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Intracranial araknoid cysts

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ABSTRACT

Intracranial arachnoid cysts, known as leptomeningeal cysts, are benign lesions that develop after the collections of cerebral spinal fluid (CSF) wrapped by a wall of collagen and arachnoid cells congenitally (real) or acquired (sourced by bleeding, tumor, trauma, or meningitis). Most of these cysts are asymptomatic. Symptomatic cysts are detected in the first 20 years of life, and three-quarters of these cysts become symptomatic during childhood. Typical symptoms are headache, nausea, vomiting, epilepsy, sudden loss of consciousness depending on cyst rupture or bleeding, macrocephaly, hydrocephalus, endocrinological disorders, psychiatric disorders, and focal findings of the lesion occupying space within the head (weakness, cranial neuropathy). They are usually diagnosed using radiological imaging methods including X-Ray, transfontanel ultrasonography, computed tomography, magnetic resonance imaging, and scintigraphy. Surgical treatment is not recommended for arachnoid cysts unless they cause a mass effect or symptoms and they are annually follow-up with CT or MR scan is an appropriate method in asymptomatic patients. Interestingly, no standard surgical intervention method has been recommended for the cyst. Therefore, the patient's clinical and radiological findings, age, size, and localization of the cyst are still the most important factors in the decision-making of the surgical intervention method. Surgical intervention methods include cyst resection and fenestration of the cyst into the basal cisterns by craniotomy; endoscope-assisted cyst fenestration through the burr hole; cystoperitoneal shunt or ventriculoperitoneal shunt.

Keywords: Arachnoid cyst, cerebral, management

INTRODUCTION

Intracranial arachnoid cysts, known as leptomeningeal cysts, are benign lesions (1, 2). In all radiological imaging methods, the cyst filled with clear, colorless fluid has almost the same density as CSF and is often diagnosed incidentally following trauma or headache (3). The majority of arachnoid cysts are located supratentorial and are usually seen in the middle fossa. Less frequently, they may be seen in the suprasellar region, cerebral convexities, cerebellopontine angle, quadrigeminal cisterns, and cisterna magna (4).

The signs and symptoms of arachnoid cysts vary according to their size and intracranial location. Most patients with small arachnoid cysts are usually symptomatic, and they can be followed up conservatively. Unfortunately, the natural history of asymptomatic arachnoid cysts is not yet fully understood. However, an increase in cyst size may be detected in 2.5% of the patients during their follow-up period, and this enlargement may increase the intracranial pressure and may produce some symptoms including headache, nausea, vomiting, focal neurological deficits, or hydrocephalus (5). On the other hand, larger cysts in 5.3% of patients can be seen and these cysts may however exert massive compression on neurovascular structures and cause neurological symptoms including headache (66% of the patients), dizziness, nausea, vomiting, mood deterioration, mental status changes, ataxia, seizures, and hearing loss, and therefore surgical intervention may require for these symptomatic patients (4, 6, 7, 8). Depending on the location of the arachnoid cyst and the experience of the surgeon, different surgical techniques such as microsurgery, endoscopic intervention, and cystoperitoneal shunt can be performed in the surgical treatment of symptomatic arachnoid cyst patients (9).

EPIDEMIOLOGY

Approximately 60% of arachnoid cysts are seen in the pediatric age group (10). Recent studies have shown that the prevalence of arachnoid cysts is 2.6% in children and 1.4% in adults (4, 11). Population studies estimate that arachnoid cysts are mostly detected in the left hemisphere in the pediatric population and constitute at least 1% of intracranial space-occupying lesions and they are seen between 0.3% and 2.6% of the population while they are

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found in 5 of 1000 autopsies. Its incidence is higher in men than in women (male / female = 4/1) (1, 4).

Etiology/ Pathogenesis

Arachnoid cysts that can be congenital or acquired account for 1% of all intracranial space-occupying lesions (12). It has been suggested that congenital cysts arise from the separation of arachnoid membranes during the fetal period and that they occur after the accumulation of cerebrospinal fluid in these spaces. However, acquired cysts are rare and may usually develop after trapping of the CSF in the arachnoid scar tissue followed by trauma, intracranial surgery, intracranial bleedings, tumors, infections, and meningitis (1, 5). Acquired arachnoid cysts. The accumulation of hemosiderin in the cyst wall suggests the traumatic origin (13, 14). Cyst walls may contain trapped choroid plexus remnants, arachnoid granules, or subdural neuroepithelium (3).

