Chapter 5
Arachnoid Cysts

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5.1 Introduction

Arachnoid cysts (AC) could be located either cranial or spinal. ACs are present in around 2.6% of the population [1]. The authors reviewed 11,738 consecutive MRI studies of a pediatric population aged between 0–18 years and found in 309 arachnoid cysts to be present. The male to female ratio was 1.8:1. Most frequent was a location in middle fossa (49%), followed by posterior fossa (38%), quadrigeminal plate (6%), convexity (4%), sellar-suprasellar (2%), anterior fossa (2%), interhemispheric (1%), and intraventricular (0.3%). There was a preference noticed for side in only left middle fossa with a ratio of 1.7:1 [2]. Over a mean follow-up of 3.5 years of 111 ACs, 11 (9,9%) increased in size (three becoming symptomatic), 13 (11.7%) diminished, and 87 (78.4%) were unchanged. The younger the patient at diagnosis the more likely the need for an operation [1].

The first reports of successful treatment of spinal intradural cysts were reported by Spiller in 1903 [3] and Skoog in 1915 [4]. Later, case reports described rarities which were fatal if it located in the upper cervical medulla or at the craniocervical junction [5]. Spinal AC are very infrequent in all age groups. Age and gender do not play a role in the incidence of AC in the first two decades of life. The incidence of diagnosed AC has increased, very likely due to the better quality of magnetic resonance imaging (MRI). The introduction of 3 Tesla MRI has provided further anatomic resolution allowing better preoperative planning [6].

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5.2 Classification

5.2.1 Cranial ACs

AC can be classified as to location, size, and etiology. Location incidence, as previously mentioned mainly in the middle and posterior fossa with a wide distribution of the rest. The middle fossa AC have been classified regarding their size into small, moderate, and large (Galassi I, II and III respectively) [7]. A recent classification has been proposed for suprasellar AC [8]. Most ACs are primary. Secondary ACs are related to infection, previous operation, hemorrhage, trauma, and metastatic disease. The incidence of secondary AC is not well reported.

5.2.2 Spinal ACs

The classification of spinal ACs is more complicated, a study simplified the classification of spinal meningeal cysts into three major categories: extradural cysts without nerve root fibers (Type I) subdivided into IA-extradural arachnoid cysts and IB-sacral meningoceles or occult meningoceles; extradural cysts with nerve root fibers (Type II); and intradural cysts (Type III) [9]. Spinal meningeal cysts are most often located in the mid- to lower thoracic area [6, 10, 11], and are found predominantly in males, and tend to be symptomatic during the second decade of the patient's life [10, 11]. The arachnoid cysts in the sacral spinal canal can enlarge the bony canal, but are distinctly different from closed neural tube defects in which the dura mater extends beyond the confines of the spinal canal through a deficit in the posterior vertebral arches [12]. It is suspected that some of the intradural AC in patients with an open neural tube defect were secondary to infection. Those AC associated with a split cord malformation are more likely to be congenital.

5.3 Relation between Arachnoid Cysts and Hydrocephalus

Cranial ACs can cause blockage of the internal cerebrospinal fluid (CSF) pathways and thereby cause hydrocephalus [13]. An example is in case of suprasellar arachnoid cysts. Unlike other arachnoid cysts, these are usually associated with continuous increase in size. This occurs as the basal membrane around the basilar artery exerts a valve mechanism action and with every arterial pulsation more CSF is pushed into the cyst that may not flow out again. The growth occurs thereby slowly but continuously and takes a long time to cause compression of the third ventricle and occlusion of the aqueduct forming a classic occlusive hydrocephalus picture

