Arachnoid Cysts

State-of-the-Art

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Foreword

Very few nosographic entities have attracted the attention of pediatric and adult neurosurgeons like arachnoid cysts (ACs). There are several reasons for such an interest, namely the heterogeneous nature of these lesions due to their different locations, their variable impact on the neurocognitive development, the possible association with disturbances of the cerebrospinal fluid (CSF) dynamics, the induced endocrinological alterations, and seizure disorders.

The treatment of an AC can be challenging. Indeed, a conservative management can be adopted in some specific cases; in others the surgical indication depends not only on the mere presence of the malformation but also on the age of the patient at the time of its diagnosis. A typical example is provided by the quite common temporal ACs that interfere with the cerebral development in early life, thus requiring a prompt surgical excision if recognized. However, these cysts often reach a stable spatial equilibrium with the brain and tend to remain unchanged after childhood and adolescence. Consequently, in the case of accidental late recognition a wait-and-see policy should be taken into account. A similar consideration could be made for asymptomatic and accidentally discovered interhemispheric ACs diagnosed at a late age. On the other hand, the surgical indications cannot be ignored in cases of chiasmatic region or posterior cranial fossa ACs because of their obvious role in causing severe endocrinological and CSF dynamics disturbances.

A further limitation on the correct surgical indications can be in some cases due to the difficulty in the interpretation of the clinical manifestations attributed to the presence of the cyst. Once more, this limit concerns the temporal ACs when associated with seizure disorders. Often, the epilepsy does not resolve even after the complete surgical excision of the lesion, thereby demonstrating that the cyst was not the cause of the seizures but rather a manifestation of an underlying developmental malformative process of the temporal lobe. Also, those cases of surprising re-bleeding of ACs operated on because of intracystic hemorrhage can be interpreted on the grounds of the abovementioned consideration.

For several years the debate on the most appropriate surgical modality has been centered on the choice between the craniotomic direct approach for the surgical excision of the lining membranes or for the diversion of the cyst fluid content into the ventricular system and subarachnoid spaces or an indirect approach, in most cases a cysto-peritoneal shunt. The advent of endoscopy

has deeply modified the surgical management of ACs providing a further and effective management tool that is apt to reduce the surgical risk for several patients.

On the grounds of these considerations and the still ongoing discussion of the more appropriate management of ACs, the editors should be congratulated for the timely chosen subject, still open to research, of their well-organized book as well as for the numerous and widely respected coauthors that they were able to gather.

I wish them and the book the deserved success.

Pediatric Neurosurgery
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Concezio Di Rocco

Contents

Part I

General Information About Arachnoid Cysts

Editor's Summary

In this "Part I: General Information About Arachnoid Cysts," there are a total of six chapters describing the general aspects of the arachnoid cysts (ACs), noncancerous collections of cerebrospinal fluid (CSF) localized by the arachnoid membranes of the brain or spinal cord. Even today, unlike many other space-occupying lesions, they are usually asymptomatic and their management can be challenging for neurologists, neuroradiologists, and neurosurgeons due to the heterogeneous nature of these lesions. The first of the chapters in this section compiles the historical and etymological aspects of ACs in light of the current literature. In the second chapter, the authors review both the gender distribution and lateralization of ACs, which can present in many different locations within the central nervous system. In the subsequent two chapters, the embryological and histological features of ACs that influence their cerebral development in early life, the authors illustrate this process with demonstrative histological figures and special drawings. Interestingly, these cysts usually reach a stable spatial equilibrium with the brain or spinal cord, and they tend to remain unchanged all throughout life. The fifth chapter addresses a variety of pathophysiological mechanisms for their formation and their possible association with disturbances of the CSF dynamics, such as fluid secretion and slit-valve mechanism. The last remaining chapter provides the biochemical findings of the AC fluid in detail for the readers of the book.

History and Etymology of Arachnoid Cysts

Timothy Beutler

1 Introduction

The history of arachnoid cysts (ACs) is difficult to trace through the literature. The nomenclature has changed over time, and some of the earliest reports likely mixed in with different pathologies including resolving subdural hematoma and infections. Early descriptions of ACs associate the entity with a variety of conditions ranging from spontaneous and seemingly incidental to posthemorrhagic and inflammatory conditions.

The history of ACs begins in the early nineteenth century with postmortem autopsy studies. ACs in most of these patients were incidental findings and not related to the cause of death. In the early twentieth century, we begin to see the earliest operative reports of patients with symptomatic ACs. By the mid-twentieth to late twentieth century, new imaging technology led to the increased discovery of asymptomatic ACs and the emergence of modern management practices.

2 Early History

The early history of ACs is derived primarily from postmortem pathology studies which became more prevalent in the nineteenth century. Many of these studies were unfortunately performed in asylum populations, so some of the literature has bias toward an association with mental illness. Additionally, early studies also tended to describe any cyst or collection located beneath the dura as an AC and most likely included additional pathologies such as resolving hemorrhages and infections.

One of earliest attempts to characterize the prevalence of ACs was completed by Crichton Brown in 1875 and estimated a 5% prevalence in his asylum population after reviewing over 1000 autopsies [7]. However, this study likely overestimated the true prevalence of the condition as some descriptions appeared to describe a variety of hemorrhagic, infectious, and inflammatory pathologies.

3 Serous Cysts in the Arachnoid

The earliest description of an AC is attributed to the English physician Richard Bright who practiced at Guy's Hospital in London in the early nineteenth century. He utilized autopsies to understand a variety of diseases and published two large volumes of medical cases with accompanying illustrations in 1831 [6]. In the section on diseases of the brain and nervous system, he reports two cases of ACs that were discovered on autopsy and publishes what is considered the first two medical illustrations of the condition. The

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first illustration shows what appears to be a large middle cranial fossa AC (Fig. 1), while the second illustration shows two smaller ACs located over the convexity (Fig. 2).

Bright observed that the cysts were apparently asymptomatic and discovered incidentally at autopsy. They are described as being located between the layers of the arachnoid or attached to the arachnoid by thin adventitious membranes. Their sizes varied from the "size of a pea to that of a large orange" [6]. Notably, he postulated that the condition was likely chronic and observed that the "bone of the skull is molded to their form" [6].

As early as the 1860s, it was recognized that cysts in the arachnoid space were not uncommon [25]. At that point, there were two prevailing theories regarding their pathophysiology that lead to occasionally confusing nomenclature in the early literature. The first theory was that they were a result of an accumulation of lymph that occurs secondary to arachnoiditis. The second theory was that they are the result of chronic changes from previous hemorrhage.

