4 years, and thus far has had no recurrence of hydrocephalus.

Abbreviation used in this paper: MR = magnetic resonance.
Second Operation and Postoperative Course. The cyst was treated using endoscopic fenestration and shunt removal. The fenestration consisted of resecting the roof of the cyst and opening its bottom into the suprasellar cistern—a procedure akin to a third ventriculostomy. The cerebral aqueduct was revealed when the cyst membrane covering it was removed. An external ventricular drain was left in place for a few days. Postoperatively, the patient suffered from transient episodes of photophobia, but his preoperative symptoms resolved. Computerized tomography and MR images of the head showed that the cyst had resolved and that the ventricles had significantly decreased in size (Fig. 3).

Discussion

Intracranial arachnoid cysts have been implicated in a variety of neurological problems. In particular, third ventricular arachnoid cysts have been associated with hydrocephalus, seizures, developmental delays, and decreased visual acuity. Yet often these cysts remain asymptomatic and undetected. In fact, unless an imaging procedure is undertaken for an unrelated issue, it is likely that an asymptomatic arachnoid cyst will remain undiscovered for the entire lifetime of the patient. As such, detailed studies of the development of arachnoid cysts are rare. To date there are only five cases of enlargement of arachnoid cysts that have been documented using serial imaging techniques.\(^7,10,12,14\)

Arachnoid cysts are generally thought to be congenital or caused by discrete events such as trauma or intracranial surgery.\(^1,4,8,10,13,15,16,22\) In this patient, the cyst did not appear to be congenital because an MR image obtained when he was 4 months of age demonstrated normal ventricle size and no obvious cyst. In addition, the patient’s parents described no serious trauma or surgery during the child’s life. The origin of this cyst thus remains obscure.

Several hypotheses have been proposed in an attempt to explain the mechanism by which arachnoid cysts form, persist, and enlarge. Starkmen, et al.,\(^7\) proposed that congenital arachnoid cysts originate from the splitting of the arachnoid membrane. Recently, attempts have been made to support Starkmen’s theory through experimental measures, including an example of an arachnoid cyst in an intraarachnoid location.\(^5,10,12\) Mechanisms proposed to explain the enlargement and persistence of cysts include secretion of fluid by ependymal cells, fluid influx from an osmotic gradient, and a one-way valve mechanism.\(^7\) The valve mechanism has been witnessed during endoscopic fenestration of arachnoid cysts and has subsequently been gaining credence.\(^10,16\)
The original insult leading to the formation of the cyst might well be congenital yet imperceptible in the early imaging studies. However, the cause of the rapid enlargement remains unknown. With the ubiquitous use of MR imaging, more documented cases of symptomatic as well as asymptomatic arachnoid cysts are surely forthcoming. Hopefully, this body of data will elucidate these commonly idiopathic and sometimes pathogenic cysts.

Conclusions

This is the first documented case of a suprasellar third ventricular arachnoid cyst that formed spontaneously during early childhood. The mechanism of cyst formation and persistence is unclear. The lesion was successfully fenestrated endoscopically, resulting in a decrease in ventricle size and resolution of the symptoms.

References


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