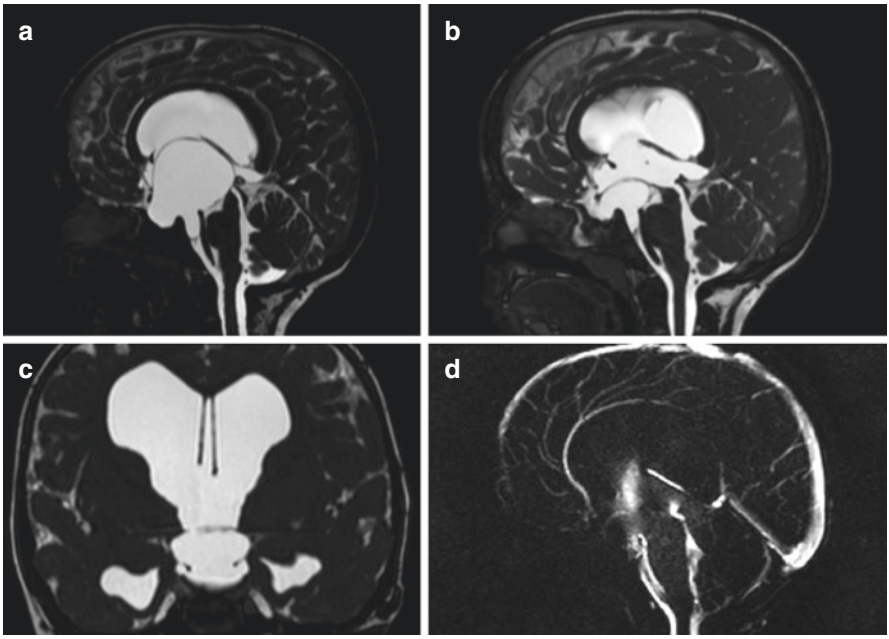

Fig. 5.1 (**a**) Midsagittal CISS MR image showing the suprasellar AC with aqueduct occlusion. (**b**) Midsagittal CISS MR image 4 years after surgery showing decrease in size of AC and free aqueduct. (**c**) Coronal CISS MR image showing the fenestration of the roof of the AC. (**d**) CINE Phase MR Image showing the flow at roof and floor of AC after successful ventriculocystocisternostomy

(see Fig. 5.1). 17–30% of all cranial ACs had an increase in intracranial pressure causing macrocephalic changes [14]. Hydrocephalus is more common with cysts of the midline and posterior fossa [15, 16]. The aim of therapy is always the fenestration of the cysts to the normal CSF spaces to achieve decompression of the cysts and at the same time a reopening of the regular CSF pathways. The implantation of a shunt should be primarily avoided as the problem is not resorptive, but rather the distorted intracranial CSF communication. For example, in cases of retrocerebellar ACs with compression of the cerebellum and 4th ventricle ventrally causing hydrocephalus, the operative choices are either a fenestration to the cisterna magna or to establish a communication to a lateral ventricle with a wide fenestration with or without stent placement. Other rare situations are septum pellucidum cysts (see Fig. 5.2) with symptoms of a periodic increase in intracranial pressure but not clearly hydrocephalic [17, 18], and intraventricular cysts [19]. In these cases, the experience of the surgeon plays a major role in the surgical decision. Basically, an endoscopic fenestration to the ventricular system is recommended, if there is local compression effect or obstruction of the CSF pathways.

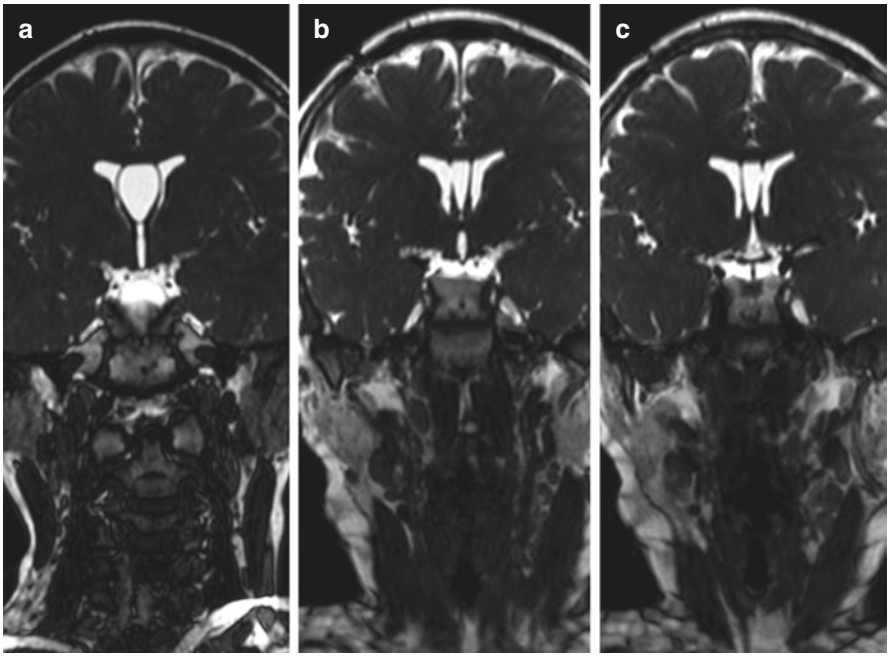

Fig. 5.2 (**a**) Coronal CISS MR image showing the septum pellucidum cyst with narrowing of both foramena of Monroi. (**b**) Coronal CISS MR image 2 years after surgery showing the fenestration of the right leaflet. (**c**) Coronal CISS MR image 2 years after surgery showing the fenestration of the left leaflet