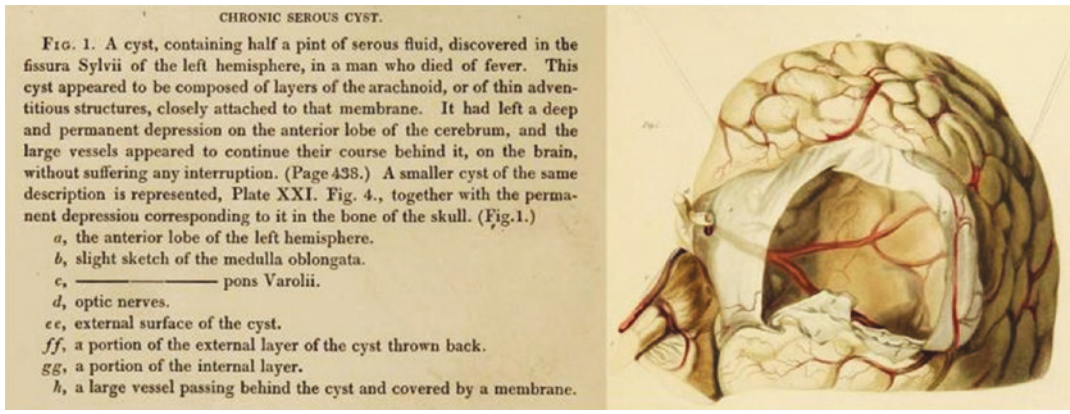
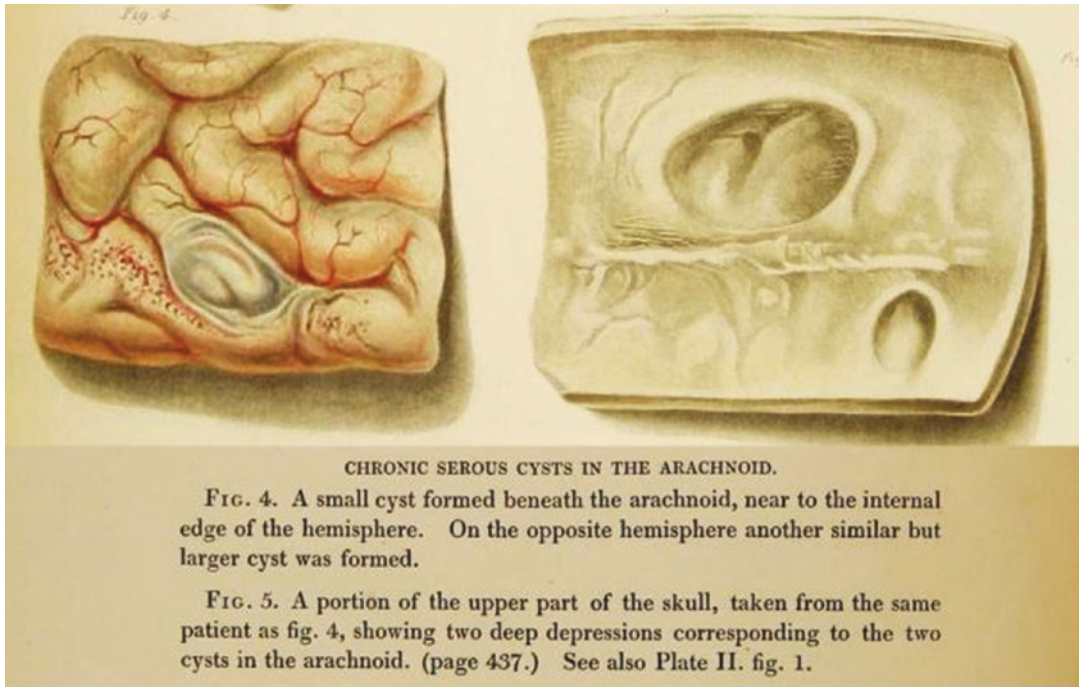

CHRONIC SEROUS CYST.

FIG. 1. A cyst, containing half a pint of serous fluid, discovered in the fissura Sylvii of the left hemisphere, in a man who died of fever. This cyst appeared to be composed of layers of the arachnoid, or of thin adventitious structures, closely attached to that membrane. It had left a deep and permanent depression on the anterior lobe of the cerebrum, and the large vessels appeared to continue their course behind it, on the brain, without suffering any interruption. (Page 438.) A smaller cyst of the same description is represented, Plate XXI. Fig. 4., together with the permanent depression corresponding to it in the bone of the skull. (Fig. 1.)

a, the anterior lobe of the left hemisphere.
b, slight sketch of the medulla oblongata.
c, ——————— pons Varolii.
d, optic nerves.
ee, external surface of the cyst.
ff, a portion of the external layer of the cyst thrown back.
gg, a portion of the internal layer.
h, a large vessel passing behind the cyst and covered by a membrane.

Fig. 1 Large middle cranial fossa cyst. (Depicted by Richard Bright in "Reports of Medical Cases, Selected with a View of Illustrating the Symptoms and Cure of Diseases by a Reference to Morbid Anatomy." Volume II "Diseases of the Brain and Nervous System: Part I": Fig. 1 on plate II and corresponding legend description. Originally published in 1831)



CHRONIC SEROUS CYSTS IN THE ARACHNOID.

FIG. 4. A small cyst formed beneath the arachnoid, near to the internal edge of the hemisphere. On the opposite hemisphere another similar but larger cyst was formed.

FIG. 5. A portion of the upper part of the skull, taken from the same patient as fig. 4, showing two deep depressions corresponding to the two cysts in the arachnoid. (page 437.) See also Plate II. fig. 1.

Fig. 2 Small AC and corresponding portion of the skull. (Depicted by Richard Bright in "Reports of Medical Cases, Selected with a View of Illustrating the Symptoms and Cure of Diseases by a Reference to Morbid Anatomy." Volume II "Diseases of the Brain and Nervous System: Part II": Figs. 4 and 5 on plate XXI and corresponding legend description. Originally published in 1831)

4 Organization of Lymph

The organization of lymph is perhaps the earliest theory regarding the pathophysiology of ACs. It is unclear from where the lymph theory originated; however, in an 1865 paper by Samuel Wilks entitled "Cyst in the Cavity of the Arachnoid," he described an AC specimen in Guy's Hospital that Thomas Hodgkin attributed to the organization of lymph [25].

Arachnoiditis was thought to lead to a spontaneous accumulation of lymph between the layers of the arachnoid leading to the formation of ACs. However, this theory was supplanted by the belief that ACs were formed as a result of an effusion of blood. While ACs were found in patients with no history of either trauma or infection, apoplexy had been commonly observed, whereas spontaneous arachnoiditis had not been seen. Even French physician Achille-Louis Foville, while classifying ACs under his "Meningite parietale," admitted that "their inflammatory nature" [was] not sufficiently well established [25].

5 Extravasation of Blood into the Cavity of the Arachnoid

In 1845, Prescott Hewett attempted to outline the pathophysiology of ACs in a paper entitled "Extravasations of Blood into the Cavity of the Arachnoid" [13]. He described four divisions of hemorrhage in the subarachnoid space ranging from simple to complex collections.

At first glance, to a modern reader, the work of Hewett appears to be unrelated to ACs and seems to better describe the pathophysiology that we commonly encounter with resolving intraparenchymal and subdural hemorrhages. However, careful examination of the manuscript reveals references to AC case descriptions provided by Hodgkin and Bright from Guy's Hospital [13].

