Intracranial Arachnoid Cysts
Clinical Features and Management of 35 Cases and Review of the Literature

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Abstract: The purpose of this study is to evaluate the distribution, clinical features, and treatment modalities of arachnoid cysts (ACs) in our clinic. The study was carried out between January 1, 1996 and November 31, 2002 at the Neurosurgery Department of Çukurova University Hospital, Adana, Turkey. Thirty-five patients (13 female and 22 male) with ACs demonstrated by cranial magnetic resonance imaging or computed tomography during this period were enrolled and discussed in this study. The middle cranial fossa was the most common location of ACs; ACs were located in the middle cranial fossa in 22 (62.9%) cases, followed by the cerebellopontine angle in 4 (11.4%) cases, the cerebral convexity in 3 (8.6%) cases, the suprasellar region in 2 (5.7%) cases, the quadrigeminal cisterns in 2 (5.7%) cases, and the retrocerebellar region in 2 (5.7%) cases. All cysts were unilateral: 25 (71.4%) were located on the left side, and 10 (28.6%) were located on the right side. The mean age at onset of clinical manifestations was 3 years, 6 months (range: 1 month to 12 years). The mean age at diagnosis was 8 years (range: 1–32 years). The most frequent clinical features on presentation were epileptic seizures (42.9%) and headache (28.6%). A few patients (9 [25.7%]) underwent surgery. Surgical intervention with microsurgical excision and fenestration was performed in 7 patients, and cystoperitoneal shunting was performed in 2 patients. The major indications for surgery of ACs are intractable seizures, intracranial hypertension, and compression of neuronal tissues. Headache only is not a surgical indication. Microsurgical excision and fenestration are safe and effective for the surgical treatment of ACs.

Key Words: arachnoid cyst, intracranial, sex, surgery, treatment

From postinflammatory accumulation of CSF in the subarachnoid space in patients with head injury, intracranial hemorrhage, or infection.4,6,8 In many instances, the recognition of an AC is an incidental finding at postmortem examination or on neuroimaging studies in symptomatic patients examined for head injury or infants with macrocrania.2,3,6,9

ACs are relatively rare. The reported incidence accounts for only 1% of all intracranial space-occupying lesions.1–6,10–17 An apparent increase in frequency and a shift in age distribution toward the first years of life have been described in recent years, likely reflecting the widespread use of computed tomography (CT), magnetic resonance imaging (MRI), and ultrasound scanning.1,6,12

We present 35 patients with intracranial ACs. The distribution, clinical features, and treatment modalities are discussed in this review.

PATIENTS AND METHODS
The study was carried out between January 1, 1996 and November 31, 2002 at the Neurosurgery Department of Çukurova University Hospital, Adana, Turkey. Thirty-five patients with ACs demonstrated by cranial MRI or CT (13 MRI cases, 15 CT cases, and 7 cases in which both CT and MRI were performed) during this period were enrolled in this study. These neuroimaging studies were performed because of head trauma or the presence of clinical manifestations such as headache and seizure. A complete electroencephalogram (EEG) with awake and sleep traces was performed in 23 cases.

Intractable seizures, intracranial hypertension, and specific signs resulting from compression of neuronal tissues were accepted surgical indications. All patients were followed for minimum of 6 months after surgery, and all complications were recorded.

The complete medical records of the patients were reviewed, and data were extracted. The data collected included information on demographics, localization, symptoms, course of treatment, complications, and outcome.

RESULTS
During the study period, 35 patients were investigated. Of these patients, 13 (37.1%) were female and 22 (62.9%)
were male. The mean age at diagnosis was 8 years (range: 1–32 years). The mean age at onset of clinical manifestations was 3 years, 6 months (range: 1 month to 12 years). The middle cranial fossa was the most common location of ACs; ACs were located in the middle cranial fossa in 22 (62.9%) cases, followed by the cerebellopontine angle (CPA) in 4 (11.4%) cases, the cerebral convexity in 3 (8.6%) cases, the suprasellar region in 2 (5.7%) cases, the quadrigeminal cisterns in 2 (5.7%) cases, and the retrocerebellar region in 2 (5.7%) cases. All cysts were unilateral: 25 (71.4%) were located on the left side, and 10 (28.6%) were located on the right side.

The presenting clinical features were epileptic seizures in 15 (42.9%) cases; headache in 10 (28.6%) cases; vomiting in 3 (8.6%) cases; temporal bulging and macrocrania in 2 (5.7%) cases; and peripheral facial palsy, proptosis, and nystagmus in 1 (2.9%) case. Headache was the sole clinical symptom in 5 (14.3%) cases and was treated with analgesics.