Except for intrasellar arachnoid cysts, all of them are adjacent to the arachnoid cisterns. They are most commonly seen in the Sylvian fissure (49%), cerebellopontine angle (11%), supracollicular region (10%), vermian (9%), sellar and suprasellar regions (9%), interhemispheric (5%), cerebral convexity (4%), clival regions (3%) and very rarely quadrigeminal cistern (15). Arachnoid cysts in the parasellar region are most common in children and most of them may cause hydrocephalus (3). Galassi et al. (16) divided Sylvian arachnoid cysts into 3 types as follows:

- Type 1: Small, lenticular, biconvex collections located at the anterior pole of the middle fossa, just behind the sphenoid ridge. It is mostly asymptomatic and does not require intervention. Annual follow-up is required.
- Type 2: They are larger, triangular, or quadrangular cysts extending from the proximal to the middle of the Sylvian fissure and may require treatment if symptomatic.
- Type 3: Large, round cysts that fill the entire Sylvian fissure. Typically, they may present with significant mass effect and midline shift, thinning of the middle fossa cranial bones, or opening of cranial sutures in younger children, and they require surgical intervention in patients with mass effect and midline shift.

On the other hand, quadrigeminal arachnoid cysts are very rare and classified into three types in the literature (17):

- Type I includes cysts with supratentorial and infratentorial extensions.
- Type II cysts with infratentorial extension (supracerebellar or supra-retrocerebellar).
- Type III includes cysts extending laterally towards the temporal lobe.

Clinical Findings

The natural course of arachnoid cysts remains unclear, however, over the years, the vast majority remains unchanged in the radiological imaging methods performed during the follow-up periods. Unfortunately, some cysts may progress and have an increasing mass effect on adjacent neural structures. These cysts are detected in the first 20 years of life, and three-quarters of these cysts become symptomatic during childhood (3, 11). Arachnoid cysts may sometimes rupture spontaneously or after minor trauma, wrestling, riding the Ferris wheel, or roller coaster. They may cause increased intracranial pressure due to either subdural hygromas or acute and chronic subdural hemorrhages (3, 18, 19, 20). Biomechanical studies show that bleeding occurs due to the force transmitted to the outer edge of the cyst by trauma, compared to the normal brain. Depending on this hypothesis, the increase in size may be considered to increase the risk of bleeding. Cress et al showed in 2013 that 6% of arachnoid cysts may have a bleeding risk after 5 years. However, studies trying to reveal the relationship between arachnoid cysts and bleeding have not provided sufficient data to require people with arachnoid cysts to limit sports or heavy activity (21, 22).

Arachnoid cysts can cause various symptoms depending on location and size. Typical symptoms are headache, nausea, vomiting (related to the increased intracranial pressure), dizziness, epilepsy, mental status changes and sudden loss of consciousness (depending on cyst rupture or bleeding), ataxia, macrocephaly, hydrocephalus, endocrinological disorders, psychiatric disorders (mood deterioration, etc.), focal findings of the lesion occupying space within the head (weakness, cranial neuropathy, hearing loss, etc.) (1). In a study involving 485 patients (116 pediatrics, 369 adults), it was revealed that 92.4% of adult patients with arachnoid cysts and 72.4% of pediatric patients were asymptomatic. Headache was reported as the most common symptom (60.7%) in 28 symptomatic adults. In adult patients, cranial nerve dysfunctions (39.3%), nausea-vomiting (25%), gait disturbance (14.3%), hemiparesis (14.3%), dizziness (7.1%), dysphasia (%) 3.8) and epileptic seizure (3.8%) were detected. It was reported that the most common symptom in 32 symptomatic pediatric patients was headache (43.8%), as in adults. In symptomatic pediatric cases, macrocephaly (28.1%), nausea-vomiting (18.8%), gait disturbance (9.4%), dizziness (6.3%), papilledema (6.3%), hemiparesis (3.1%), impaired consciousness (3.1%), developmental delay (3.1%) and swelling in the skull (3.1%) were found (Table 1) (23).

Supratentorial Arachnoid Cysts

Sylvian fissure arachnoid cysts: They locates at the anterior pole of the middle fossa, just behind the sphenoid ridge

(**Figure 1**). The most common symptom is a unilateral supraorbital or temporal headache, which may be exacerbated by physical effort and breath-holding. Headaches may rarely be associated with other signs and symptoms of increased intracranial pressure such as nausea, vomiting, and papilledema. Other associated symptoms are focal, complex-partial, or generalized epileptic seizures. Rare symptoms include headache during chewing, proptosis, temporal bone deformity, speech disorder, and tic-like movement disorders (4, 21, 22).