Hewett describes the pathology of ACs in the following excerpt:

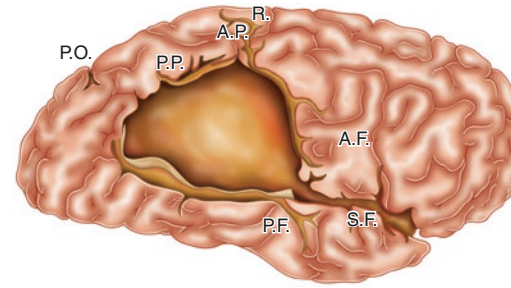
> Thus organized, these cysts possess all the physiological characters of an original serous membrane; they secrete, they absorb; they have been found filed with clots of fibrine and blood-tinged serum; sometimes they contain serum alone, of various colours, and often times, in the cavity of the same cyst, are found coagula of blood of various hues, some recently effused, and others of long standing [13].

By 1865, Samuel Wilks wrote that the work of Hewett was generally accepted as the correct explanation for the origin of ACs. He further describes the end result of the cyst after all the blood had been reabsorbed:

> The interior of the cyst, if it be old, has generally lost all traces of blood, and contains merely shreds of fibrous material of the same structure as the walls, together with a limpid fluid and cholesterine [25].

6 Chronic and Asymptomatic

Even from the earliest descriptions by Richard Bright in 1831, there was the belief that ACs were generally chronic and asymptomatic entities. The earliest cysts were discovered incidentally on autopsy studies, and so they were associated with a benign and indolent course. During the early pre-surgical era, even large supratentorial ACs were discovered. One case report from 1879 by D.J. Cunningham described discovering a cystic cavitation in the right parietal lobe "large enough to hold a hen's egg" in a patient who passed away from diabetes [9]. What was remarkable at the time was that the cyst extended posteriorly to the Rolandic fissure and the patient was not known to have any weakness (Fig. 3). Observations such as this one reinforced the belief that ACs were asymptomatic and chronic entities. However, our understanding of the etiology of ACs changed again in the early twentieth century with the emergence of neurosurgery as a surgically discipline.



Profile of right hemisphere to show position of cyst (reduced to a little more than half size). SF, Sylvian fissure; R, Fissure of Rolando; PO, Parieto-occipital fissure; PF, Parallel fissure; AF, Ascending frontal convolution; PP, Postero-parietal convolution; AP, Ascending parietal convolution.

Fig. 3 Large right middle fossa AC. (Depicted by D.J. Cunningham in "A large sub-arachnoid cyst involving the greater part of the parietal lobe of the brain." Journal of anatomy and physiology originally published in 1879)

7 Early Surgical Experience

In the early twentieth century, due largely in part to advancements in anesthesia and the localization of neurologic disease, case reports of successful surgery for symptomatic ACs begins to emerge in the literature. X-rays were only invented by Wilhelm Rontgen in 1895, so localization of symptoms based on a detailed neurologic exam was paramount for the successful surgical treatment of a patient with neurologic disease.

8 Spinal Arachnoid Cysts

Some of the earliest reports of successful surgery for ACs were for spinal ACs [5, 21, 23]. The earliest American report of a spinal AC was written by Spiller, Musser, and Martin in 1903 [23]. They presented a case of a lumbar AC in a young woman who presented with progressive left leg pain, weakness, and paresthesias. They describe treatment with an L2–L4 laminectomy

and report encountering a cyst measuring approximately an inch and a half in length after opening the dura. The walls of the cyst were described as of a "flimsy consistency" and filled with clear fluid which they reported was under "unusual intradural pressure" [23]. After surgery, they reported the gradual improvement in the patient's preoperative symptoms with near complete recovery after 6 years. Their description is unique from many other craniospinal cysts described during this era which were secondary to parasitic infections from cysticercus and echinococcus.

By 1915, there were at least six spinal ACs that had been described in the surgical literature [21]. These cysts ranged in location throughout the cervical, thoracic, and lumbar spine. Diagnostic tools were limited at this time, and much emphasis was placed on thoroughly documenting a neurologic exam for localization of spinal symptoms. Rachiocentesis was also recommended as part of the workup and sometime completed at levels above and below where the suspected lesion was located. Outcomes for these early patients were often reported as good as long as the surgery was completed early in course of the patient's symptoms. Those who presented with severe symptoms such as paraplegia did not have outcomes as good as those who presented with only pain and sensory changes.

In a review of the first six cases of spinal ACs, Skoog describes all the cases having been found in women aged 17 to 50 years [21]. Histological reports are limited from the early literature as the thin walls of the cysts were difficult to preserve during surgery. Trauma and infectious etiologies were still thought to be the most likely provocative factors contributing to cyst formation. Skoog writes regarding their pathogenesis:

> I believe that there may be several provocative agents in the different cases and that from one of the irritating factors there are formed adhesions with the endothelial cells and fibers of the arachnoid. This is followed by an exudate or secretion from the delicate endothelial cells lining the fibrils or covering the interior of the cyst. Thus there is a very slow accumulation of fluid followed by the gradual development of the cord compression [21].

9 Posterior Fossa Cysts

In 1893, William Krause presented a case of a 36-year-old man with worsening posterior headaches, nausea, vomiting, vertigo, and an unsteady gait [16]. While the patient's symptoms were correctly localized to the cerebellum, he unfortunately passed away before any treatment. Upon autopsy, a large cystic cavity was found in the right cerebellum filled with an amber-colored fluid. While this cyst was likely not a primary AC, Krause wrote his article to show that the constellation of symptoms for posterior fossa lesions was specific and distinct enough to support surgical intervention.

By the 1930s, a series of cases had been described by multiple surgeons of ACs in the posterior fossa [3, 10, 14]. At this time, theories of infection and hemorrhage still remained common despite little evidence to support either consideration. Most of the cysts in the posterior fossa were reported either in a midline location or in the cerebellopontine angle (CPA). Posterior fossa cysts would often present with findings consistent with elevated intracranial pressure and obstructive hydrocephalus. Those cysts in the CPA were often associated with inflammation and otitis media. Many of the midline cerebellar cysts, including some of Cushing's cases described by Horrax, appear more consistent with arachnoiditis or dilation of the cisterna magna than with well-circumscribed cysts [14].

10 Primary Arachnoid Cysts

While ACs were described in earlier literature, there was little distinction between what a modern reader would recognize as an AC and any other kind of cystic pathology. The early literature was often limited to gross anatomic descriptions of lesions, and histologic reports of the lesions are more difficult to find because the walls of many cysts were thin and difficult to preserve. The histologic descriptions that were available varied greatly with some reporting structures similar to normal arachnoid, some reporting resolving hemorrhage, and others describing nonspecific inflammatory changes.

Throughout the first half of the twentieth century, diagnostic imaging techniques continued to advance. As a result, preoperative workup progressed to include a number of techniques including cerebral angiography, air encephalography, and electroencephalography [20].

For example, in the case of large middle cranial fossa cyst, plain X-rays may show thinning and expansion of the temporal squama and surrounding frontal and parietal bone enlarging the middle cranial fossa and elevating the lesser wing of the sphenoid. Cerebral angiography may show displacement of the arteries on the ipsilateral side with the middle cerebral arteries being shifted upward and medially and the anterior cerebral artery shifted contralaterally. Air encephalography could show some contralateral shift of the lateral ventricles as well as a truncated or dorsally displaced ipsilateral temporal horn. Electroencephalogram may be unremarkable or show some generalized slowing in the region of the cyst.

