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Examination of intracranial arachnoid cysts in children, symptomatic or asymptomatic

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Keywords
Arachnoid Cysts; Intracranial Hemorrhages; Magnetic Resonance Imaging; Epilepsy

Abstract

Background: Arachnoid cysts (ACs) are non-neoplastic, intracranial cerebrospinal fluid (CSF)-filled cavities lined with arachnoid membranes. Most of them are found incidentally and are asymptomatic. The aim of this study was to describe the clinical and demographic characteristics of ACs and to investigate the frequency of complications.

Methods: ACs were classified as cerebral and cerebellar, and were evaluated according to location and size using the Galassi classification.

Results: Evaluation was made of 103 patients with AC admitted to the pediatric neurology department. The patients comprised 62.1% boys and 37.9% girls with a mean age of 10.1 ± 4.9 years. Headache was present in 33% of patients, epileptic seizures in 22.3%, and neuromotor developmental delay in 14%. Spontaneous intracranial hemorrhage (ICH) was observed in 6 patients (5.8%), with 3 (50%) having a cyst diameter ≥ 5 cm. ACs frequently involved the retrocerebellar and temporal lobes. Epilepsy was diagnosed in 26 patients (25.2%) and in only one of them, epileptic discharge and AC originated from the same region. The risk of bleeding was found to be higher in patients with cyst diameter ≥ 5 cm (P = 0.032).

Conclusion: Although ACs are generally considered harmless, a cyst size ≥ 5 cm is important in terms of the risk of bleeding and should be followed up regularly. Although it has been reported in the literature that ACs may be associated with epilepsy, no significant correlation was found between seizure type and electroencephalography (EEG) recordings in this study.

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Introduction

Arachnoid cysts (ACs) are non-neoplastic, intracranial cavities lined with arachnoid membranes and filled with cerebrospinal fluid (CSF). They are collections of CSF within the arachnoid membranes and are secreted by the arachnoid cells lining the cyst.[1] ACs constitute approximately 1% of intracranial lesions. The incidence of AC in children is thought to be approximately 1%, and the pathogenesis is not fully known. Although most are sporadic, rare familial forms have also been described.[2,3] The majority are found incidentally and are usually considered asymptomatic. A very small proportion of patients become symptomatic and require surgical intervention. Clinical symptoms of an AC may include headache, signs of increased intracranial pressure, hydrocephalus, local mass effect, or cyst rupture.[4] The majority of ACs found in the pediatric population are asymptomatic.[5]

In the majority of asymptomatic lesions, imaging and neurological examinations are sufficient. However, surgery is indicated if there are symptoms of increased intracranial pressure, seizures, focal neurological deficits, or cognitive impairment.[6]

This study aimed to describe the clinical and demographic characteristics of AC and investigate the frequency of complications.

Materials and Methods

The study included 103 patients with AC who were admitted to the Department of Pediatric Neurology of S.B.U Tepecik Training and Research Hospital, Izmir, Turkey, between December 2021 and December 2023. The clinical and demographic characteristics of the patients included in the study were analysed.

Electroencephalography (EEG) was performed on patients with seizures or suspected seizures. The ACs were classified anatomically using the Galassi classification, which categorizes cerebral ACs based on location and size.[7] In the Galassi classification, middle fossa cysts are categorised in three types. Type I cysts are usually asymptomatic and located in the anterior middle cranial fossa. Type II cysts extend upward along the sylvian fissure and sometimes displace the temporal lobe. Type III cysts are very large and occupy the entire middle cranial fossa, not only displacing the temporal lobe but also disrupting the parietal and frontal lobes (Table 1).[8,9]

Clinical findings, the need for surgical intervention, and the development of complications of the ACs were analyzed according to Galassi classification. The patients were separated into 2 groups as those with AC diameter < 5 cm or ≥ 5 cm according to the maximum cyst diameter. The incidence of spontaneous intracranial hemorrhage (ICH) was analyzed.

Results

Evaluations were made of 103 patients, comprising 39 (37.9%) girls and 64 (62.1%) boys, with a mean age of 10.1 ± 4.9 years. The indications for brain magnetic resonance imaging (MRI) were headache in 34 (33%) patients, epileptic seizure in 23 (22.3%) patients, differential diagnosis of seizure in 16 (15.5%) patients, further investigation for neuromotor developmental delay in 14 (13.6%) patients, head trauma in 5 (4.9%) patients, increased intracranial pressure in 4 (3.9%) patients, ICH in 3 (2.9%) patients, intrauterine diagnosis in 1 (1%) patient, and other reasons in 3 (2.9%) patients. The clinical and demographic characteristics of the patients are summarized in table 2.

The ACs were divided into two groups as cerebral and cerebellar. Cerebral ACs were found in 53 (51.5%) patients and cerebellar ACs in 50 (48.5%) patients. When the age at which AC was diagnosed and the rates of being symptomatic were analyzed, it was seen that the likelihood of being symptomatic was higher, especially in patients aged > 12 years (P = 0.036).