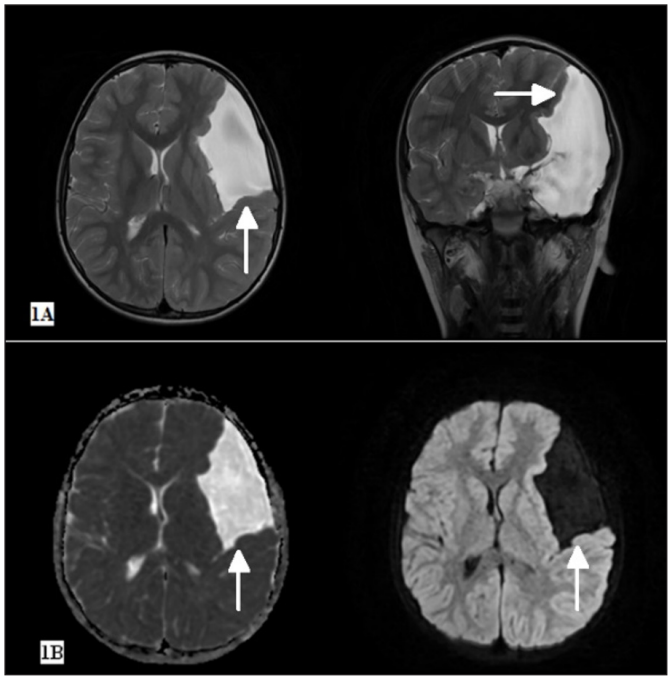

Figure 1. Axial and coronal T2-weighted (1A, 1B); ADC (apparent diffusion coefficient) (1C); and DWI (diffusion-weighted imaging) (1D) MR scan indicate an arachnoid cyst located in the Sylvian region

Table 1. The table demonstrates the symptoms in patients with symptomatic arachnoid cyst (23).

Symptoms	Pediatric patients N (%)	Adult patients N (%)
Headache	14 (43.8)	17 (60.7)
Nausea/ vomiting	6 (18.8)	7 (25.0)
Cranial nerve dysfunction	2 (6.3)	11 (39.3)
Macrocephaly	9 (28.1)	0 (0.0)
Gait unsteadiness	3 (9.4)	4 (14.3)
Hemiparesis	1 (3.1)	4 (14.3)
Dizziness	2 (6.3)	2 (7.1)
Papilledema	2 (6.3)	0 (0.0)
Cognitive decline	1 (3.1)	0 (0.0)
Developmental delay	1 (3.1)	0 (0.0)
Dysphasia	0 (0.0)	1 (3.8)
Seizures	0 (0.0)	1 (3.8)
Skull swelling	1 (3.1)	0 (0.0)

Studies have shown that there is a relationship between bilateral Sylvian arachnoid cysts and glutaric aciduria type I (**Figure 2**). It is very rare to find cysts both in the supratentorial and infratentorial regions and to detect multiple or bilateral cysts. Rarely, familial cases have been reported (24).

Parasellar arachnoid cyst: The most common parasellar region arachnoid cysts occur within the suprasellar cistern. About 50% of suprasellar cysts are diagnosed before 5 years of age, and 20% before 1 year of age. The most common symptom in arachnoid cysts of this region is hydrocephalus, which develops as a result of the cyst's elevation of the third ventricle and the disruption of CSF passage through the foramen of Monro. Other symptoms include endocrine dysfunction, visual impairment, ataxia, visual field defects, opisthotonus, and bobblehead doll syndrome. 10-60% of patients with suprasellar arachnoid cysts may present with endocrinologic disorders. Short stature can be seen in children due to precocious puberty or growth hormone deficiency. More than half of children with suprasellar cysts have mental retardation and they fail the neurocognitive tests (25, 26).