Advancements in imaging as well as greater attention to the histology of ACs ultimately led to the development of one of the earliest classifications of ACs.

In a case reported by Leslie Oliver in 1958, he described a 21-year-old man who presented with worsening headaches, vomiting, and diplopia 10 weeks after sustaining a traumatic sports injury to the chest [19]. On physical exam, he experienced diplopia when looking to the right and was noted to have left upper facial weakness. Fundoscopic exam revealed bilateral papilledema, and the lumbar puncture showed an opening pressure of 30 cm of H_2O. Ventriculography showed that the ventricles were displaced to the left. The patient underwent a right-sided craniotomy, and a large AC was discovered filled with yellow fluid. The cyst separated the Sylvian fissure and extended well into the anterior and middle fossa.

Histologic examination of the cyst wall showed "arachnoid membrane thickened by fibrosis, and showing (a) considerable recent focal haemorrhage, and (b) areas of greatly increased vascularity associated with chronic inflammatory infiltration and considerable numbers of macrophages containing iron pigment" [19].

Because of the patient's recent trauma and histologic description, the etiology of the cyst was originally attributed to trauma. However, air encephalography completed 8 weeks after surgery revealed a persistent empty space in the area formerly occupied by the cyst, which raised the question of whether the cyst had been present for much longer.

Oliver's discussion of the AC is significant because he proposed classifying ACs as either primary or secondary. Primary ACs were defined as congenital lesions that were formed by the absence rather than displacement of nervous tissue, whereas secondary ACs were those lesions that formed due to injury, inflammation, or tumors.

The same year that Oliver's paper was published, an autopsy series of four cases of ACs was published by Starkman, Brown, and Linell [22]. In this report, they described how these idiopathic cysts differed in their anatomic relationships from those cysts that are associated with inflammation and other secondary causes. They concluded that idiopathic or primary ACs likely result from a focal derangement in the normal development of the leptomeninges.

11 Temporal Lobe Agenesis

Congenital and developmental theories concerning AC formation varied in the lateral half of the twentieth century. While Starkman, Brown, and Linell thought these cysts formed due to aberrant development of the leptomeninges, others such as Robinson thought that cyst formation was due to a defect in brain formation. Robinson referred to this process as temporal lobe agenesis [20].

Patients with temporal lobe agenesis usually presented at a young age with local bulging of the skull with underlying large intracranial collections of fluid. These lesions were described as "bluish transparent cyst[s]" containing clear fluid [20]. They often were so large that upon opening the outer cyst wall, the tentorium, optic nerve, and carotid artery were visible without any retraction of the brain. In a literature review by Robinson, he found that these lesions were often described as

subdural hygromas by previous authors and thought to be secondary to minor head injuries [20]. The problem with the trauma theory was that the brain tissue often appeared normal without displacement or compression. Robinson's impression of these cases was that the temporal pole was congenitally absent and called this developmental abnormality temporal lobe agenesis.

The theory of temporal lobe agenesis framed the pathophysiology of ACs as an underdevelopment of the temporal lobe as opposed to an abnormal development of the leptomeninges. Because many of these patients presented only with misshaped heads and nonspecific headaches, this theory ultimately contributed to decreasing the rates of surgery for the condition because there was no role for decompressive surgery if there was no compression of the brain. However, opponents to this theory argued that it could not easily account for the expansion of the skull. Other variants of agenesis of the brain were typically associated with a corresponding lack of skull development. Temporal lobe agenesis also could not easily explain the elevated intracranial pressure that was associated with some ACs. Today, the primary agenesis hypothesis has been largely abandoned, in favor of more widely accepted congenital leptomeningeal developmental theories.

12 Arachnoid Cysts in the Age of Modern Imaging

The 1970s revolutionized the treatment of neurologic conditions. In 1971, Hounsfield obtained the first images of the brain with a computed tomography (CT) scanner. Later in 1978, clinical teams at EMI laboratories lead by Ian Robert Young and Hugh Clow developed magnetic resonance imaging (MRI) imaging. These new imaging techniques allowed for the structure of the brain to be imaged for the first time. With the development and widespread adoption of these imaging techniques, ACs could be identified and diagnosed much more easily.

Modern imaging techniques have allowed for the distinct diagnosis of primary ACs from a variety of other pathologies with which they had historically been associated such as hemorrhages, hygromas, infection, tumors, and arachnoiditis.

Imaging also made it possible to start classifying ACs beyond just describing their anatomic location. In 1982, Galassi published his classification of middle cranial fossa ACs [12]. Middle cranial fossa ACs are thought to represent approximately half of all cranial ACs. Galassi classified these lesions into three types. Type I cysts were small, spindle-shaped lesions that were limited to the anterior portion of the middle cranial fossa, typically below the sphenoid ridge. These lesions were typically asymptomatic and did not require surgical intervention. Type II cysts extended superiorly to the Sylvian fissure and displaced the temporal lobe resulting in slowed flow of cerebrospinal fluid (CSF) in the subarachnoid space. Type III cysts filled the entire middle cranial fossa and often displaced not just the temporal lobe but also the frontal and parietal lobes. Type III cysts could present with midline shift and were associated with impaired CSF flow.

Modern imaging techniques and access to imaging led to easier diagnosis and the re-emergence of incidental asymptomatic ACs. Similar to the early literature, where postmortem studies first identified ACs, CT and MRI led to the discovery of a wide variety of small asymptomatic ACs located throughout the neuroaxis. Modern retrospective reviews of imaging have estimated the prevalence of ACs being found on brain imaging as 1.4% in adults and 2.6% in children [1, 2]. While this is a lower prevalence than reported in older autopsy studies, it likely represents a more homogenous group of pathologies.

13 Modern Presentation, Management, and Pathogenesis

Today, ACs are found throughout the brain and spine. They can still be classified as primary cysts which form due to congenital splitting of the arachnoid or as secondary cysts which form as a result of trauma, surgery, infection, hemorrhage,

or inflammation [11]. Due to prevalence and availability of imaging, over 90% of ACs that are discovered are asymptomatic and do not require any surgery [15].

ACs that are symptomatic can present with diverse symptoms depending on their size and anatomic location. Cranial lesions have been described presenting with obstructive hydrocephalus, headaches, elevated intracranial pressure, endocrinopathies from compression of the hypothalamic-pituitary axis, seizures, remodeling/deformity of the temporal bone and/or orbit, as well as cranial nerve and brain stem symptoms such as tinnitus, hearing loss, facial palsy, nystagmus, and vertigo. Some middle fossa ACs have also have been associated with acute and/or recurrent subdural hematomas and may present with focal neurologic weakness. Spinal ACs may be associated with pain or progressive neurologic symptoms such as weakness or paresthesias.

Treatment is typically recommended for symptomatic ACs. Although, on occasion, it may be difficult to determine if patients are symptomatic from their AC, there are three primary treatment options that are available: open microsurgical cyst resection and fenestration, endoscopic cyst fenestration, and cyst shunting. The optimal choice of treatment depends on a patient's symptoms and the location of the cyst. Subsequent chapters of this book will explore the various presentations of ACs as well as their treatment options in more details.