5.4 Clinical Presentation

5.4.1 Cranial ACs

In infancy, the presence of an intracranial AC is often accompanied by an abnormal increase in head circumference secondary to enlargement of the AC or secondary to CSF flow obstruction [20]. Rare finding is an outward bowing of the calvarial bone overlying the arachnoid cyst due to chronic longstanding local pressure. The most common symptoms relate to raised intracranial pressure are headache, nausea, vomiting, lethargy, and papilledema, followed by abnormal increase in head circumference. Less common symptoms are seizures, cerebellar signs, cranial nerve deficits, hemiparesis, visual disturbances, and endocrinopathy according to its location and the nearby structures affected through local space occupying effect of the AC. A number of reports have noted that rupture or hemorrhage can occur into an AC or the adjacent subdural space spontaneously or after mild head trauma [21]. The incidence of seizures as a presenting symptom is reported to be in the range of 5%–20% [1, 22, 23]. Abnormal electrical activity if present may or may not relate to the location of the AC. Koch et al. simply stated that: “Arachnoid cysts are congenital cystic brain malformations associated with epilepsy”[24].

5.4.2 Cranial ACs and Cognitive Impairment

Eleven studies have subjected relatively large series of symptomatic AC patients to systematic neuropsychological investigations and one additional study have looked at mental functions in elderly asymptomatic patients. Ten of these studies reported significant mental impairment, mostly, but not solely, in cognition, and the seven that report both pre- and postoperative results found a clear postoperative normalization. These data highlight the importance of using neuropsychological tests in these patients, as suggested by Soukup et al. [25], that the cognitive measures may provide an alternative functional index of outcome efficacy, rather than reliance on the traditional outcome measures (i.e. anatomical decompression or resolution of clinical symptoms) as they may underestimate the efficacy of surgical intervention for these patients. The present studies indicate that ACs indeed affect mental functions and that they do so in a reversible manner. The preoperative clinical complaints seem to be not associated with the size of the cyst but with the intracystic pressure; the higher the pressure, the stronger the complaints [26]. Another common misconception is that the postoperative improvement must be correlated with the postoperative cyst volume reduction.

Most ACs are considered congenital; therefore, the effects exerted by the cyst pressure upon the surrounding brain tissue have been lifelong. There have been some prospective studies suggesting that the pressure from the cyst on the surrounding brain parenchyma does not necessarily cause a permanent destruction of brain tissue, but more likely a reversible suppression of brain functions, which probably is associated with disturbed perfusion. This explains the postoperative cognitive improvement as the perfusion of the involved brain structures is normalized after the pressure from the cyst is removed. Additionally, structural neuroimaging studies have shown that the temporal lobe adjacent to an AC is smaller and less metabolically active than the contralateral temporal region [27], that language areas within the left hemisphere is displaced by an AC, but not to the contralateral hemisphere [28], and that there is a thinning of cortical tissue around an AC [29]. Most importantly, it has also been demonstrated that a cyst may reduce the perfusion and metabolism in the surrounding cortical regions [14, 30–35], and that these changes are reversible after the cyst has been decompressed, hence explaining the cognitive improvements seen in the same patients. These findings are important, as they clearly demonstrate the association between a functional improvement and a normalized metabolism through improved perfusion in the corresponding cortical areas following cyst decompression.

5.4.3 Spinal ACs

In case of spinal AC, the usual clinical presentation is either myelopathy, radiculopathy, or both with the symptoms usually being insidious and very rarely acute. The presenting symptoms in order of decreasing frequency were: pain, lower

extremity weakness, gait disturbance, scoliosis, spasticity, sensory loss, and a neurogenic bladder [36]. An AC presenting as scoliosis is often associated with other symptoms whereas scoliosis secondary to syringomyelia more often has little or no neurological deficit. As the thoracic spinal canal is the longest and smallest in diameter, AC in this location may manifest earlier than those in the cervical or lumbosacral region.