The brain lobes in which the ACs were observed are presented in detail in table 2. When cerebral and cerebellar ACs were compared, no significant difference was found between the age at presentation, effect on neuromotor development, frequency of epilepsy, and complication rates (Table 3).

Developmental delay was determined in a total of 30 patients.

Table 1. Galassi classification[8]

Type 1	Type 2	Type 3
Small Usually asymptomatic In the anterior, middle cranial fossa	Located at the top along the sylvian fissure Displacing the temporal lobe	The cysts are very large and occupy the entire middle cranial fossa, not only displacing the temporal lobe but also disrupting the parietal and frontal lobes

Table 2. Clinical and demographic characteristics of the patients with arachnoid cysts (ACs)

Family history of AC	**n (%)**	
History of NICU hospitalization	15 (14.6)	
Developmental delay	30 (29.1)	
Epilepsy	26 (25.2)	
Age range at diagnosis of AC and rate of being symptomatic (year)		
0-6	26 (25.3)	1 (3.8)
6-12	40 (38.8)	0 (0)
12-18	37 (35.9)	5 (13.5)
Physical examination findings		
Papilledema	10 (9.7)	
Spastic tetraparesis	4 (3.9)	
Microcephaly	3 (2.9)	
Hemiplegia	2 (1.9)	
Brain region affected		
Cerebral	53 (51.5)	
Frontal	4 (7.5)	
Parietal	3 (5.7)	
Temporal	41 (39.8)	
Occipital	1 (1.0)	
Multiple lobe involvement	4 (3.9)	
Cerebellar	50 (48.5)	
Galassi classification		
Type 1	40 (75.5)	
Type 2	10 (18.9)	
Type 3	3 (5.7)	

AC: Arachnoid cyst; NICU: Neonatal intensive care unit

When the relationship between AC localization and developmental delay was examined, developmental delay was found in 15 (30%) of 50 patients with cerebellar involvement, 13 (31.7%) of 41 patients with temporal lobe involvement, and 2 (50%) of 4 patients with multiple lobe involvement.

Epilepsy was diagnosed in 26 (25.2%) patients, and was determined to be resistant in only one of these patients. EEG was performed in 23 patients with epileptic seizures, 16 patients with suspected seizures, and 3 patients who were started on prophylactic antiepileptics after ICH (42 patients in total). Of these patients, 29 (69%) had normal EEG. Generalized epileptic discharges were detected in 6 (14.3%) and focal epileptic discharges were detected in 7 (16.7%) of 13 (31%) patients with abnormal EEG. In only one patient with focal epileptic discharges, epileptic discharge and the AC originated from the same region.

When neuromotor development, epilepsy development, and complications of cerebral ACs were analyzed according to the Galassi classification, it was observed that type 2 and type 3 ACs had a higher risk of spontaneous ICH. Galassi staging did not predict the development of epilepsy and neuromotor retardation (Table 4).

The ACs were separated into two groups as < 5 cm and ≥ 5 cm according to the maximum diameter. Cyst diameter ≥ 5 cm was determined in 6 (13.6%) patients. Spontaneous ICH was observed in 6 (5.8%) patients, of whom 3 had maximum diameter of the AC of ≥ 5 cm. A comparison of the risk of spontaneous ICH between patients with AC diameters < 5 cm and ≥ 5 cm showed that the risk of bleeding was higher in patients with a cyst diameter ≥ 5 cm (P = 0.032).

Table 3. Comparisons of clinical and demographic characteristics of cerebral and cerebellar arachnoid cysts (ACs)

Variable	**Cerebral (n = 53)**	**Cerebellar (n = 50)**	**P**
Age (year) (mean ± SD)	9.8 ± 5.1	10.5 ± 4.6	0.450
Gender (girl) [n (%)]	20 (37.7)	19 (38.0)	0.978
Developmental delay [n (%)]	15 (28.3)	15 (30.0)	0.850
Epilepsy [n (%)]	13 (25.4)	13 (26.0)	0.908
Headache [n (%)]	20 (37.7)	15 (30.0)	0.407
Spontaneous ICH [n (%)]	4 (7.5)	2 (4.0)	0.442

ICH: Intracranial hemorrhage; SD: Standard devation

Table 4. Comparisons of Galassi classification and clinical findings

Variable	Type 1 (n = 40) [n (%)]	Type 2-3 (n = 13) [n (%)]	P
Epilepsy	8 (20.0)	5 (38.5)	0.196
Developmental delay	11 (27.5)	4 (30.8)	> 0.999
Spontaneous ICH	1 (2.5)	3 (23.1)	0.042

ICH: Intracranial hemorrhage

Discussion

ACs are usually detected incidentally during brain imaging studies performed for other reasons. The frequency of detecting these cysts has increased with the widespread use of neuroimaging techniques.[5] ACs account for approximately 1% of intracranial masses.[10]

The incidence of ACs is approximately 2:1 higher in men than in women.[10-12] In this study, the incidence of ACs was higher in boys, similar to the literature. The detection rate of ACs increases with age, and they are more likely to be symptomatic in children.[12,13] In the current study, similar to the general literature, the detection rate of ACs increased with age, but unlike the literature, the likelihood of being symptomatic increased with increasing age (especially above 12 years).

