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Intraventricular congenital lesions and colloid cysts

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Intraventricular congenital lesions and colloid cysts comprise a rather large spectrum of different pathologic conditions, which are outlined in the following article. Treatment is not warranted in most cases unless there is progressive ventricular obstruction with hydrocephalus or growth of the lesion itself, making tissue biopsy and histopathologic diagnosis necessary. Accordingly, a precise neuroradiologic evaluation is of the utmost importance, because most lesions, if not symptomatic, only require clinical and radiologic follow-up.

Developmental midline intracranial cysts constitute a separate entity and are usually associated with malformation disorders of the brain. They can be intraventricular, paraventricular, or intraarachnoid in location. Detailed evaluation with neuroimaging is beneficial because it directly affects subsequent management. The presence of communication with the ventricular system, associated hydrocephalus, and mass effect are features that help the neurosurgeon to plan appropriate treatment.

Neurosurgical techniques, such as endoscopy, allow a more direct approach for fenestration of either the cyst or the ventricular system if there is associated hydrocephalus. The move toward these less invasive methods of treatment demands detailed anatomic roadmaps that can be provided by appropriate neuroimaging.

Before we can fully appreciate congenital intraventricular lesions, it is important to recognize cavities that exist as normal variants, such as the cavum septum pellucidum (CSP), cavum vergae, and cavum veli interpositi (CVI).

Embryology

The neural crest begins to form at 3 weeks of gestation of the trilaminar embryo (containing ectoderm, mesoderm, and endoderm) by folding of the neural plate into the neural tube. The anterior neuropore closes at 18 days of gestation; by the end of the fourth week, the primary brain vesicle exists and the hindbrain flexures outline the primary divisions of the brain. The forebrain divides into two secondary vesicles during the fifth week, forming the telencephalon and the diencephalon. With further invagination of the telencephalon and growth of the cerebral hemispheres in a dorsorostral direction, the frontal, temporal, and occipital poles form around the diencephalon during the seventh week. The lateral ventricles follow accordingly and are drawn from the frontal pole to the temporal pole in a C-shaped manner.

A vascular layer of the pia mater fuses with the ependyma to form the tela choroidea, which subsequently invaginates into the ventricles through the choroidal fissure as the choroid plexus is developing. Cerebrospinal fluid (CSF) production starts, and, finally, in the second trimester, the thin roof of the fourth ventricle bulges and

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ruptures in three locations, giving rise to the foramina of Luschka and Magendie and allowing CSF access into the subarachnoid space.

Normal variants

Cavum septum pellucidum

As a single midline structure, the septum pellucidum separates the two anterior horns of the lateral ventricles (Fig. 1). The CSP is demarcated by the genu of the corpus callosum anteriorly , by the columns and body of the fornix posteriorly, by the body of the corpus callosum superiorly, and by the rostrum of the corpus callosum inferiorly. It consists of an ependymal lining toward the ventricles and contains neuronal and glial cell elements. These cell elements have connections to the hypothalamus and the hippocampus. At birth, the two layers of the septum pellucidum are separate and enclose a cavum. Later in life, these two layers typically fuse into a single septum. Autopsy and imaging studies have shown that all premature infants and 97% of term infants have a CSP, with the incidence dropping to 41% by 3 months of age and to 15% by 6 months of age [1]. The CSP may remain dilated in the context of some congenital disorders with arrest of normal brain development or may secondarily enlarge with repetitive brain trauma, such as in boxers. In such instances, obstructive hydrocephalus results from compression at the foramina of Monro and may require neurosurgical treatment.

Cavum vergae

If the layers of the septum pellucidum posterior to the columns of the fornix do not merge, they leave a cavum vergae, which is commonly seen in combination with a CSP (see Fig. 1) [2]. It is not clear whether the cavum vergae is the posterior portion of the CSP or whether it develops independently and communicates with the CSP. The cavum vergae is bordered by the body of the corpus callosum superiorly, by the hippocampal fissure inferiorly, by the crus of the fornices laterally, and by the splenium of the corpus callosum posteriorly. This anatomic variant is present in about one third of newborns and persists only rarely until adulthood. Interestingly,