5.5 Diagnosis

5.5.1 Cranial ACs

MRI is the gold standard for diagnosis. A very common differential diagnosis is the enlargement of the anterior temporal subarachnoid space (SAS) which is a normal common variant as is the size of the cisterna magna. Two factors help in determining whether the finding is an AC, the first being displacement of adjacent structures (i.e., mass effect), and the second is the presence of signal flow voids on MRI-T2 imaging that may indicate communication with adjacent CSF spaces. A new MRI technique, Time-SLIP, developed from modification of arterial spin labelling, has the ability, with videos, to show qualitative CSF flow between adjacent CSF spaces [37, 38]. This technique can help determine if an enlarged SAS at the anterior temporal region or a large cisterna magna is in communication with the surrounding SAS, thereby confirming the presence or absence of an AC. This MRI sequence can also establish the patency of an AC fenestration in further follow up.

As prenatal ultrasound studies are now part of routine monitoring during pregnancy, AC could be detected in utero with two-thirds of AC during the second trimester and the remaining one-third in the third [39]. The size of the AC rarely increases disproportionately to fetal growth. Unless the AC is associated with an incidental CNS malformation, the prognosis for normal neurologic development is excellent. Size and location of the AC are most often not major factors except in case of suprasellar location, in which hydrocephalus, visual impairment, and endocrinopathy can occur. It is rare for hydrocephalus to develop secondary to AC in non-suprasellar locations.

5.5.2 Spinal ACs

The use of MRI, especially with 3T machines, makes the diagnosis of spinal AC relatively clear. Contrast enhancement may be indicated in the first study only but is infrequently needed subsequently. Most useful are the T2-weighted images, as they visualize AC and delineate signal flow artifact that indicate CSF movement and signal intensity changes within the spinal cord. The recently introduced MRI

technique to visualize CSF communication to nearby CSF spaces is very beneficial. CT is infrequently needed specially in pediatric population due to long term hazards of radiation, but if done, can help delineate such bony changes as enlargement of the spinal canal, pedicle erosion, and increase in foraminal size, findings that usually are associated with extra- and not intradural AC. CT myelograms are only rarely needed, but can be useful in establishing patency between CSF spaces, especially in the presence of spinal instrumentation that produces a significant artifact on MRI and CT.

Intramedullary AC are very rare. The origin of this form of AC is not known nor is the mechanism of its enlargement. These AC are reported to present with progressive quadriparesis or paraparesis with pain not being mentioned as a prominent feature. The differential diagnosis includes syringomyelia or a spinal cord tumor associated cyst, both of which should be distinguishable by MRI.

5.6 Indications for Treatment

5.6.1 Cranial ACs

An AC can produce displacement of the adjacent brain especially in the middle fossa where an AC can become quite large. The question arises if such displacement/compression has long term effects on neurologic functions especially in an infant. If so, then diminishing the size of the AC could be postulated to improve long term neurologic function [33, 40–42].

In a recent study by Mørkve et al., adults who underwent surgery for fenestration of a middle fossa arachnoid cyst were given multiple questionnaires to evaluate if their quality-of-life improved following fenestration. Cyst size before surgery and its reduction thereafter had no correlation as to outcome. The majority of patients indicated that their quality of life was improved. The authors of this study thought that headache, dizziness, and cognitive impairment were significantly improved enough to outweigh the risk of operative intervention. We still need more studies in order to substantiate this hypothesis [40].

5.6.2 Spinal ACs

In case of spinal AC, the decision to treat is mainly depending on symptoms and the dynamic change in size noticed, if any. The decision to make surgery is therefore relatively easier in cases of spinal ACs due to the usual presentation with symptoms as well as the definite mass effect exerted on the spinal cord regarding the small spinal canal in comparison to the cranial cavity.

5.7 Operative Treatment

5.7.1 Cranial ACs

Surgical treatment of arachnoid cysts has been performed using both open [23, 26, 40, 41, 43–45], endoscopic techniques [43, 46–49] and also using various shunting procedures [43, 50, 51]. Successful treatment, regardless of approach, requires fenestration of the cyst wall creating communication between the cyst and the normal SAS (see Fig. 5.3) or an insertion of a cystoperitoneal shunt, thus relieving pressure from the cyst on the surrounding tissues. No randomized controlled trial has been undertaken to compare these different surgical approaches, and the choice of surgical procedure for a particular patient might differ from center to center based on experience and preference of the treating surgeon.

