Suprasellar Arachnoid Cyst: A 20-Year Follow-Up after Stereotactic Internal Drainage: Case Report and Review of the Literature

Approximately 9 to 15% arachnoid cysts occur in the sellar or suprasellar region. The optimal management of symptomatic suprasellar cysts continues to pose a challenge to neurosurgeons. This case report describes a patient with a suprasellar arachnoid cyst who presented at the age of 3 and was followed for 20 years after cystoventriculostomy. A girl was referred to neurosurgery department by a pediatric clinic because computed tomography demonstrated a suprasellar cystic lesion. The neurological examination and all routine blood tests including hormone profiles were normal. The physical examination was unremarkable except premature thelarche. Stereotactic ventriculocystostomy was performed using a catheter providing permanent internal drainage. Postoperative cystoventriculography showed contrast medium in both the cyst and the ventricles. Control computed tomography confirmed that the ventricular cyst catheter was within the cyst. During a follow-up of 20 years, signs of precocious puberty disappeared and the girl showed normal sexual development. Endocrine profiles and visual function remained normal. The stereotactic approach to suprasellar arachnoid cysts is a safe procedure in experienced hands.

KEY WORDS: Cystoventriculostomy, Premature thelarche, Stereotactic management, Suprasellar arachnoid cyst

ÖZ

ANAHTAR SÖZCÜKLER: Kistoventrikülostomi, Stereotaktik cerrahi, Suprasellar araknoid kist
INTRODUCTION

The majority of arachnoid cysts arise in the middle cranial fossa with approximately 9 to 15% occurring in the sellar or suprasellar region (7,11,30). Sellar and suprasellar arachnoid cysts may be asymptomatic or may cause headache, optic nerve compression, psychomotor retardation, endocrine dysfunction, or hydrocephalus (3,7,27,37,40). The optimal management of symptomatic sellar or suprasellar arachnoid cysts continues to present a challenge to neurosurgeons (5,7,9,37). We here report a patient with a suprasellar arachnoid cyst who presented with premature thelarche and who was followed up for 20 years after cystoventriculostomy. We have not encountered in the literature any other case report of a suprasellar arachnoid cyst with a 20-year follow-up after treatment.

CASE REPORT

A 3-year-old girl was admitted to a pediatric hospital with signs of precocious puberty. There was no previous head trauma, central nervous system infection or any other illness. The neurological examination was normal and all routine blood tests including hormone profiles were within normal limits. Her physical examination was unremarkable except for premature breast enlargement. A skull X-ray revealed widening of the dorsum sellae. Computed tomography (CT) demonstrated a suprasellar cystic lesion isodense with cerebrospinal fluid (CSF). The lesion was approximately 35x25 mm in size and compressed the hypothalamus and pons (Figure 1A). Cerebral angiography showed caudal displacement of the basilar artery (Figure 1B).

Stereotactic Management

After attachment of the stereotactic frame (Riechert system), the co-ordinates of the cyst were localized by means of the CT image data. According to its localization, the cyst was approached with a 13-gauge cannula via a frontal precoronal paramedian burrhole. The procedure was performed in the routine manner and included:

1. Visualisation of the cyst by filling with air and iodine contrast medium under fluoroscopic control (Figure 1C). 2. Endoscopic inspection of the cyst through a rigid endoscope (2.8 mm diameter) with straight and 70-degree views. 3. Implantation of a Rickham silicone catheter with additional holes to establish a connection between the cyst content and the ventricular system for drainage (Figure 1D).

The operation was carried out under general anesthesia. The cyst wall was punctured and CSF drained spontaneously through the cannula probe. 5 ml of clear, colorless fluid was removed. Routine laboratory studies of the aspirated fluid were consistent with CSF. There was no problem during surgery and the postoperative course was uneventful. CT cystoventriculography performed 9 months postoperatively showed contrast medium in the cyst and in the lateral and third ventricles. CT examinations performed postoperatively, 1, 3 and 9

Figure 1A: Preoperative CT scan revealing an intra- and suprasellar cystic lesion with the same density as CSF. The lesion is seen to compress the hypothalamus and midbrain.