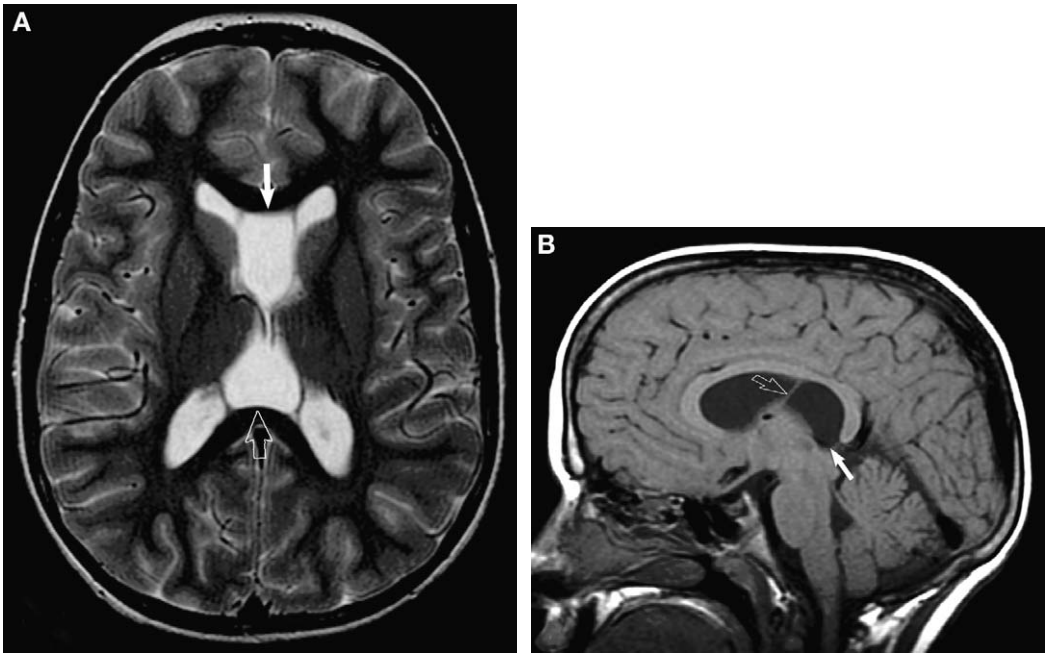

Fig. 1. Cavum septum pellucidum (CVP) and cavum velum interpositum (CVI). (*A*) Axial T2-weighted image demonstrates the CVP anteriorly (*arrow*) and CVI posteriorly (*open arrow*). (*B*) Sagittal T1-weighted image demonstrates anterior and superior displacement of the fornix (*open arrow*) distinguishing the CVI from the cavum vergae. Note characteristic inferior displacement of the internal cerebral veins (*arrow*).

the cavum vergae disappears before the CSP. Cystic enlargement of the cavum vergae may cause hydrocephalus by obstruction of either the foramen of Monro or the body of the lateral ventricle.

Cavum veli interpositi

This rare variant develops through an anterior extension of the pia-arachnoid membrane that arises from the quadrigeminal plate cistern. The CVI is situated between the crus of the fornices and lies inferior to the hippocampal commissure and the corpus callosum and superior to the roof of the third ventricle [3]. The CVI may extend as far as the columns of the fornix. It is formed from a double layer of pia mater, the tela choroidea, which covers the ependymal roof of the third ventricle, and results in fluid accumulation within the potential space of these two layers when the posterior end of the tela choroidea remains open. The internal cerebral veins and the medial posterior choroidal artery lie within the two layers and can be displaced by cystic expansion of the CVI inferolaterally. Cystic enlargement of the CVI requiring treatment is exceptional, with only a few case reports in the literature [4,5].

Intraventricular cysts

The development of intracranial cysts, especially those in the midline, can occur in isolation or may be associated with further anomalies of the brain, such as dysgenesis of the corpus callosum, and incomplete forebrain cleavage within the spectrum of holoprosencephaly [6,7]. Midline cysts may be composed of ependymal, glioependymal, epithelial, neuroepithelial, or true arachnoid features.

There are several classification systems of interhemispheric cysts, with most being based on the communication with the ventricular system and location in relation to brain parenchyma. More recently, interhemispheric cysts have been classified according to a combination of criteria, including the presence of further developmental anomalies or clinical characteristics [8,9].

Colloid cysts

Third ventricular colloid cysts constitute 0.5% to 1% of all intracranial mass lesions and arise from the anterior roof of the third ventricle. There is a slight male predominance [10] and a predilection for the third and fourth decades of life. The clinical symptomatology may vary considerably. Sudden death as the result of acute obstruction of the foramen of Monro has been reported [11]. Most patients present with headaches that have a sudden onset with change in head position. An explanation for this phenomenon is acute ventricular obstruction from cyst occlusion of the foramen of Monro. The bobblehead doll syndrome typically described in the context of colloid cysts has the same underlying pathophysiology [12]. Because these cysts are located between the columns of the fornix, an association with short-term memory deficits is not uncommon [10].