Previous decades have debated whether shunting or craniotomy was the optimal method to treat AC. Most of the current literature regarding treatment of AC is comparing craniotomy versus endoscopic techniques [23, 48, 52–54]. Taking into consideration the concept of minimally invasive neurosurgery, the less the operative manipulation the better, thus favouring the increasing use of endoscopy [55]. Despite our preference of endoscopy, recent studies comparing shunting, craniotomy, and endoscopy found no difference in outcome or complications [43, 52].

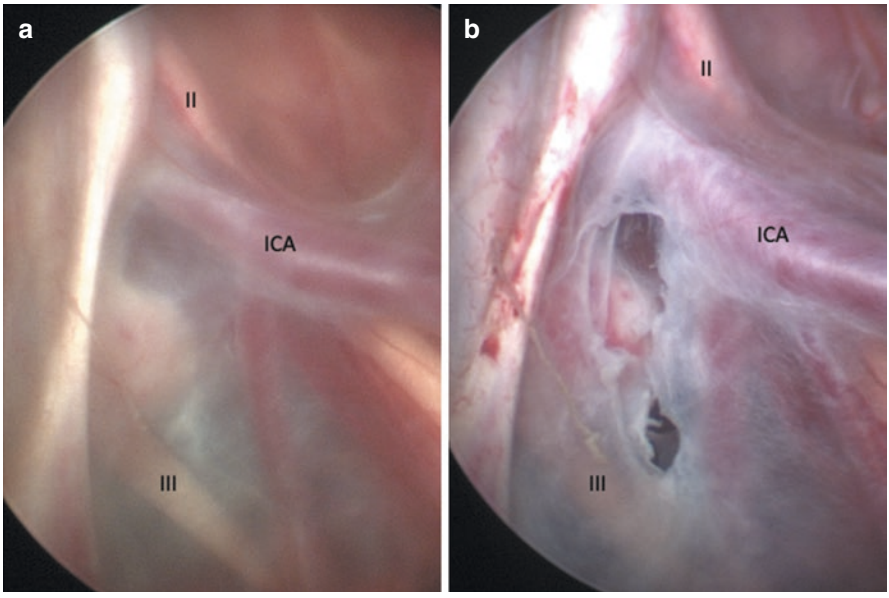

Fig. 5.3 (**a**) Endoscopic view of medial wall of perisylvian arachnoid cyst showing optic nerve (II), internal carotid artery (ICA) and oculomotor nerve (III). (**b**) Endoscopic view after successful fenestration of the cyst to the basal cisterns

Others have proven a clear-cut advantage when operating an intraventricular AC [56]. Shunting may have more long-term complications which will be discussed in this chapter later.

An individualized treatment strategy should also be applied when it comes to selecting patients for surgical treatment. Whether or not patients with arachnoid cysts should be operated, has long been a matter of controversy [40, 44, 57–61]. In these patients surgery is usually not lifesaving, but rather recommended aiming to reduce symptoms and increase quality of life, and thus should only be undertaken if the risk of complication is very low. Complication rates in surgical series have been described in 6% up to 20% [23, 40, 44, 49, 50]. This has led several authors to prefer a conservative approach [59, 60] and although most authors now would agree that symptomatic arachnoid cysts necessitate surgical treatment [40, 42–44], some reserve surgery for patients with overt symptoms of hydrocephalus, raised intracranial pressure, or other objectively verifiable symptoms [59]. A clear reduction in headache and dizziness have shown a significant improvement in patient's quality of life [40].

In case of objective symptoms of hydrocephalus, raised intracranial pressure, or focal neurological deficits that are to be expected through a potential space occupying effect, the indication and decision to offer surgical treatment is fairly straightforward. In cases where all or most symptoms are non-specific, however, it is the responsibility of the treating surgeon to identify the patients with symptoms severe enough to justify the risk of undergoing intracranial surgery. We recommend performing a neuropsychological assessment, especially in the pediatric age group, prior to surgery in order to be able to quantify an improvement after surgery if done. Regardless of which methods are applied for assessment, however, the patients and their parents in case of children must always receive unbiased and quantified information about risks and potential benefits of both surgical treatment as well as conservative management before they themselves make the final decision whether the symptoms are debilitating enough that they accept the calculated risks of undergoing surgical treatment or not.