The ACs appeared hypointense on T1-weighted sequences and hyperintense on T2-weighted images on MRI (Figs. 1–3). In all cases, the cyst fluid produced the signal characteristics of CSF. There was no enhancement of the cyst contents or cyst wall after the intravenous administration of gadolinium. CT also demonstrated an extra-axial hypodense lesion with sharp borders. According to the classification system of Galassi et al., most (16 [72.2%]) of the middle fossa cysts were type II or smaller. The diagnosis was confirmed by tissue biopsy in surgical patients with microsurgical excision and fenestration (Fig. 4).

Background EEG activity was in the normal range in 8 cases. Focal epileptiform discharges, mainly arising from the central or temporal region, were found in 14 of 22 middle cranial fossa ACs, and an origin from the frontal region was found in 1 of 3 cerebral convexity ACs. We also found that the frequency of seizures varied and was independent of cyst size. The response to antiepileptic drugs was variable. Seven of 15 (46.6%) patients with epileptic seizures receiving monotherapy were seizure-free. The remaining 8 (53.4%) patients had intractable seizures despite numerous adjustments in antiepileptic drugs.

A few patients (9 [25.7%]) underwent surgery. Other patients were followed clinically and radiologically with MRI or CT and did not require surgical therapy. Intractable seizure was the major indication for surgery (8 [88.8%]). Only 1 (11.2%) patient was operated on for peripheral facial palsy. Surgical intervention with microsurgical excision and fenestration was performed in 7 patients, and cystoperitoneal shunting was performed in 2 patients for treatment of ACs. No patients were operated on for headache alone.

The mean period of monitoring was 30 months (range: 6 months to 5 years). Microsurgical excision and fenestration were performed in 6 intractable seizure patients, which resulted in a reduction in cyst size and the disappearance of epileptic seizures in all patients. Proptosis and nystagmus, together with intractable seizures, were not improved in 1 patient, however. Microsurgical excision and fenestration were also performed in peripheral facial palsy patients, but clinical improvement was not seen. Cystoperitoneal shunting was performed only in 2 intractable seizure patients, which resulted in a partial reduction of seizure frequency. These patients could not discontinue antiepileptic drugs, however. Surgical complications were not encountered in any surgical patients. At long-term follow-up, all patients are able to carry on a normal life.

**DISCUSSION**

The cause of ACs is unknown. Nevertheless, there are 3 postulated mechanisms by which ACs form: 1) congenital,
whereby abnormal CSF flow occurs through the leptomeninges, leading to the development of a diverticulum in the arachnoidal membrane; 2) an inflammatory reaction caused by infection, leading to the formation of ACs; and 3) a history of trauma, particularly during birth.1,6–10,12–17 Our patient had no previous history to suggest a cause other than congenital.

ACs are nearly always sporadic and single. Most ACs are now detected in the first 2 decades of life. In a worldwide literature survey of patients diagnosed using imaging techniques, these cysts were demonstrated to occur nearly twice as often on the left side than on the right side, and there was also a marked male predominance (3:1 male-to-female ratio) as in our series.2,18

Intracranial ACs are apparently not distributed randomly within the neocranium. In some reports, 90% of ACs were found in supratentorial locations and 10% were found in the posterior fossa. The most common supratentorial site of ACs is the middle cranial fossa, particularly the temporosylvian area.3–7,10,11 Rengachary16 and Rengachary and Watanabe14 compiled patient data from previous publications and found that the middle cranial fossa is the site of nearly half (49%) of all intracranial ACs. Sakai et al19 also found that 65% of ACs were located in the middle cranial fossa. The CPA is the second most common location for ACs, with an approximately 10% frequency.2,4,5,10,11,18,20 Other sites include the posterior fossa, suprasellar cistern, collicular region, quadrigeminal plate, and cerebral or cerebellar hemispheres.3,10,21–23

FIGURE 2. Axial magnetic resonance imaging (MRI) scans showing a left cerebellopontine angle mass with cerebrospinal fluid intensity. T1-weighted (A) and T2-weighted (B) axial MRI scans.

FIGURE 3. Axial magnetic resonance imaging (MRI) scans showing a right frontal arachnoid cyst with cerebrospinal fluid intensity. T1-weighted (A) and T2-weighted (B) axial MRI scans.
The distribution of ACs in our series was found to be concordant with that reported in the literature.

Symptoms of intracranial ACs vary by age and location. Most (60%-90%) ACs become symptomatic in early childhood. The onset of symptoms and signs is usually a result of 3 primary factors: cortical irritation, compression of the cerebral parenchyma, and obstruction of CSF circulation. Occasionally, they may produce symptoms because of bleeding. The common neurologic features are headache, seizures, hydrocephalus, developmental delay, psychomotor retardation, specific signs resulting from compression of neuronal tissues, or elevated intracranial pressure, focal compensatory enlargement of the skull, visual impairment, proptosis, endocrine dysfunction, cerebellar signs, nystagmus, and gait disturbance. Epileptic seizures and headache were the most common presenting clinical features in our series.