Modern pathogenesis of ACs remains quite similar to many of the early prevailing theories. Most common explanations given for ACs involve congenital, traumatic, and infectious/inflammatory etiologies. Given the diverse pathology of ACs in the nervous system, there are likely multiple pathways through which they can form. In large part due to modern imaging, we now have documented reports in the literature of ACs forming after traumatic injuries, infections, and inflammatory conditions as well as by spontaneous formation during early childhood development without any obvious inciting events [8, 24].

There is much that is still not understood of about the development of ACs. While there have been some studies that have linked different genetic polymorphisms to the development of both cranial and spinal ACs, more research is needed to understand how these lesions develop [4, 17, 18].

14 Conclusion

ACs represent a diverse pathology of lesions that can be found throughout the central nervous system. Throughout the years, a variety of pathophysiologic mechanisms have been associated with these lesions including congenital, developmental, traumatic, infectious, and inflammatory etiologies. Most of these theories have been present in some form or another since shortly after ACs were first described in the early nineteenth century.

The history of ACs can be divided into three primary eras: pre-surgical history, early surgical history, and the modern era (Fig. 4). While some of the nomenclature and terminology may have changed, many early observations of ACs still hold true. ACs can vary greatly in size and location. Most ACs are incidental findings and do not require any intervention. Decompression of symptomatic lesions can provide symptomatic relief. Future study into the imaging characteristics, pathology, and genetic expression of these lesions may help us understand more about their diverse pathology.

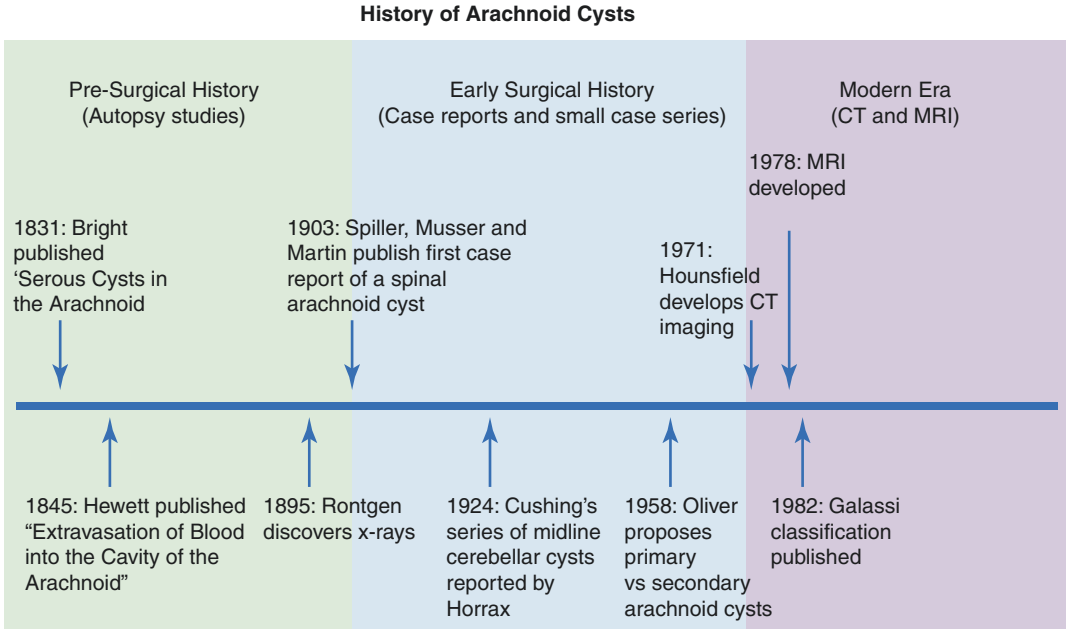


Fig. 4 History of ACs. Three distinct eras of literature can be found. Pre-surgical history primarily consists of autopsy studies of asymptomatic patients. Early surgical history consists of small case series of symptomatic patients. The emergence of computed tomography and magnetic resonance imaging marks the beginning of the modern era of management and classification of ACs

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Gender Distribution and Lateralization of Arachnoid Cysts

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1 Introduction

Cerebrospinal fluid (CSF) is a transparent serous liquid/fluid that provides essential mechanical hydrostatic and hydrodynamic medium for the central nervous system. Produced by the intraventricular choroid plexus, infilles the ventricular system, flows from the lateral ventricles, through Monroe foramina into the third ventricle, continues through the aqueduct towards the fourth ventricle and fluxes through the aperture mediana Magendy and lateral foramina Lushka, into the subarachnoid compartment spreaded on the skullbase, convexity of the brain and cerebellum and spinal canal, finalyresorpted in parasagitalPaccioni granules/bodies. The CSF fluxes through naturally creating a space formatted in subarachnoid cisterns and canals, comprising the subarachnoid compartment. Evolutionarily, it is created by the mesenchymal arachnoid—leptomeninga, carring the blood vesels. Physiologically, no nutritive, but hydrostatic, mechanical, gravity balancing role of CSF is defined.

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On that pathway, from the origin up to resorption, the CSF circulates through the subarachnoid compartment, driven by the capillarity and flow pressure gradient and provided by the pulsatile brain activity, providing medium for the blood vessels and neurologic elements, without direct/intimate encounter with the neurons of the brain, cerebellum, brain stem, and medulla. The separation/distinction is provided by the visceral arachnoid and pia mater, most deep intimate to the cortex leptomeninga.

By numerous, diverse influences, and etiological factors, as genetics, idiopathic, mechanical/physical, biological infectious, and chemical noxes, the normal CSF flux can be disturbed and compromised. This story is to depict the abnormal arachnoid collection defined as arachnoid cyst (ACs). The supreme delicate system of the subarachnoid compartment and CSF flux can be interrupted most frequently by mechanical means, aggravating, obstructing, and compromising the flux and finally accumulating similar to CSF, creating the morphologic issue, i.e., AC. The basic content is similar but not the same with CSF. Most likely, it is CSF plus protein, peptide, crystalloids, micro-blood pigment residua, or detritus of arachnoid desquamation. Mostly, they are occasional/incidental findings in the skull and spinal canal during neuroimaging studies. Thus, they can be primary, congenital idiopathic, or secondary provoked by trauma, i.e., mechanical rupture of the arachnoid, infection, and vascular and oncologic lesion, as well as a result of sur-

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gery, provoking stasis and accumulation of the CSF synergically with the pulsatile brain activity, locally creating valve mechanism obstruction, leading to the accumulation of the CSF-like content, defined as AC.

The natural evolution of the ACs as benign lesion is still not clear, with variety of dimension, location, distribution and variable degree of communication with the subarachnoid compartment.

Clinically, they can present with local compressive phenomenon, focal neurologic deficit, or more often symptoms of generally increased intracranial pressure (headache, nausea and vomiting, dizziness) or clinical signs of suboptimal cerebral function, i.e., epilepsy, mental disturbances, psychiatric disorders, dementia, impaired cognition, etc.

With the evolution of the diagnostic technologies, e.g., magnetic resonance imaging (MRI), certainly, the evidence-based knowledge concerning the predilection of AC location, side, and sex distribution emerged more unveiled and clear.