Figure 1B: Preoperative angiography showing the basilar artery displaced posteriorly.
months and one year after the intervention revealed a stable cyst and demonstrated absence of hydrocephalus or subdural effusion. The cyst volume was 22ml. The ventricular cyst catheter was in the cyst. Thereafter, the patient was followed up by CT every year until 5 years postoperatively, then by CT once every two years and subsequently once every three years. All follow-up CT scans confirmed a stable cyst (Figure 2 and 3). Breast enlargement disappeared gradually in a year. In the course of follow-up, there was normal growth velocity and her body mass index increased in an age-appropriate manner while other endocrine parameters remained within normal limits.

**DISCUSSION**

Congenital intracranial arachnoid cysts are found in 0.16% of all newborns (17), comprising 1% of all intracranial mass lesions (13). More than 75% of arachnoid cysts arise in the supratentorial
compartment (40–50% in the sylvian fissure) and approximately 9 to 15% occur in the sellar region (11,26,30). Sellar arachnoid cysts are divided into suprasellar and intrasellar cysts. Suprasellar arachnoid cysts are by far the more common type. Most suprasellar arachnoid cysts occur in children and there is a known male prevalence (12).

The etiology and pathophysiology of arachnoid cysts remain debated. Several theories have been postulated regarding their formation. Starkman et al. consider the cysts to be of intra-arachnoid origin and occur as a result of splitting of the arachnoid membrane during early embryonic development (37). Rengachary and Wartamabe also noted that anomalous splitting of the arachnoid membrane occurs during the process of the complex folding of the primitive neural tube and the formation of normal subarachnoid cisterns (30). Smith has suggested that arachnoid cysts enlarge as a result of either a secretory mechanism of the arachnoid cells or as a consequence of a “ball-valve” mechanism, allowing fluid into the cyst but blocking its outflow (35). Other authors have postulated that subtle osmotic gradients contribute to the growth of arachnoid cysts (26,34). Some authors attribute cyst enlargement to a slit-valve mechanism (6,11,32,34). Caemaert et al. were the first to endoscopically observe a slit-valve in a suprasellar arachnoid cyst (6). Schroeder and Gaab clearly identified a slit-valve-like structure formed by an arachnoid membrane around the basilar artery that opened and closed with arterial pulsation (33).

The diagnostic information obtained by CT and/or magnetic resonance imaging (MRI) is helpful in deciding about therapeutic management by a stereotactic intervention. CT scans depict arachnoid cysts as non-enhancing extra-axial hypodense lesions with sharp borders (21). On MRI the cysts have low signal intensity on T1-weighted sequences and a high signal on T2-weighted images with the cyst fluid having the same signal characteristics as CSF. There is no enhancement of any part of the lesion after contrast agent administration (26). The differential diagnosis of suprasellar cysts includes cystic craniopharyngiomas, Rathke’s cleft cysts, epidermoid tumours, empty sella, cystic gliomas, cystic pituitary adenomas, inflammatory cysts, and ependymal cysts (8,18,22,24). These entities are commonly of congenital origin and asymptomatic. Choi and Kim postulated that head trauma in infancy or perinatal trauma may contribute to the development of arachnoid cysts, including those of the suprasellar region, in some cases (8).

Suprasellar arachnoid cysts are uncommon lesions and their optimal management is still controversial (9,20). The aim of surgical treatment is to normalize CSF flow by establishing a permanent communication between the cyst cavity and the intraventricular or/and subarachnoid space. Various treatment options have been advocated. Different surgical measures are available and should be selected on an individual basis. The surgical options include cyst aspiration, wall fenestration, wall excision, cystoventriculostomy, and cystoperitoneal shunting (3,9,23,25-29,32).

Repeat operations are often required regardless of the initial surgical approach (28). The complication rates reported after surgery for arachnoid cysts range from 15 to 58% (1,26). Reported complications include seizures, neurological injury, infection, intracystic haemorrhage, catheter occlusion, cyst reaccumulation, subdural hygroma, hyponatraemia, and failure to alleviate the symptoms that led to the intervention (16,26).