The preoperative diagnosis of an AC can be easily made with MRI or CT. The AC appears hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences, and it demonstrate signal characteristics similar to CSF on all sequences, including fast spin echo and proton-density images on MRI scanning. There was no enhancement of the cyst contents or cyst wall after the intravenous administration of gadolinium. MRI is also adequate to evaluate the compression of cysts on neural structures. The differential diagnosis of AC includes epidermoid tumor and other cystic collections such as craniopharyngioma, astrocytoma, and chronic subdural hematoma. On MRI, however, the combination of extra-axial location, morphologic features, and signal intensity matching that of CSF allows one to make the diagnosis of an uncomplicated AC with confidence. The epidermoid tumor can be indistinguishable on routine CT and MRI; however, because their management differs considerably, further MRI sequences are required when an AC is suspected. ACs show a CSF signal on the diffusion-weighted sequences, excluding an epidermoid tumor. A fast spin echo technique known as fluid-attenuated inversion recovery is highly T2-sensitive for water in solid tissue and is able to suppress signals arising from CSF spaces so that they remain black, whereas an epidermoid tumor becomes hyperintense.

Because ACs are congenital, it is possible to identify them in the fetus. Widespread use of ultrasound has resulted in antenatal diagnosis of ACs, but the diagnosis is difficult and they may be confused with other cystic structures. Some antenatally detected ACs have disappeared before birth. After birth, many are asymptomatic and remain quiescent for years, although others expand and cause symptoms.

The indications for surgery and the type of surgical treatment are still matters of debate. An intensive review of the literature revealed mostly positive results regarding surgical treatment. Asymptomatic ACs do not require treatment, however. Patients with an asymptomatic cyst should be followed clinically and radiologically. Headache, intractable seizures, intracranial hypertension, progressive hydrocephalus and growth of the cyst, specific signs resulting from compression of neuronal tissues, or refractory symptoms referable to a cyst have been reported as indications for surgery in the literature. Intractable seizures and specific signs resulting from compression of neuronal tissues were specifically accepted as indications for surgery in our series. An excellent response to antiepileptic drugs was obtained in a significant portion of our patients with focal paroxysmal discharges on EEG. We suggest that antiepileptic drugs be tried in these patients before surgery is considered.

Headache was reported as a major indication for surgery by Westerl. ACs presenting with headaches only require careful evaluation to determine whether the cyst is actually symptomatic and treatment is indicated, however. Headache as the primary symptom was present in 5 cases and was treated successfully with analgesics in our series. Headache alone was not accepted as a surgical indication.

Different modes of surgical approaches have been reported. Cyst fenestration, stereotactic puncture, endoscopic cyst fenestration, cystoperitoneal shunt, cyst marsupialization into the subarachnoid space, and complete or partial resection of the cyst wall are the main options. The advantages and disadvantages of each surgical treatment have been discussed extensively. Most authors prefer craniotomy and fenestration or marsupialization of the cyst wall. The greatest advantage of this option is that patients are independent of a shunt. Performing a craniotomy allows the surgeon to inspect the cyst wall, coagulate any arachnoid blood vessels, and confirm the diagnosis with tissue biopsy. Reclosure of the cyst wall and insufficient fenestration resulting in adequate drainage into the subarachnoid space are the primary reasons for recurrence and regrowth. Only sporadic reports of clinical recurrence after fenestration can be found in the literature. Cystoperitoneal shunting carries a
considerable rate of obstruction and/or recurrence, however. Nevertheless, cyst shunting has good results, with a low rate of recurrence, for ACs located in the middle cranial fossa associated with hydrocephalus. Endoscopic cystostomometry is also reported in the literature. This is a minimally invasive technique that has been reported to be safe and effective; in the case of failure, more aggressive surgery is still possible.

Microsurgical excision and fenestration were performed on 7 patients in our series and resulted in a reduction in cyst size and the disappearance of epileptic seizures in 6 patients. Clinical improvement was not seen in peripheral facial palsy, proptosis, and nystagmus, however. We considered that some neurologic symptoms such as nerve palsy attributed to long-term compression of ACs might not improve by means of surgery. Cystoperitoneal shunting was performed only in 2 intractable seizure patients, which resulted in a partial reduction of seizure frequency in our series. These patients could not discontinue antiepileptic drugs, however.

CONCLUSIONS

The study indicates that ACs occur more commonly in the middle cranial fossa, on the left side, and in male patients. We also conclude that the major indication for surgery in patients with ACs is the presence of intractable seizures, intracranial hypertension, and compression of neuronal tissues. Headache only is not a surgical indication. Microsurgical excision and fenestration are safe and effective for surgical treatment of ACs.

REFERENCES