Colloid cysts are filled with thick gelatinous fluid that is rich in cholesterol and contains calcifications. They are typically hyperdense on CT and hyperintense on T1-weighted images (Fig. 2) [13]. The cyst appearance on MRI can vary considerably, and the signal intensity is directly related to the viscosity of the cyst, which can be helpful in deciding whether stereotactic aspiration or open craniotomy for removal should be performed. It is not unusual to find colloid cysts at autopsy as an incidental finding; however, those individuals who become clinically symptomatic require surgical intervention. Definitive cure can be achieved only by microsurgical resection either by an endoscopic route or via open transcallosal or transcortical resection [14]. Cyst aspiration alone carries the risk of reaccumulation of cyst material with subsequent re-expansion of the cyst and recurrence of symptoms. Memory impairment as a consequence of endoscopic or open surgery is not an uncommon postoperative phenomenon caused by trauma to the fornices. The callosotomy should not be longer than 2.5 cm and should remain in the anterior third to avoid disconnection syndromes. Seizures can occur after a transcortical route, and sagittal sinus thrombosis is a feared complication, albeit rare, after a transcallosal approach [10].

Arachnoid cysts

Arachnoid cysts occur most frequently in the middle cranial fossa (30%–50%) and less commonly over the convexity (10%), in the suprasellar region (10%), in the quadrigeminal plate (10%), in the cerebellopontine angle (10%), or in the midline posterior fossa (10%). In rare cases, these cysts may occur unrelated to the cerebral cisterns, such as in the ventricles or the diploë of the