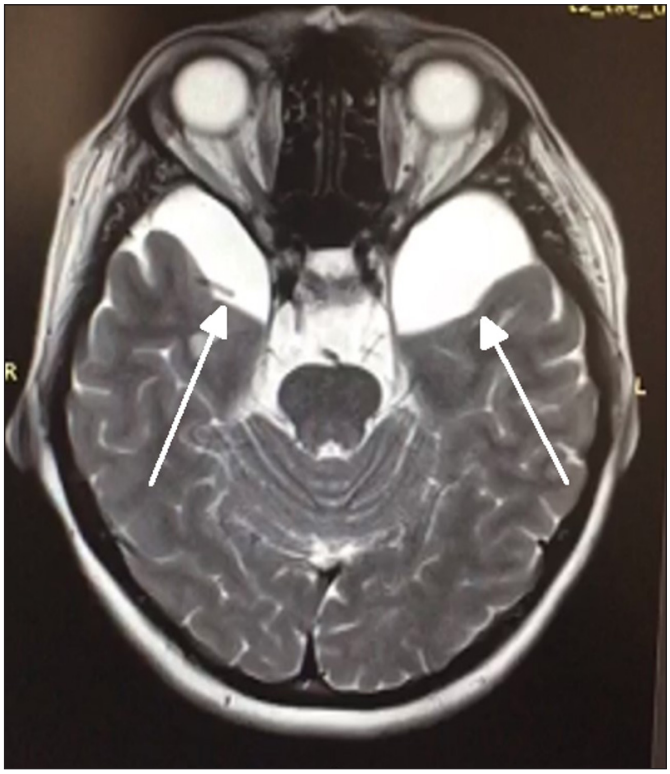

Figure 2. Axial T2-weighted MR image demonstrates the bilateral temporal arachnoid cyst associated with glutaric aciduria type 1

Intrasellar arachnoid cysts are rare in children and more often present with symptoms in the fourth or fifth decade of life. The most common symptom is headache, but bitemporal hemianopsia and endocrinologic disorders have also been reported. Most of these cysts tend to cause hemorrhages and are difficult to distinguish from Rathke's cleft cysts (27).

Convexity arachnoid cyst: Arachnoid cysts of cerebral convexities usually present with headaches, seizures, or both. Age, size, and location typically determine its

clinical manifestations. Small cysts may focally compress the adjacent brain and thin the bone whereas large cysts may cause asymmetrical enlargement of the calvarium, suture diastasis, and hemispheric distortion (28).

Interhemispheric arachnoid cyst: They are often associated with agenesis of the corpus callosum. Their clinical signs are mostly in the form of macrocephaly and asymmetrical enlargement of the calvarium. They tend to be associated with increased intracranial pressure, growth retardation, hemiparesis, and epilepsy (29).

Infratentorial Arachnoid Cysts

Posterior fossa arachnoid cysts: They are less common than supratentorial cysts (**Figure 3**). Unlike the Dandy-Walker malformation, posterior fossa arachnoid cysts are extra-axial and separate from the CSF circulation. The most common symptoms are a headache at the vertex and suboccipital region accompanied by hydrocephalus secondary to fourth ventricle compression. There may be macrocrania with asymmetry of the skull base. Teenage patients may describe a headache that increases with sports activities, similar to the symptoms of Chiari malformation Type I (30).

Cerebellopontine angle arachnoid cyst: They can cause tinnitus, vertigo, facial weakness, facial sensory loss, hearing loss, torticollis, difficulty swallowing, or ataxia (**Figure 4**). It has also been reported to cause trigeminal neuralgia and hemifacial spasm (31-33).

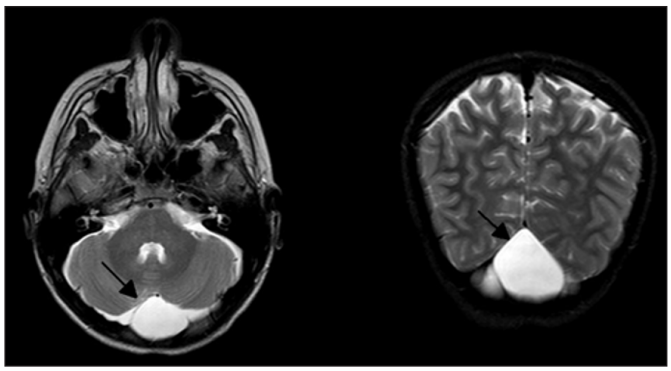

Figure 3. Axial and coronal T2-weighted MR images show an infratentorial arachnoid cyst

