Progressive symptomatic increase in the size of choroidal fissure cysts

Report of 4 cases

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Cysts of the choroidal fissure are often incidentally identified. Symptoms from such cysts appear to be exceedingly rare. Herein, the authors report a case series of symptomatic enlargement of choroidal fissure cysts that were surgically treated. Although cysts of the choroidal fissure do not normally become symptomatic, the neurosurgeon should be aware of such a complication. Based on the authors’ experience, surgical fenestration of such cysts has good long-term results.

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Key Words • cyst fenestration • choroidal fissure • treatment efficacy • pediatric neurosurgery • vascular disorders

The tela choroidea invaginates through the choroidal fissure to reach the lateral ventricles, and anomalous development may lead to the formation of a cyst along this fissure. Such a cyst formation may occur from an ependymal diverticulum that becomes isolated from the ventricular system, or from sequestration of neuroectoderm and its vascular pia mater.5 Cysts of the choroidal fissure are typically ovoid and have a long axis that is parallel to the choroidal fissure in the antero-posterior plane.6 Progression of such cysts is very rare. In this article we report our experience with the progression of choroidal fissure cysts. This very small cohort is derived from more than 75 surgical cases of intracranial arachnoid cysts treated over a 16-year period, with the exception of 1 patient who remains surgically untreated and asymptomatic.

Case Reports

Case 1

This boy initially presented at the age of 7 years. He had a history of autism and a prior neurosurgical procedure for a choroidal fissure cyst that was found when he was 8 months old. He was followed with serial imaging...
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and was found to have a symptomatic interval increase in cyst size, requiring an operation. The initial procedure, open fenestration with placement of a cystoperitoneal shunt for a choroidal cyst, was performed when he was 8 months old. Since the time of the initial procedure the patient had experienced no complications, and was not seen again until he was 7 years old, when CT studies found the cyst to be decreased in size. The patient continued follow-up for 5 years, when he began to develop headaches (at 12 years of age). Compared with earlier radiographs, the size of the choroidal cyst at the time of his presentation with headache was unchanged from the size of the cyst after fenestration and shunt placement. The patient continued to experience headaches until the age of 16 years, despite no change in the radiographic appearance of the lesion over the course of 15 years.

Case 2

This 6-year-old girl was referred from an outside institution for a right temporal choroidal fissure cyst (Fig. 1A). The finding of the cyst was incidental following a traumatic head injury after she fell down a set of stairs. On initial evaluation, the child had normal results on her neurological examination and an unremarkable medical history. A CT scan was obtained, which showed a right-sided choroidal fissure cyst measuring $2.2 \times 2.2 \times 2.5$ cm, with slight enlargement of the right temporal horn. The patient presented 2 years later with Valsalva-induced head pain and a radiographically confirmed increase in the size of her choroidal fissure cyst (Fig. 1B). Due to trapping of the anterior temporal horn and Valsalva-induced headaches, an endoscopic fenestration of the cyst into the ambient cistern was performed. There were no postoperative complications, and a CT scan obtained 3 years later showed that the cyst had decreased to a size of $1.5 \times 1.5 \times 1.5$ cm (Fig. 1C). The patient continued to be followed until she was 17 years old, when she was last seen for new complaints of temporal pain and headaches. An MRI study was ordered, and the size of the cyst had not changed from its size 3 years postoperatively.