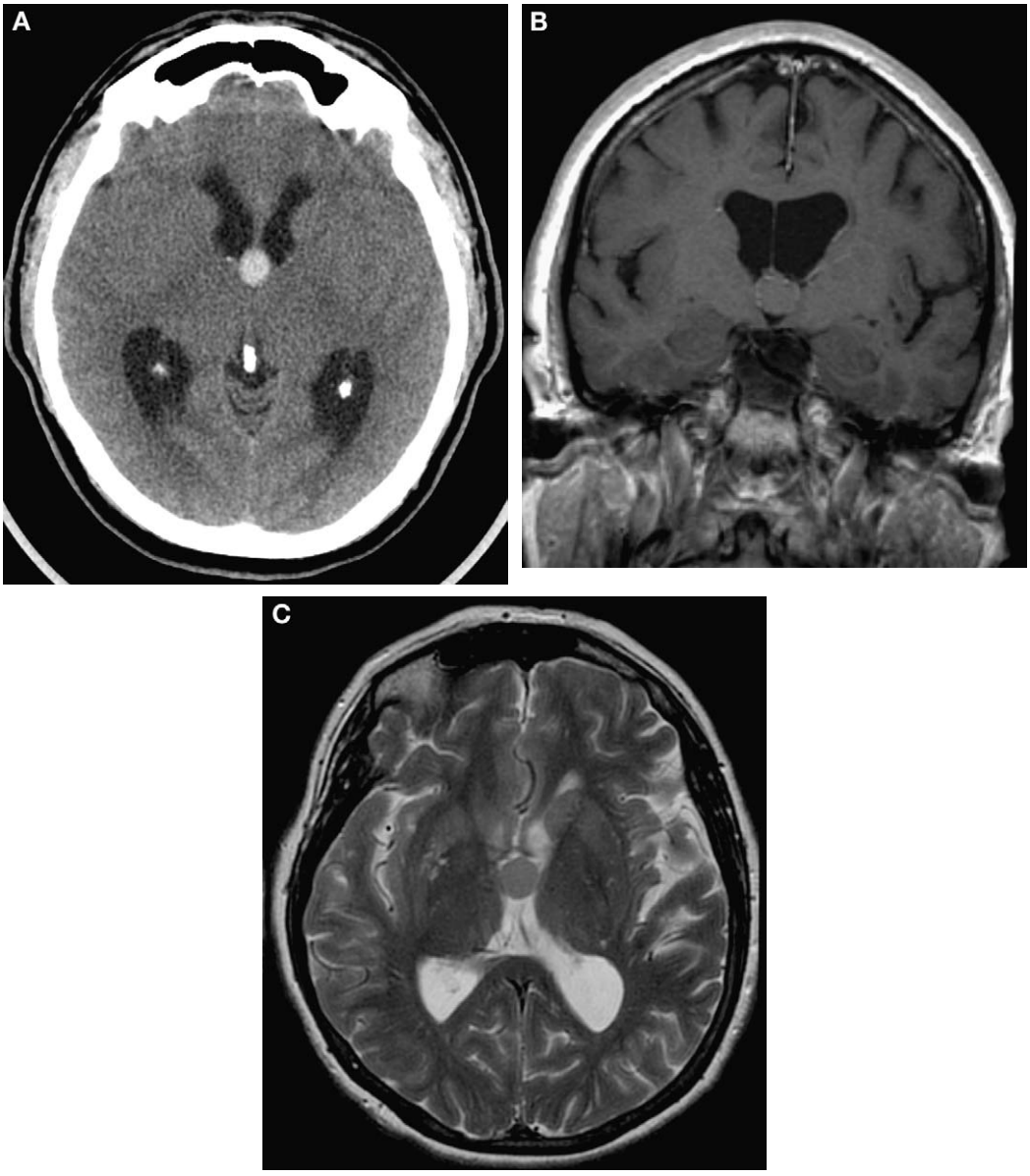

Fig. 2. Colloid cyst. (*A*) Unenhanced CT in an adult demonstrates a homogeneous hyperdense lesion in the region of the foramen of Monro. The ventricles are mildly enlarged. (*B, C*) Coronal T1-weighted postcontrast and axial T2-weighted images in another adult patient show a nonenhancing cyst with medium signal intensity in the roof of the third ventricle causing mild ventricular enlargement. The unusual signal characteristics and location of the lesion are typical of a colloid cyst.

skull. Intraventricular arachnoid cysts are mainly located in the lateral ventricles and only rarely in the third or fourth ventricle [15,16]. Those in the lateral ventricle develop by invagination of arachnoid through the choroidal fissure into the choroid plexus [17] and appear as CSF-filled thin-walled lesions in the atrium of the ventricle (Fig. 3) [18]. They can be classified as either primary or secondary. Primary intraventricular arachnoid cysts arise in the lateral or fourth ventricle, whereas secondary cysts originate extra-axially and extend secondarily into the ventricular

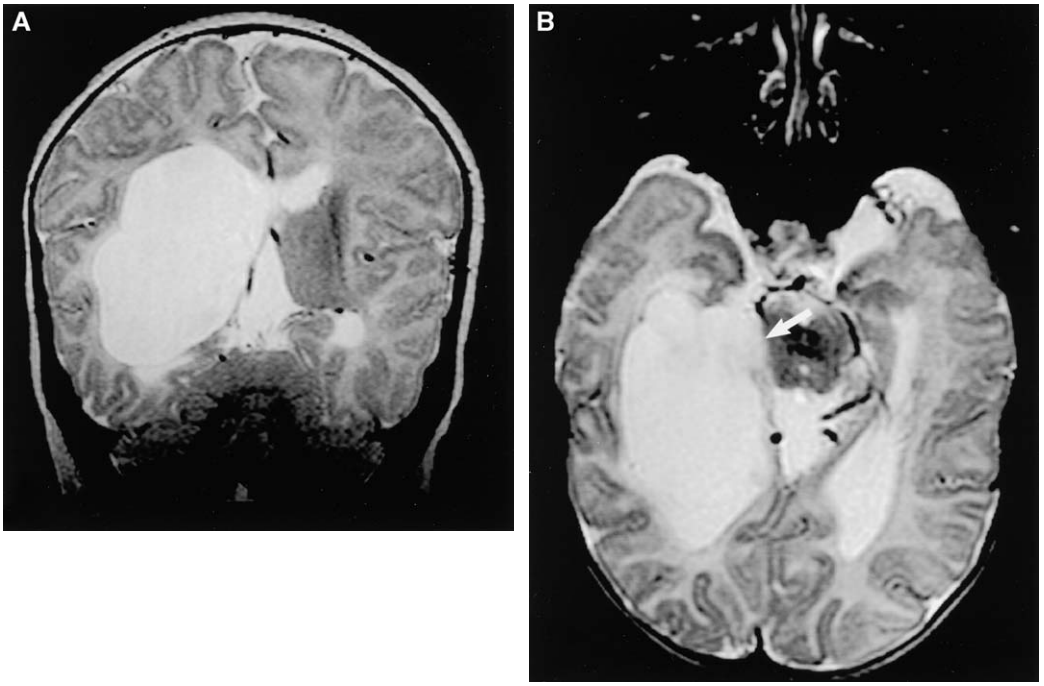

Fig. 3. Intraventricular arachnoid cyst. (*A*) Coronal T2-weighted image demonstrates a large trigonal arachnoid cyst. (*B*) Axial T2-weighted image demonstrates involvement of the perimesencephalic cistern via the choroidal fissure (*arrow*), indicating a secondary rather than primary arachnoid cyst.

