Case Report

Endoscopic Treatment of a Suprasellar Arachnoid Cyst Causing Precocious Puberty

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Summary

Suprasellar arachnoid cysts are uncommon developmental anomalies that are most often diagnosed in childhood. They constitute approximately 9% of all arachnoid cysts. These cysts require special consideration because of the unique location where they more commonly become symptomatic. Precocious puberty is rarely the presenting sign of suprasellar arachnoid cyst. A rare case of suprasellar arachnoid cyst presenting with precocious puberty is reported. Treatment was via endoscopic ventriculocystocisternostomy. The case is described and the advantages of endoscopic treatment of suprasellar arachnoid cyst is emphasized.

Key words: Arachnoid cyst, endoscopy, suprasellar, precocious puberty

INTRODUCTION

Arachnoid cysts are developmental anomalies of the arachnoid membrane with intraarachnoidal collection of cerebrospinal fluid. They can arise in any part of the central nervous system. Suprasellar region is a rare location for arachnoid cysts. The majority of the suprasellar arachnoid cysts become symptomatic in early childhood. Precocious puberty is an uncommon presentation with the few cases reported in the pertinent literature. Several surgical procedures are used for the treatment of these cysts. However, suprasellar arachnoid cysts are amenable to endoscopic surgery because of their location in the anterior portion of the third ventricle and the hydrocephalic changes they create. We report a case of suprasellar arachnoid cyst presented with precocious puberty and treated endoscopically.

CASE PRESENTATION

A 7 year-old-girl presented with a 4 months history of headache, breast enlargement and axillary and pubic hair development. Her neurological examination was normal. Physical examination revealed development of breast tissue as classified Tanner's stage 3, both pubic and axillary hair development. The bone age by skeletal survey of the
hand was rated as 8 years 10 months. Endocrinological examination showed normal serum levels of luteinizing hormone, follicle-stimulating hormone and estradiol. Gonadotropin releasing hormone (GnRH) test was done and elevated plasma concentrations of luteinizing hormone, follicle-stimulating hormone and estradiol were found. Magnetic resonance imaging (MRI) showed a suprasellar cyst which was hypointense on T1 weighted image and hyperintense on T2 weighted image with no contrast enhancement (Figure 1). The patient underwent surgery.

**Operation.** A single burr hole was placed and slightly enlarged using rongeurs 1 cm just in front of the coronal suture and approximately 3 cm from the midline. A rigid endoscope was passed into the right frontal horn of the lateral ventricle, revealing a large cystic mass with a semitranslucent, blue membrane obstructing the foramen of Monro (Figure 2A). A combination of bipolar electrocautery and sharp dissection was used to create a large fenestration in this region of the cyst. The endoscope was then navigated into the lesion, where the infundibular stalk, dorsum sella, both internal carotid arteries and posterior communicating arteries as well as the ventral surface of the brainstem could be identified (Figure 2B). The endoscope was navigated into the preptone cistern, where the bilateral abducent nerves and basillary artery were identified. The membrane at the basal surface of the cyst was identified and blunt dissection was used to create a fenestration in front of the basillary artery between the dural surface of the clivus and the ventral surface of the brainstem. This was enlarged using a No.3 French embolectomy catheter. The membrane became pulsatile with excursions of the cardiac cycle and it was believed that a complete fenestration of the cyst had been achieved (Figure 2C).

The postoperative course was uneventful. Headache improved after surgery. Precocious puberty did not resolve. She was discharged and monitored at our pediatric endocrine clinic. Long-acting GnRH analogue treatment was administered. At 3., 6., 12. months follow-up, no significant decrease in cyst volume was observed in MRI (Figure 3). No further surgery was performed. Serum levels of luteinizing hormone, follicle-stimulating hormone and estradiol significantly decreased. She is still at follow up.