Cyst fenestration for internal drainage (31) performed via an open craniotomy has a 60% recurrence rate with significant risks of mortality and morbidity (10,36). The establishment of a single CSF space, by the surgical creation of a communication between the cyst and the ventricular system or the basal cisterns, appears to offer the best chance of success in the treatment of suprasellar arachnoid cysts (12).

Neuroendoscopic cyst fenestration or cystoventriculostomy has the advantage of reducing complications related to brain shifts, which occur with open craniotomy. However, reports in the literature suggest that there is only a moderate or slight reduction in cyst volume following this procedure while the artificially created communication tends to close with time. The need for a second procedure after cystoventriculostomy is reported to be as high as 38% (2,13,14,28,36,38,41). A summary of published cases of suprasellar arachnoid cysts, their therapeutic management, complications, and outcome is given in the table (Table I).

Precocious puberty is rarely the presenting sign of an arachnoid cyst. It is an uncommon presentation
### Table I: Published series of suprasellar arachnoid cysts, therapeutic approaches, and outcome.

<table>
<thead>
<tr>
<th>Authors (Year)</th>
<th>Number of patients</th>
<th>Therapeutic approach</th>
<th>Outcome and complications</th>
<th>Follow-up (years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Behrans P et al. (1993)</td>
<td>5</td>
<td>Stereotactic cystoventriculostomy</td>
<td>2 CVA shunt recurred</td>
<td>3</td>
</tr>
<tr>
<td>Buxton N et al. (1999)</td>
<td>3</td>
<td>Endoscopic fenestration</td>
<td>-</td>
<td>2.5</td>
</tr>
<tr>
<td>Caemaert J et al. (1992)</td>
<td>4</td>
<td>Endoscopic cystoventriculostomy and cystocisternostomy</td>
<td>1 re-operation</td>
<td>2</td>
</tr>
<tr>
<td>Charalampaki P et al. (2005)</td>
<td>13 (7 had shunt before)</td>
<td>Cystoventriculostomy and cystocisternostomy</td>
<td>Shunt-free 1 meningitis, 1 psychomotor disturbance</td>
<td>0.5-6</td>
</tr>
<tr>
<td>D’Angelo V et al. (1999)</td>
<td>1</td>
<td>Stereotactic cystoventricular shunt</td>
<td>-</td>
<td>2.3</td>
</tr>
<tr>
<td>Decq P et al. (1996)</td>
<td>2</td>
<td>Endoscopic ventriculostomy or ventriculocystocisternostomy</td>
<td>1 re-operation</td>
<td>1.5-2</td>
</tr>
<tr>
<td>Desai KI et al. (2003)</td>
<td>3</td>
<td>Craniotomy cystocisternostomy</td>
<td>-</td>
<td>3.1</td>
</tr>
<tr>
<td>DiRocco C et al. (2005)</td>
<td>2</td>
<td>Endoscopic cystoventriculostomy</td>
<td>-</td>
<td>3</td>
</tr>
<tr>
<td>Golash A et al. (2001)</td>
<td>1</td>
<td>Endoscopic ventriculostomy and cystocisternostomy</td>
<td>-</td>
<td>2</td>
</tr>
<tr>
<td>Gupta SK et al. (1999)</td>
<td>2</td>
<td>Craniotomy cystocisternostomy VPS</td>
<td>Shunt revision and craniotomy</td>
<td>?</td>
</tr>
<tr>
<td>Hofmann HJ et al. (1982)</td>
<td>8</td>
<td>5 VPS 2 craniotomy (cystocisternostomy) 1VPS+craniotomy</td>
<td>3 VPS 2 craniotomy needed 1 hypopituitarism</td>
<td>?</td>
</tr>
<tr>
<td>Kirollos RW et al. (2001)</td>
<td>10</td>
<td>(previous 3 cyst or VPS removed) 7 Neuroendoscopy fenestration cystoventriculostomy and cystocisternostomy 3 cystoventriculostomy combined with third ventriculostomy</td>
<td>-</td>
<td>1.5</td>
</tr>
<tr>
<td>Meyer FB et al. (1987)</td>
<td>13</td>
<td>Csym excision, fenestration (transsphenoidal)</td>
<td>1 death (meningitis) 2 hormone deficiency</td>
<td>4.5</td>
</tr>
</tbody>
</table>
with only sporadic cases reported in the literature (4,7,15,28). Puberty is a complex developmental process culminating in sexual maturity. This transitional period begins in late childhood and is characterized by maturation of the hypothalamic-pituitary-gonadal axis, the appearance of secondary sexual characteristics, acceleration of growth, and, ultimately, the capacity for fertility (19). Although 10 to 40% of patients with suprasellar arachnoid cysts have been reported to experience signs and symptoms of precocious puberty despite surgical treatment, the origin of this condition is unclear (38). It is believed that pulsatile damage to the hypothalamus before treatment may in some way enhance the onset of puberty in children (36). Some patients with suprasellar arachnoid cysts presenting with precocious puberty have shown good response to surgical treatment (4,15,39).