5.7.2 Spinal ACs

As almost all extradural AC reported in the literature were dorsally located and thus, surgically accessible. Usually, the wall of the AC can be separated from the dura mater. The important feature is assuring complete closure of the communication between the SAS and the extradural cyst. This usually can be done with suturing alone and, if needed, reinforced with fat or a dural substitute. As 50% of the intradural AC are dorsally located, they are more readily fenestrated than those located anterior to the spinal cord. The other half of intradural AC is anterior to the spinal cord and more of a surgical challenge (see Fig. 5.4). Usually after opening the dura, the spinal cord bulges upward through the dural opening. The aim is to go laterally to expose the

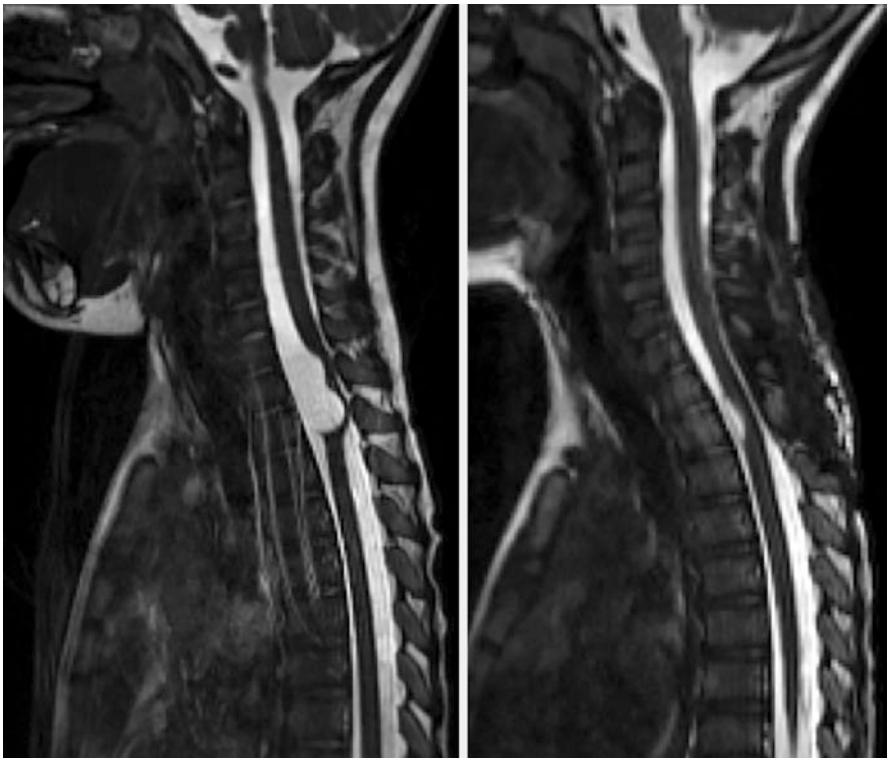

Fig. 5.4 Left: Sagittal CISS MR image of the cervical spine in a 3 years old child showing a ventrally located intradural arachnoid cyst. Right: Sagittal MR image 6 months after successful microsurgical fenestration through a 3-level laminoplasty

wall of the AC which is opened allowing drainage of CSF. After release of the pressure inside, exposure of the cyst would become easier specially after cutting the dentate ligaments and mobilising the spinal cord further medially. Additional areas should be excised on both lateral sides of the spinal cord by going between nerve roots. Intramedullary AC are almost in reality syringomyelia. If the cyst is very large and is surrounded by a thin rim of spinal cord, an approach would be to do a myelotomy in the region where the rim of spinal cord tissue is the thinnest with or without placing a stent to drain the syrinx continuously to the spinal SAS. Somatosensory and motor evoked potentials are routinely used for fenestration of AC. Nerve root stimulation might be of some benefit when dealing with AC surrounding an exiting nerve root.