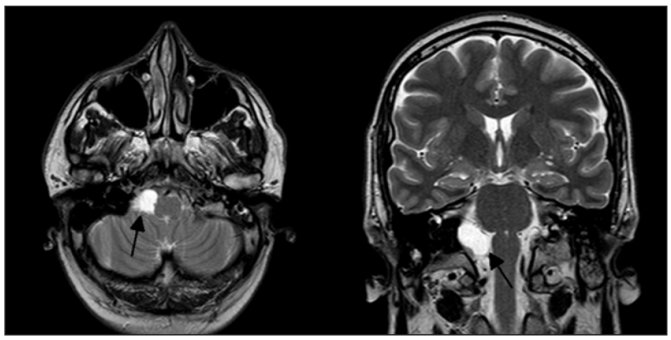

Figure 4. Axial and coronal T2-weighted MR images show a cerebellopontine angle arachnoid cyst

Quadrigeminal arachnoid cysts: They may cause hydrocephalus by applying pressure on the Aqueductus Sylvii in the early period. They must be surgically treated as symptoms include macrocrania, headaches, vomiting, lethargy, papilledema, upward gaze disorder, and other ocular disorders (17, 28).

Radiological Presentation

X-ray: It can reveal thinning of the calvarium bone adjacent to the cyst, an increase in the size of the middle fossa, and elevation or displacement of the sphenoid wing (1).

Trans-fontanel USG: It can demonstrate supratentorial arachnoid cysts, hydrocephalus, and the mass effect of the cyst on the cerebral tissue in newborn patients (1).

Computed tomography (CT): It can show an extra parenchymal lesion with soft borders, no calcifications, smooth edges, and the same density as CSF. It may also distinguish chronic processes if it is accompanied by enlargement of the adjacent bone tissue with ventriculomegaly (64% of supratentorial cysts, 80% of infratentorial cysts) (1).

Magnetic resonance (MR): Using the MR scan, the relationship of the arachnoid cyst with the basal cisterns can be demonstrated in three dimensions. Most arachnoid cysts appear hyperintense on T2-weighted imaging and hypointense on T1-weighted imaging like CSF. This evaluation can easily detect mass effects on adjacent structures and associated anomalies. Constructive interference in the steady-state (CISS) sequence of the MR scan may provide high-resolution imaging of the arachnoid cyst wall. Furthermore, the vascular anatomy surrounding the cyst can be visualized quite well on the MRI with gadolinium. The higher resolution of MRI allows better detection of smaller cysts and cysts adjacent to bony structures, particularly in the sellar region. Unlike diffusion restriction in epidermoid tumors, which is included in the differential diagnosis of arachnoid cysts in diffusion-weighted MR, suppression is not observed in arachnoid cysts (3,14,34). CT and MR cisternography that is performed with an intrathecal contrast material can distinguish whether there is a flow of the CSF into the cyst (i.e. communicated or non-communicated arachnoid cyst). On the other hand, with the use of cine-phase contrast MR, this distinction can be made non-invasively. CSF flow-sensitive MR sequences and cine MR sequences can generally show the connection between intracranial arachnoid cyst and CSF spaces in 95% of patients (35).

Treatment Modalities

Surgical treatment is not recommended for arachnoid cysts unless they cause a mass effect or symptoms. Annual

follow-up with CT or MR scan is an appropriate method in asymptomatic patients. The most important step in the decision-making of the surgical treatment of the cyst is to determine the correct indication (**Figure 5**). The patient's clinical and radiological findings, age, size, and localization of the cyst are the most important factors in the decision-making of the surgical intervention method. A definite and clear relationship should be established between the patient's symptoms and lesion. No standard surgical intervention method has been recommended for the cyst. The surgical treatment of arachnoid cysts aims to decompress the cyst and establish a connection between normal and pathological CSF spaces (23).