![Figure 1: MRI showing a large suprasellar arachnoid cyst with obstructive hydrocephalic changes A. Axial T1 weighted image, B. Sagittal T1 weighted image.](image-url)
Figure 2: A. Endoscopic view of a large cystic mass with a semitranslucent, blue membrane obstructing the foramen of Monro (ac: arachnoid cyst, cp: choroid plexus, sv: septal vein), B. Endoscopic view within the cyst (BA: basillary artery, DS: Dorsum sella, NIII: oculomotor nerve, PCA: Posterior cerebral artery, PComA: Posterior communicating artery), C. Endoscopic view showing the stoma at the base of the suprasellar arachnoid cyst surrounding the basillary artery (BA: basillary artery, S: Stoma), D. Endoscopic view showing the second stoma at the base of the suprasellar arachnoid cyst behind the dorsum sella (BA: basillary artery, DS: Dorsum sella, S: Stoma).

Figure 3: Follow-up MR scan at 12 month showing no reduction in size of the cyst.
DISCUSSION

Suprasellar arachnoid cysts arise from an anomaly of the diencephalic membrane of Liliequist, either as a diverticulum or from splitting of a membrane and secretion of cerebro-spinal fluid (CSF) within the cavity. This diverticulum could increase in size following inflammatory, haemorrhagic or developmental events. Alternatively a ball valve like mechanism may allow ingress but not egress of CSF from the cyst\(^{14,11}\).

Typically, suprasellar arachnoid cysts expand from the prepontine space, displacing the floor of the third ventricle upwards, the pituitary stalk and optic chiasm upwards and forwards, and the mamellar bodies upwards and backwards. As the cyst increases in size it fills and occludes the third ventricle, and distorts and blocks the aqueduct, which finally results in hydrocephalus\(^6\).

Two different types of suprasellar arachnoid cyst are described: a non-communicating intra-arachnoid cyst of the diencephalic membrane of Liliequist, and a communicating cyst that is a cystic dilatation of the interpeduncular cistern. Cystic dilatation of the interpeduncular cistern occurs between the two leaves of Liliequist's membrane and basillary artery bifurcation lies inside the cyst. In case of the occurrence of intra-arachnoid cyst of the diencephalic membrane, the interpeduncular cistern would be compressed, leaving the basillary artery bifurcation behind the posterior wall of the cyst\(^{3,6,11}\).

60-90% of the reported patients are children\(^4\). Obstructive hydrocephalus, visual disturbance and endocrine dysfunction are the common symptoms. The incidence of precocious puberty in suprasellar arachnoid cysts is uncertain. It is an uncommon presentation with only few cases reported in the literature\(^{1,4,5,7,8,10-14}\). The pathogenesis of precocious puberty in suprasellar arachnoid cysts is unclear. It is believed that the mass effect of the arachnoid cyst upon the hypothalamus is responsible for the development of precocious puberty\(^{11,14}\).

Magnetic resonans imaging is the diagnostic method of choice in the recognition and differential diagnosis of suprasellar arachnoid cysts. The cyst appears hypointense on T1-weighted images and hyperintense on T2-weighted images with no contrast enhancement. The MR signal intensity is similar to that of CSF. The marked dilatation of the third and lateral ventricles gives a typical “Mickey Mouse” appearance on axial images.

Several surgical procedures, including craniotomy and cyst fenestration or resection, cystoperitoneal shunting and stereotactic aspiration were used for treating this pathology in the past. In recent years, in the presence of hydrocephalus endoscopy is considered to be the best treatment modality due to its less invasiveness and low complication rates. Successfull results have been reported in gradually increasing in number in the treatment of suprasellar arachnoid cysts by endoscopic procedures\(^{2-6,9,11}\).

There is still a controversy about the type of endoscopic surgery whether ventriculocystostomy or ventriculocystocisternostomy. In a study by El-Ghandour NM et al they compared endoscopic ventriculocystostomy or ventriculocystocisternostomy in 25 cases and found that both procedures are almost equally effective clinically and radiologically. But long term follow up showed low recurrence rates with ventriculocystocisternostomy\(^4\).

Considering these results, we performed ventriculocystocisternostomy in our case.

One of the most striking feature during follow-up period is the inability to resolve endocrin dysfunction. Isolated endocrine
dysfunction is reported to be an contraindication to surgery\(^{(6,11)}\).

In conclusion, suprasellar arachnoid cyst should be kept in mind in patients presenting with precocious puberty. Neuroendoscopic procedure is a safe, effective and minimally invasive method in the treatment of these cysts.

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