![Fig. 1. Case 2. Admission axial MRI study (A) revealing a choroidal fissure cyst. Follow-up MRI study (B) demonstrating that the cyst had enlarged over time; it had also become symptomatic. Axial MRI study (C) obtained after fenestration, showing that the cyst size had decreased.](image)

Case 3

This 8-week-old girl was referred for the incidental finding of a right-sided temporal cyst. The cyst was first identified during a perigestational ultrasound. The patient had been delivered vaginally without complications. After delivery, an extruterine ultrasound demonstrated a right-sided cyst at the level of the temporal horn measuring $0.9 \times 1.1 \times 1.1$ cm. The initial evaluation of the child at 2 months of age showed no neurological deficits and no history of seizures. Additionally, the patient had an age-appropriate head circumference and no clinical evidence of hydrocephalus. A CT scan that was obtained when the child was 3 months old demonstrated a cyst size of $1.5 \times 1.6$ cm, with no appreciable communication to the right temporal horn, midline shift, or intracranial hemorrhage. When the child was 9 months old, a follow-up CT study was obtained, showing an increase in the size of the cyst to $2.3 \times 1.8$ cm, with irregular borders. The patient remains asymptomatic, with a normal head circumference, and the patient’s family was given the option for surgical intervention based on the radiographic change. The case is currently ongoing.

Case 4

This 2-year-old boy was referred for evaluation of a right-sided choroidal fissure cyst (Fig. 2A). This was an incidental finding during an evaluation for sleep apnea. The patient was subsequently found to have pertussis, was treated, and his apnea resolved. On imaging, the cyst measured $0.9 \times 0.8$ cm. Two years later, the child was evaluated for trauma to the head after being struck with a bat. After the event, the patient returned to baseline with no deficits. A head CT scan obtained at this time found that the choroidal fissure cyst had doubled in size over the prior 2 years, to $2.3 \times 2.0$ cm (Fig. 2B). Due to the increase in size over a 2-year period, it was suggested to the family that fenestration of the cyst should be performed. The patient underwent cyst fenestration with no postoperative complications. Pathological examination of a portion of the cyst wall demonstrated normal meninge-
al tissue consistent with arachnoid mater. At the 1.5-year follow-up visit, imaging demonstrated that the size of the cyst had decreased to 1.4 × 1.4 cm (Fig. 2C). The patient remains asymptomatic and seizure free.

**Discussion**

The choroidal fissure extends from near the foramen of Monro to the hippocampal fissure. This channel is usually shallow and curves posterosuperiorly from the anterior temporal lobe to the atrium of the lateral ventricle. This fissure is separated from the temporal horn only by a layer of epithelium derived from the roof plate of the telencephalon, known as the tela choroidea, and contains the choroid plexus of the inferior horn. The choroidal fissure, the cleft along which the choroid plexus of the temporal horn is attached, is situated at the junction of the roof and lateral wall of the middle incisural space between the fimbria below and the pulvinar above. The inferior choroidal point, at the inferior end of the choroidal fissure, is located immediately lateral to the lateral geniculate body and posterior to the hippocampal head, where the anterior choroidal artery enters into the temporal horn.

Sherman et al. reviewed the MRI studies of choroidal fissure cysts and reported 26 cases, mostly adults, with neurological sequelae such as epilepsy, migraines, gait disturbance, tremor, paresthesias, and hemiparesis. They concluded, however, based on imaging and basing surgical intervention on the degree of compression, that following the cyst size with serial imaging and basing surgical intervention on the degree of compression is an appropriate course of treatment.

In the aforementioned case reports, it is important to note that choroidal fissure cysts do not communicate with the ventricular system, so a VP shunt alone would not be expected reliably to reduce the size of a choroidal fissure cyst. Okamura et al. reported 1 case of a cyst of the choroidal fissure in a 42-year-old man with seizures and vertigo. The cyst was treated with partial resection of its wall, and surgical findings showed that it was attached to the choroid plexus, which the investigators believe indicated that it derived from arachnoid mater that invaginated into the choroidal fissure. Histochemical analysis and light microscopy confirmed that the cyst walls were composed of arachnoid mater, as was the case with 1 patient of ours for whom histological examination of the cyst wall was performed.

Our experience demonstrates that choroidal fissure cysts that increase in size may be effectively treated with fenestration and possible VP shunting, with a low risk of complication. How such cysts are different histologically or clinically is not clear based on the current literature.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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