Definitely, the recent static and dynamic diagnostic high-technology procedures are essential tool for diagnosis, estimation, and evaluation of the ACs.

Furthermore, accurate analysis of the patient series, based on "double individual concept" comprising of the factors of the patient and factor of the therapist, has the mission of the most important conclusive prediction of the natural evolution, with or without active neurosurgical treatment of this phenomenon.

The basic neurosurgical questions—"what, when, and how"—should be answered by a high pool of accumulated direct and collateral systematized information comprising knowledge for the ACs.

Certainly, the statistical analysis of the gender distribution, location, and lateralization of the ACs should contribute to their surgical or not surgical solution, accomplishing the patient's well-being as our duty, which requires all of our commitment without distraction.

ACs represent fluid-filled duplications of the arachnoid membrane with a content which is similar but not equal to the CSF [45]. Another definition states that the ACs are loculated cavities within the arachnoid mater without discrimination of the wall or cyst content, meaning that the wall is composed by arachnoid cells connected with a normal arachnoid membrane [12, 14]. They do not communicate with the ventricles or subarachnoid space and may be unloculated or may have septations.

On the basis of their origin, the ACs can be primary (also known as "true") and secondary ACs. The primary ACs are considered to be congenital in nature, [4] nontumoral, extra-axial, intradural masses, characterized by a single layer of arachnoid cells covered by a vascularized collagen membrane in continuity with the arachnoid [11], while the secondary or acquired ACs result from a primary traumatic, hemorrhagic, tumoral infection (meningitis) or iatrogenic event that causes a prevalent inflammatory process leading to a loculation of the subarachnoid space [29, 30, 31, 38, 46].

ACs usually occur as sporadic and single malformations. However, the occurrence of bilateral or multiple ACs is not exceptional, especially in patients with metabolic disorders or syndromes [12, 28]. In particular, the occurrence of bilateral Sylvian ACs combined with macrocrania and cerebral palsy-like dystonia may suggest a condition known as glutaric aciduria type I (GAT1) [25, 26]. A recently identified gene located on the sixth chromosome (6q22.31-23.2), with an autosomal recessive pattern of inheritance, has been linked to the formation of congenital ACs. The gene was identified in a consanguineous family, and there did not appear to be a pattern in the location or size of the cysts [5]. Certain hereditary syndromes, such as Marfan syndrome [21], neurofibromatosis [27, 41, 47], glutamic aciduria type I [19], autosomal dominant polycystic kidney disease [3], and tuberous sclerosis [7], have demonstrated a higher incidence of ACs than the general population.

Population studies estimate that ACs make up approximately 1% of intracranial space-occupying lesions [9, 10, 36] and are found in approximately 1.7% of the adult population [1] but also represent the most common type of intracranial cyst in the clinical practice [8].

2 Lateralization and Frequency

There have been quite a few studies in the last two decades trying to estimate the prevalence of the ACs. Some large, hospital-based, and population-based studies have demonstrated a prevalence of ACs among the population of about 2%, varying from 0.23% to 2.6% [1, 2, 15, 34, 39, 40]. Some autopsy studies demonstrated a lower prevalence of ACs at 0.1% of individuals [37]. Two peaks in prevalence at 1 and 5 years (3.8% and 4.6% of pediatric cases, respectively) have been reported [2, 29]. Males are affected twice than females (M/F ratio: 2/1) [28]. It is more frequently diagnosed in the pediatric population where the prevalence of ACs is 2.6% [2].

The true incidence of ACs in the population can hardly be known, due to the possible asymptomatic course throughout life, which makes them hardly detectible, but also the different availability of radiodiagnostic tools in the different countries, which likely account for the different rates of their incidental recognition and the difficulty in obtaining specimens for the histological diagnosis, which may result in their underreporting [28].

3 Gender Distribution

A male predominance (2:1 males to females) is noted in both adults and children [1, 2, 29]. As described in a paper by Wester, ACs seemed to occur more frequently in males and on the left side. According to the author, this latter phenomenon could be explained by a greater significance attributed to symptoms from the dominant hemisphere as a justification for invasive procedures in the pre-computed tomography (CT) era. Based on CT studies, it has been determent there is a significant tendency for these cysts to occur in males, with a male/female ratio of nearly 3:1. This preponderance toward males could not be explained by the somewhat higher frequency of associated subdural hematomas that was found in male patients. The survey also showed that middle fossa ACs occur or are detected significantly more frequently on the left side than on the right, with a ratio of 1.8:1 [42].

However, in one of the largest cohorts, Al-Holou et al. studied a total of 48,417 patients who underwent brain MRI. ACs were identified in 661 patients (1.4%). Men had a higher prevalence than women ($p < 0.0001$). Multiple ACs occurred in 30 patients. The most common locations were middle fossa (34%), retrocerebellar (33%), and convexity (14%). Middle fossa cysts were predominantly left-sided (70%, $p < 0.001$). Thirty-five patients were considered symptomatic and 24 underwent surgical treatment. Sellar and suprasellar cysts were more likely to be considered symptomatic ($p < 0.0001$). Middle fossa cysts were less likely to be considered symptomatic ($p = 0.01$) [1].

In a study of pediatric ACs, Al-Holou et al. reviewed a consecutive series of 11,738 patients who were 18 years of age or younger during an 11-year period. In their study, 309 ACs (2.6% prevalence rate) were identified with an increased prevalence of ACs in males ($p < 0.000001$). Of those, 111 patients met all criteria for inclusion in the natural history analysis. The patients were followed for 3.5 years, after which 11 ACs increased in size, 13 decreased, and 87 remained stable. The authors concluded that younger age at presentation was significantly associated with cyst enlargement ($p = 0.001$) and the need for surgery ($p = 0.05$). No patient older than 4 years of age at the time of initial diagnosis had cyst enlargement, demonstrated new symptoms, or underwent surgical treatment, meaning that an older age at the time of presentation is associated with a lack of clinical or imaging changes over time [2].

4 Gender Distribution and Lateralization According to Location

On the basis of their location, the ACs can simply be divided into intracranial and spinal ACs. The intracranial ACs can further be subdivided into supratentorial and infratentorial, whereas the spi-

nal ACs can be divided into extraspinal and intraspinal cysts and may also be both intradural and extradural masses [11, 24]. A previous study on spinal ACs in the pediatric age group has demonstrated an association with neural tube defects, such as myelomeningocele and diastematomyelia [35].

In the era before CT and MRI, a literature review demonstrated 49% of cysts in the middle fossa or Sylvian fissure; in the cerebellopontine angle (CPA), 11%; quadrigeminal plate area, 10%; vermis, 9%; suprasellar area, 9%; interhemispheric fissure, 5%; cerebral convexity, 4%; and interpeduncular area, 3% [34].

About 80–90% of the ACs are located in the supratentorial space, including the quadrigeminal plate region [2, 12, 9] (see Table 1).

The great majority of them originates from the Sylvian fissure (50–60% of all intracranial ACs) and occupies the temporal fossa, followed by those arising from the chiasmatic and the quadrigeminal plate cistern. The interhemispheric fissure and the subarachnoid spaces of the convexity represent less common sites of origin. With regard to the infratentorial space, the lateral (CPA) and the midline retrocerebellar locations are quite equivalent, while the interpeduncular and prepontine cisterns are a rare site of origin of ACs [28].