5.8 Postoperative Complications

A wide spectrum of complications can occur with or following any cranial operative procedure, whether craniotomy or endoscopy, any and is dictated to a degree by the location of the AC. The most common complication related to the disease itself is impairment of CSF circulation that leads to having to do another procedure to treat

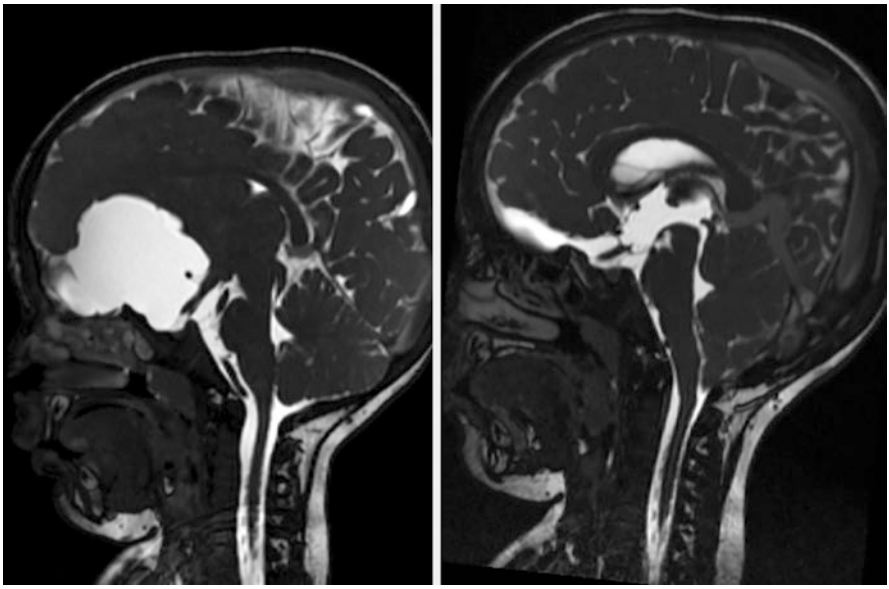

Fig. 5.5 Left: Midsagittal CISS MR image of a one-year-old child with huge perisylvian arachnoid cyst. Right: Midsagittal CISS MR image one year after insertion of a cystoperitoneal shunt showing the tonsillar herniation and the engorged venous sinuses

the hydrocephalus. A study has noted that this complication is much higher in infants under 2 years of age [20]. Other complications associated with CSF diversion via a shunt are also possible. Cystoperitoneal (CP) shunting was for decades the preferred treatment option due to its familiarity in neurosurgical practice [62]. In addition to other common complications of CSF shunting (infection, obstruction, disconnection, etc.), CP shunt insertion is also capable of producing shunt dependency [63, 64]. Current publications have clearly defined the hazards derived from CP shunt utilization, particularly those related to excessive drainage, for example, orthostatic headache, slit cyst syndrome [64], posterior fossa crowding [65], craniocerebral disproportion [65–67] and acquired (pseudo) Chiari malformation [68, 69].

Few studies tried to explain the cascade of these complications. The primary event involving overdrainage in shunted AC seems to be CSF hypotension. After cyst shunting, the cerebral ventricles initially enlarge and are displaced toward the cyst while CSF is drained. Later on, the brain aims to fill up the space left by the decompressed cyst. The intracranial venous system becomes dilated and engorged producing meningeal congestion that, in turn, will evolve to causing meningeal and sutural fibrosis. Subsequently, the skull bones thicken by inward apposition of bone and the paranasal sinuses expand to fill the gap due to volume depletion and to diminution of ICP and cerebral pulse pressure. These osseous changes also affect the posterior fossa leading to overcrowding and to tonsillar herniation (see Fig. 5.5). All these features contribute to reduce CSF reabsorption too. Finally, craniocerebral disproportion occurs when the skull becomes rigid which is usually irreversible in nature obliging the neurosurgeon to undertake more radical surgeries, such as decompressive and expanding procedures [65, 70].

To conclude, at first glance, CP shunting seems to be a safe procedure with which most neurosurgeons are familiar. However, on long terms CSF volume depletion, together with the resultant reduction in cranial capacity, leads to a cascade of events that are responsible for shunt dependency. Changes in cerebral CSF flow, brain, meninges, veins, and venous sinuses, and finally thickening of the skull bones produce a constellation of the overdrainage syndromes. All these conditions are difficult to manage and each one of them requires an individualized treatment. Hence, nowadays most neurosurgeons try to avoid placing CSF shunts and to resort to using microsurgical techniques or neuroendoscopic procedures for cyst fenestration instead.

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