In the case reported here, stereotactic cystoventriculostomy by catheter implantation (for drainage) led to clinical recovery by stabilizing the size of the suprasellar arachnoid cyst and preventing the development endocrinologic problems (3). Implantation of a reservoir may be useful in patients who are candidates for repeat aspiration.

**CONCLUSION**

In the case presented, stereotactic cystoventriculostomy by catheter implantation for establishment of an internal drainage may lead to complete recovery with permanent resolution of clinical symptoms and prevented the development of

<table>
<thead>
<tr>
<th>Murakami M et al. (2003)</th>
<th>1</th>
<th>Transsphenoidal excision</th>
<th>Re excision via craniotomy</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pierre-Kahn A et al. (1990)</td>
<td>20</td>
<td>1 Transcranial 1 VPS 15 Percutaneous ventriculocystostomy (in 2 case additionally cystoventriculostomy) 3 no surgery</td>
<td>1 SAH VPS 1 Neurological complication 2 cysts removed with subsequent VPS</td>
<td>6</td>
</tr>
<tr>
<td>Raimondi AJ (1980)</td>
<td>5</td>
<td>4 craniotomy (resection) plus VPS 1 EVD and VPS</td>
<td>1 Repeat VPS 1 CPS (cyst recurved) plus VPS</td>
<td>0.5-1</td>
</tr>
<tr>
<td>Rappaport ZH et al. (1993)</td>
<td>5</td>
<td>cystoventriculostomy cystocisternostomy VPS</td>
<td>1 death (sepsis) bilateral extradural fluid collections secondary operation. After C VPS. 3 CPS, VPS Cyst fenestration basal system Shunt infection, dysfunction</td>
<td>1</td>
</tr>
<tr>
<td>Schroeder HD et al. (1997)</td>
<td>1</td>
<td>Endoscopic cystoventriculostomy</td>
<td>-</td>
<td>?</td>
</tr>
<tr>
<td>Sood et al. (2005)</td>
<td>8 (pre-existing shunt: 4)</td>
<td>Endoscopic fenestration cystocisternostomy and coagulation</td>
<td>- 5 independent shunt</td>
<td>4.5</td>
</tr>
</tbody>
</table>

_CPS:_ Cysto-peritoneal shunt, _CVA:_ Cysto-venriculoatrial shunt, _VPS:_ Ventriculo-peritoneal shunt, _EVD:_ External ventricular drainage, _SAH:_ Subarachnoid haemorrhage.
endocrinologic problems. This procedure obviated the need for open cyst fenestration or cysto-peritoneal shunting. Our results show that early stereotactic cystoventriculostomy is useful to prevent expansion of a suprasellar arachnoid cyst and progression of endocrine symptoms. The stereotactic approach could be a minimal invasive therapeutic option in treating suprasellar arachnoid cysts.

Acknowledgement
Aslan Güzel is supported by TUBITAK (Scientific and Technical Research Council of Turkey) and DFG (Deutsche Forschungsgemeinschaft).

REFERENCES