Different from children, the retrocerebellar location is often found to be as common as the middle fossa location in adults [1]. While ACs of the middle cranial fossa are often discovered incidentally and tend to remain asymptomatic, those arising from other regions are more frequently associated with clinical signs and symptoms and are often associated with hydrocephalus [23]. Indeed, some ACs, namely, the suprasellar and the quadrigeminal plate ACs, may grow toward and even within the cerebral ventricles.

Wester et al. investigated 123 patients with 129 intracranial cysts, consecutively admitted to the Department of Neurosurgery in Bergen 1988–1997. Data were analyzed with regard to intracranial location and gender distribution. Cysts were much more commonly located in the temporal fossae than one would expect if the distribution were random; 68.1% of patients had temporal cysts. Temporal cysts were significantly more frequent in males than in females (3.9:1), while cysts of other locations did not show preponderance for a specific gender. The authors of this study described a connection between gender distribution and sidedness: the significant predominance of left-sided temporal cysts was found only in males. In patients with a unilateral temporal cyst, the left/right ratio was 2.0:1 (males 44 left and 20 right, females 8 left and 6 right) [44].

Table 1 Synopsis of the distribution of ACs and their related clinical findings

Location	Rate	Main clinical features
Supratentorial space	67–80%	Mainly asymptomatic, rupture, headache, seizures, developmental delay, bone scalloping
Sylvian fissure/middle fossa	50%	
Sellar/suprasellar region	10%	Hydrocephalus, headache, vomiting, macrocrania, hormonal deficit, bobble-head doll syndrome, visual loss
Interhemispheric fissure	5%	Macrocrania, headache, seizures, focal deficits
Convexity	5%	Headache, seizures, bone scalloping
Tentorial space	10%	Hydrocephalus, headache, vomiting, macrocrania, precocious puberty, Parinaud' syndrome
Quadrigeminal plate	10%	
Infratentorial space	10–23%	Cerebellar signs, hydrocephalus
Midline	8%	Vertigo, tinnitus, hearing loss, facial sensory loss or weakening, cerebellar signs
CP angle	10%	
Other (e.g., intraventricular, clival region/interpeduncular cistern)	2%	Hydrocephalus, brain stem signs, cranial nerve deficits

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In another study by Wester, 126 patients with 132 ACs were studied. The cysts had a strong predilection for the middle cranial fossa; 86 patients (65.2%) had cysts in this location. Of 106 cysts with clearly unilateral distribution, 64 were located on the left side and 42 on the right side. This significant difference resulted solely from the marked preponderance of middle fossa cysts for the left (left-to-right ratio, 2.1:1). There were significantly more males than females (92 males/34 females). This difference was exclusively due to male preponderance of unilateral middle fossa cysts (66 males/14 females, ratio, 4.7:1). For all other cyst locations, there was no difference between the two sexes (26 males/20 females) or the two sides (10 left, 16 rights). The marked left-sidedness for middle fossa cysts was found only in males. Females had an even distribution between the two sides. The author concludes that the strong predilection of ACs for the middle cranial fossa may be explained by a meningeal maldevelopment theory: the arachnoid coverings of the temporal and frontal lobes fail to merge when the Sylvian fissure is formed in early fetal life, thereby creating a noncommunicating fluid compartment entirely surrounded by arachnoid membranes. Why males develop more middle fossa cysts on the left side remains a mystery (Neurosurgery 45:775–779, 1999) [43].

A case series from The Johns Hopkins Hospital analyzed a group of 20 patients with ACs that underwent surgery. Of the patients enrolled in the study, 20% had supratentorial ACs, 5% infratentorial, and 25% spinal. Gender and age of the patients enrolled in the study were as follows: 12 male (60%) and 8 female (40%) patients, ranging in age from 2 weeks to 39 years (mean age of 10.9 years) at the time of surgery. Twenty-five percent of the cysts were Sylvian, 20% intraventricular, and 20% suprasellar. Supratentorial cysts were treated endoscopically in 73% of patients and with open resection in the remaining 27%. Symptoms at presentation included headache (41%), weakness (23%), seizure (14%), hydrocephalus (9%), scoliosis (4%), cognitive decline (4%), and visual loss (4%) [32].

4.1 Supratentorial ACs

Sylvian fissure ACs (also referred to as middle or temporal fossa cysts) are the most common type of intracranial cysts. Roughly, half of pediatric cysts are located in the middle fossa, and they tend to occur more frequently in males and on the left side (66%) [2]. Although slightly less prevalent in adults (34% of cases), the distribution between left and right is similar to children [1]. Helland and colleagues described a similar prevalence, sex, and side distribution but added that sex differences did not demonstrate statistical significance outside of the temporal fossa [20].

In a study performed by Helland et al., there was a strong predilection (198 patients [66.2%]) for intracranial ACs in the temporal fossa. Forty-two patients had cysts overlying the frontal convexity, 36 had cysts in the posterior fossa, and 23 patients had cysts in other different locations. Of 269 cysts with clearly unilateral distribution, 163 were located on the left side and 106 on the right side. This difference resulted from the marked preponderance of temporal fossa cysts on the left side (left-to-right ratio, 2.5:1; $p < 0.0001$ [adjusted <0.0005]). For cysts in the CPA, there was preponderance on the right side ($p = 0.001$ [adjusted = 0.005]). Significantly more males than females had cysts in the temporal fossa ($p = 0.002$ [adjusted = 0.004]), whereas in the CPA a significant female preponderance was found ($p = 0.016$ [adjusted = 0.032]). For all other cyst locations, there was no difference between the two sexes. The authors also conclude that there is a sex dependency for some intracranial locations of ACs, with temporal cysts occurring more frequently in men, and CPA cysts found more frequently in women. Furthermore, there is a strong location-related sidedness for ACs, independent of patient sex. These findings and reports from the literature suggest a possible genetic component in the development of some ACs [20].

In a landmark paper from 1988 by Galassi et al., ACs of the middle cranial fossa (also known as Sylvian cysts) were shown to represent the

most common type of intracranial leptomeningeal malformation. Among the 102 intracranial ACs that underwent surgery, 77 cases (75%) were located in the middle cranial fossa. The ACs reviewed in Galassi's paper had higher incidence in the first two decades of life (51 cases) as well as marked predilection for male sex (60 cases) and the left hemispheric location (55 cases). As for clinical presentation, cranial deformities, symptoms of raised intracranial pressure, and epilepsy constituted the most frequent features. In 13 patients, a complicating lesion was associated: subdural or intracystic hematomas in 7 cases, subdural hygromas in 4 cases, and extradural hematomas in 2 cases. Based on the appearance of the CT scan and the results of the CT cisternography, the authors proposed a classification into three basic types of increasing severity and different pathophysiologic conditions. All of the patients underwent craniotomy, excision of the cyst walls, and perforation into the basal cisterns. There was one postoperative death (mortality rate of 1.3%) due to meningitis. The remaining clinical results were gratifying in all three types of lesion; on follow-up CT scans, the cysts of type I and II exhibited a steady tendency to reduction or obliteration, while cerebral reexpansion seemed less evident in the third, most severe, type [17].