This has been attributed to the widespread use of imaging modalities and increased accessibility of imaging modalities, as well as physicians using imaging modalities more frequently to increase diagnostic accuracy due to increasing malpractice lawsuits in recent years.

The detection of ACs in patients presenting at healthcare institutions with headache complaints is most likely a clinically insignificant and incidental finding.[10,14]

When ACs are symptomatic in adults, the most common symptom is headache. According to previous studies, 66% of adults with ACs have headache symptoms.[15] In a review of 45 pediatric patients aged 2 to 17 years, headache was the primary symptom in 69% of patients.[16]

Coskuner et al. reported that 29.4% of patients with ACs in the pediatric age group presented at hospital with headache.[17] In the current study, headache was the most common reason for presentation and the primary symptom at the rate of 33%, consistent with the findings in the literature. It has also been predicted that the incidence of headaches may increase with age.

ACs are structures that are rarely detected in the prenatal period and can occur as extra-axial cysts in any part of the brain, usually after the second trimester. Ultrasonography and MRI should be used to differentiate these cysts from other brain cysts and tumors.[18] In the current study, an AC was diagnosed intrauterine in one case, indicating that such cysts can be detected in the prenatal period.

Lang et al. examined the effect of ACs on cognition and found cognitive impairment in 9 of 10 patients with temporal or temporo-frontal cysts.[19] Kwiatkowska et al. reported that children with ACs in the middle cranial fossa performed worse on the Stanford-Binet Intelligence Scale compared to the general population. Deficits were observed in visual-spatial reasoning, quantitative reasoning, long-term memory, and learning process.[9,20]

There are also reports that ACs in the temporal region are associated with delayed psychomotor development.[21] Similar to the literature, the frequency of developmental delay in the current study was found to be higher in patients with temporal lobe involvement, and the frequency of developmental delay was also higher in patients with cerebellar involvement. This was attributed to the high frequency of incidental detection of ACs and the widespread use of brain MRI as part of the etiological investigation in patients with developmental delays.

Although epileptic seizures have been reported in patients with ACs,[22-24] there are also studies indicating that these cysts may not be associated with a specific seizure type or EEG focus.[25,26] Epilepsy was diagnosed in 26 patients (25.2%) and 13 of these patients had normal EEG results, while epileptic focus was detected in 13 patients. In only one of the patients with focal epileptic discharges, the AC and the involved area were the same. This finding supports the results reported in the literature that ACs are not associated with a specific seizure type and EEG findings.[25,26]

It is known that ACs frequently involve the cerebellar region and temporal lobe.[1] Consistent with these findings, temporal and cerebellar lobes were the most frequently involved lobes in the current study.

Spontaneous or traumatic rupture is a rare complication of ACs. Rupture of the cyst leads to subdural hygroma formation and increased intracranial pressure, necessitating surgical

intervention.[27,28] Surgical treatment in asymptomatic patients should be evaluated on a patient-by-patient basis, taking into account the potential risk of rupture. Reported surgical approaches include craniotomy, cyst shunting, and endoscopic fenestration.[29] In addition to all these issues, there are also reports that ACs, especially those formed after trauma, resolve spontaneously.[30]

The bleeding rate of ACs in the pediatric population is low (0.3%-6%). Risk factors for bleeding include trauma and large cyst size.[31-33] Cress et al. found that the risk of bleeding was higher in patients with a cyst diameter > 5 cm.[31] The rate of spontaneous ICH was found to be 5.8% in the current study, which is consistent with the results reported in the literature. In addition, a higher rate of bleeding was observed in patients with a cyst diameter ≥ 5 cm.

Limitations: Since this study was retrospective in design, follow-up imaging could not be performed. Only patients with seizures or suspected seizures could undergo EEG.

Conclusion

The increasing number of malpractice lawsuits in recent years has led to increasing use of preventative medicine practices and to the more frequent use of imaging methods by physicians to increase diagnostic accuracy. This has led to an increase in the frequency of detection of ACs. Although mostly considered harmless, cyst size ≥ 5 cm is an important determining factor for the risk of bleeding. Therefore, patients should be followed up at regular intervals. Although the literature reports an association between AC and epilepsy, this study found no significant correlation between seizure type and EEG recordings.

Conflict of Interests

The authors declare no conflict of interest in this study.

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