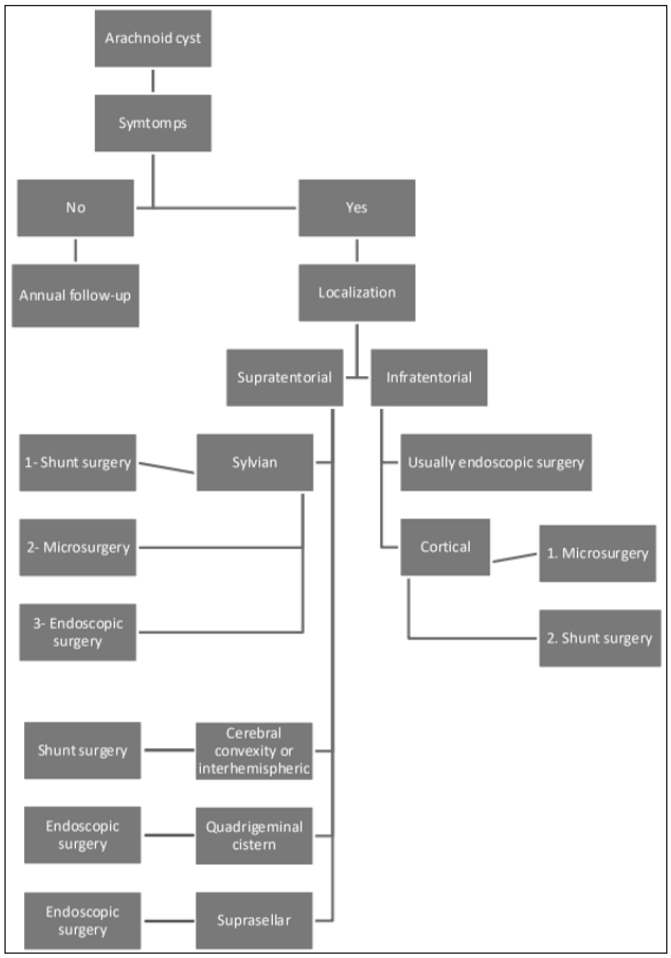

Figure 5. Graphic shows the follow-up and treatment algorithm of intracranial arachnoid cysts

For this purpose, the following surgical treatment methods can be applied (23,28,36):

- Cyst resection and fenestration of cyst to the basal cisterns with craniotomy
- Endoscope-assisted cyst fenestration through a burr-hole (such as ventriculocystostomy; ventriculocystocisternostomy)
- Cystoperitoneal shunt or ventriculoperitoneal shunt

Surgical treatment with an appropriate method is beneficial in patients with symptomatic arachnoid cysts (36). For example, whether or not to treat Sylvian-located arachnoid cysts or by which method is still unclear. Surgical indications in Galassi Type 1 and Type 2 arachnoid cysts have not been demonstrated. However, studies show that the success rates of cystoperitoneal shunt surgery (96.8%) and microsurgery and fenestration (88%) are similar and superior to the endoscopic (77%) method in Sylvian-located arachnoid cysts (14, 37, 38). On the other hand, endoscopic fenestration is quite successful and advantageous compared to other methods for suprasellar arachnoid cysts (14). The success rate of all methods is similar in the surgical treatment of arachnoid cysts located in the quadrigeminal cistern, however, the endoscopic method is usually recommended for these cysts because they are deeply localization and carries surgical risks (37). Besides, the clinical success rate of shunt surgery is 100% for the arachnoid cysts that are located in the cerebral convexity and interhemispheric region (14). Finally, the success rates of all surgical intervention methods for posterior fossa cysts are very similar to each other. Although the most appropriate method is endoscopic fenestration, cortical cysts in this region can be treated using the microsurgery method, fenestration method, or shunt (37, 38). However, some retrospective studies have reported that 86% of patients who refused surgical treatment despite being symptomatic might be recovered clinically without surgical intervention (39). It has been reported that endoscopic techniques such as cyst fenestration and removal via the suboccipital supracerebellar approach, lateral ventricle cystostomy, and third ventricle cystostomy should be preferred as a minimally invasive surgical treatment in the treatment of these cysts and that 78-93% of patients treated with these techniques are independent of a shunt (40, 41, 42).

The most common complications of surgical treatment are recurrence, subdural hematoma, subdural hygroma, CSF leakage, intralesional and/ or intraventricular hemorrhage, seizures, wound infections, meningitis, and hormonal deficiencies (10, 23). Surgical complications such as meningitis, shunt dysfunction, and cranial nerve palsy may occur in patients undergoing shunt surgery (10, 43).

CONCLUSION

Today, it is still recommended that conservative follow-up of asymptomatic arachnoid cyst patients is appropriate, however surgical treatment should be applied in symptomatic patients. In surgical treatment, marsupialization of the cyst with microsurgery, cystoperitoneal shunt placement, and endoscopic treatment methods, which are increasingly preferred, are applied. The patient's clinical and radiological findings, age, size, and localization of the cyst are proposed as

the most important factors in the decision-making of the surgical intervention method. Therefore, while neuroendoscopic interventions seem to be the most appropriate surgical treatment for arachnoid cysts in the suprasellar and quadrigeminal regions, microsurgical fenestration is still recommended for interhemispheric cysts.

ETHICAL DECLARATIONS

Referee Evaluation Process: Externally peer-reviewed.

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