Although middle fossa or Sylvian fissure cysts remain the most common in all age groups, it is possible that some of the differences in prevalence reflect the likelihood of cysts in each location to cause symptoms. In their study of adults with radiographically diagnosed ACs, Al-Holou and colleagues found a statistically significant increased rate of symptomatic cysts for cysts located in the CPA, quadrigeminal cistern, sella, and ambient cisterns [1]. Middle fossa cysts in adults were associated with a significantly lower rate of symptoms [1].

4.2 Infratentorial ACs

The infratentorial compartment represents the second most common location of arachnoid malformations [18] and is often associated with hydrocephalus [22]. When evaluating ACs in the posterior fossa, it is often advised to first rule out a Dandy-Walker syndrome in doubtful cases and to obtain information about the CSF circulation, especially before undertaking more invasive diagnostic or therapeutic measures. It is particularly important to establish the presence and type of communication of cysts with the CSF pathways. Although infratentorial cysts often communicate, they can be space-occupying masses because of increasing CSF retention, which may be due to a ball-valve mechanism or to inadequate communication. The frequently associated hydrocephalus seemed to be dependent mainly upon mechanical factors [18]. A case series by di Rocco et al. evaluating eight children who have been surgically treated for posterior fossa AC have shown that intracranial hypertension was evident in six patients; two children were clinically regarded as being affected by "arrested" hydrocephalus [13].

In a retrospective study of 26 patients with posterior fossa cysts, Arai and Sato came up with four major findings: First, posterior fossa intra-arachnoid cysts were encountered more frequently than expected and were found to be surgically treatable. Second, although IV ventricular cysts were categorized as Dandy-Walker malformation, Dandy-Walker variant, and persistent Blake's pouch in this study, the distinctions of neuroimaging findings between these three types are uncertain. Third, the diagnostic criteria for mega cisterna magna were established, and it was found to be a surgically untreatable condition. Finally, in cases with the following neuroimaging findings, surgery appears to be indicated: (1) occipital bossing or petrosal scalloping with distortion or obliteration of CSF cisterns of the posterior fossa, (2) compression and deformity of the brain surrounding the cyst, (3) radioisotope and/or computed tomography cisternography findings suggestive of disturbance of intracystic CSF circulation, (4) a noncommunicating cyst [4].

Literature review revealed no recent and/or significant study evaluating lateralization and gender distribution of posterior fossa cysts.

4.3 Spinal ACs

The spinal ACs are quite uncommon compared to the intracranial ACs. A paper by Robb et al. reviews the spinal ACs in series of 11 pediatric patients. According to the same paper, the distribution of lesions was as follows: cervicomedullary (one patient), cervical (one), cervicothoracic (two), thoracic (four), lumbar (two), and sacral (one). Four of the 11 ACs (all intradural) were located anterior to the spinal cord, 3 of which were in children with a myelomeningocele. Only two of the cysts were extradural; both were found in the lumbosacral region, and one was associated with diastematomyelia [33].

In a study of 31 pediatric patients (median age 6.9 years) who underwent operative intervention for spinal ACs between 1992 and 2008, there were 17 female patients (55%) and 14 male patients (45%). Twenty-one patients (68%) presented with symptoms of radiculopathy or myelopathy. The most common presenting symptoms were pain (42%), lower-extremity weakness (39%), gait instability (32%), spasticity (19%), sensory loss (10%), and bladder dysfunction (7%). In three patients (10%), spinal ACs were incidental findings. Intradural spinal ACs were more common (18 patients, 58%) than extradural spinal ACs (11 patients, 36%). One patient (3%) had extradural and intradural components. One patient (3%) had a purely intramedullary cyst, and one patient (3%) had both an intradural and an intramedullary component. Of the 18 intradural spinal ACs, 9 (50%) were located ventral to the spinal cord, and 9 (50%) were dorsally situated. One dorsal intradural spinal AC had an intramedullary component. All extradural spinal ACs were located dorsal to the spinal cord. Intradural spinal ACs were primarily concentrated in the cervical and thoracic regions (67%), whereas extradural cysts were more evenly distributed between thoracic, lumbar, and sacral regions. Of the 18 patients with intradural spinal ACs, 13 (72%) had significant previous CNS abnormalities, compared with 3 (27%) of 11 patients with extradural spinal ACs [6].

In a study evaluating spinal ACs, Fam et al. identified 22 patients with spinal ACs with a mean age of at presentation 53.5 years. Of those 22, 17 patients were women, representing an almost 3:1 sex distribution. Symptoms comprised back pain ($n = 16$, 73%), weakness ($n = 10$, 45%), gait ataxia ($n = 11$, 50%), and sphincter dysfunction ($n = 4$, 18%). The mean duration of symptoms was 15 months. Seven patients (32%) exhibited signs of myelopathy. The spinal ACs identified in this cohort were located in the thoracic spine ($n = 17$, 77%) and less commonly in the lumbar spine ($n = 3$, 14%) and cervical/cervicothoracolumbar region ($n = 2$, 9%). Based on imaging findings, the cysts were interpreted as intradural spinal ACs ($n = 11$, 50%), extradural spinal ACs ($n = 6$, 27%), or ventral spinal cord herniation ($n = 2$, 9%); findings in three patients (14%) were inconclusive. Nineteen patients underwent surgical treatment consisting of laminoplasty in addition to cyst resection ($n = 13$, 68%), ligation of the connecting pedicle ($n = 4$, 21%), or fenestration/marsupialization ($n = 2$, 11%). Postoperatively, patients were followed up for an average of 8.2 months (range 2–30 months). Postoperative MRI showed complete resolution of the SAC in 14 of 16 patients [16].

5 Conclusion

Numerous evidence, both empirically and clinically tested, have proven that ACs are nearly twice as more common in males and on the left side, almost 50% in the Sylvian fissure.

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Embryology of Arachnoid Mater

Alpaslan Gökçimen

1 Introduction

Structures in the central nervous system (CNS) are protected by meninges, bone, and cerebrospinal fluid (CSF) [4]. The CNS is covered by two sheaths (meninges), which are layers of connective tissue that differ from each other in structure and function; because the outer sheath is hard, it is called "pachymeninges (dura mater)." Since the inner sheath is soft, it is called "leptomeninges (arachnoid mater and pia mater)" [1].

2 Histology and Location of Meninges Brain Tissue

Brain tissues are covered from the outside inward by three membranes: (1) dura mater (hard mother/coarse mother), (2) arachnoid (cobweb-like), and (3) pia mater (tender mother) (Fig. 1).

Dura mater is the strongest of these meninges. This layer is also called the pachymeninx (thick membrane). Outside the brain, the dura mater is continuous with the periosteum of the skull and has no epidural space. In the spinal cord, it is separated from the periosteum of the spine by the epidural space. This space includes loose connective tissue, adipose tissue, and venous plexuses [4].

Fig. 1 Structures surrounding the brain: structures surrounding the brain tissue are seen

While covering the cranial cavity, dura mater is observed in two parts as "periosteal dura

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