system as with suprasellar arachnoid cysts. Primary intraventricular arachnoid cysts are rare, with less than 20 cases reported in the literature. Patients typically present with symptoms of a space-occupying lesion and with headaches, delayed psychomotor development, macrocephaly, hydrocephalus, and seizures [16,19–23].

Symptomatic cysts are usually treated with fenestration by endoscopic or open techniques [19–22,24]. During surgery, they are easy to differentiate from ependymal cysts because of their sole adherence to the choroid plexus, whereas the latter show attachment to the ventricular ependyma [25].

Choroid plexus cysts

Choroid plexus cysts are of neuroepithelial origin and are most often found in the latera ventricles according to autopsy studies (Fig. 4). They are a common finding on prenatal ultrasound during the second trimester, but most spontaneously resolve by birth or early infancy. Ventricular asymmetry may be attributed to a transient choroid plexus cyst that resolves later in life [26]. Only a few reports exist of choroid plexus cysts that

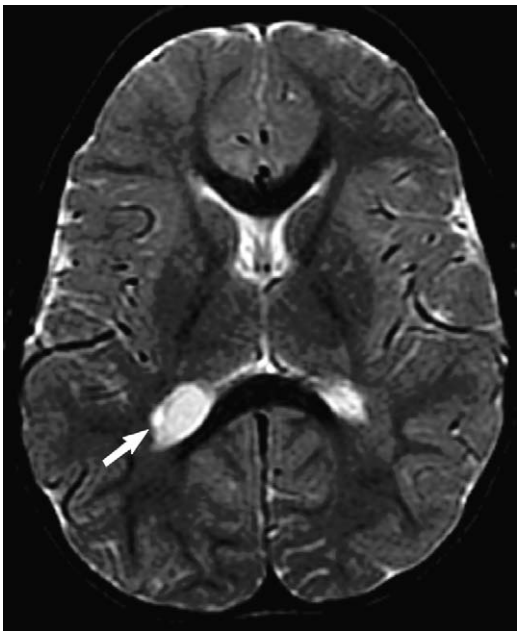

Fig. 4. Choroid plexus cyst. Axial T2-weighted image demonstrates an oval, hyperintense, intraventricular cyst in the region of the glomus of the choroid plexus (*arrow*), characteristic of a choroid plexus cyst.

persist into adulthood. An increased association with aneuploidy or trisomy 18 and 21 has been reported [27]. Shangshotti and Metsky [28] have described four stages of embryologic development of the choroid plexus and attribute the occurrence of these cysts to lobulation of the choroidal neuroepithelium in stage 2. They present as sharply marginated round lesions without septations and can be mobile on a pedicle. They may cause intermittent obstruction of CSF, particularly at the foramen of Monro. Depending on the cyst size, the symptomatic spectrum may include seizures, motor or sensory impairment, papilledema, and an altered level of consciousness [29]. Treatment is indicated in symptomatic cysts that persist past infancy.

Ependymal cysts

Ependymal cysts resemble arachnoid cysts, but the protein content of the cyst fluid is generally higher and they appear as isointense or slightly hyperintense to CSF on T1- and T2-weighted images (Fig. 5) [30]. Their wall is composed of columnar or cuboidal cells that may or may not contain cilia. They are most often found in the atrium of the lateral ventricle. Occasionally, they occur within the central white matter of the frontal or temporoparietal lobes adjacent to the CSF space and may clinically present with signs of increased intracranial pressure or seizures [31–33]. Ependymal cysts are thought to develop as a sequestration of parts of the primitive ependymal lining into the cortical mantle or the perimedullary mesh, and they never communicate with the ventricular system. They are incidental findings on imaging studies of the brain most of the time, but if they become large enough to entrap the ventricle and cause obstructive hydrocephalus, treatment becomes necessary and can be accomplished either by draining of the cyst or resection [34].

Pineal cysts

Pineal cysts are considered a normal anatomic variant and can be found incidentally on MR studies in 2% to 4% of the general population. An even higher incidence of 40% has been reported at autopsy [35]. There is a certain predilection for women in the third decade of life. These cysts

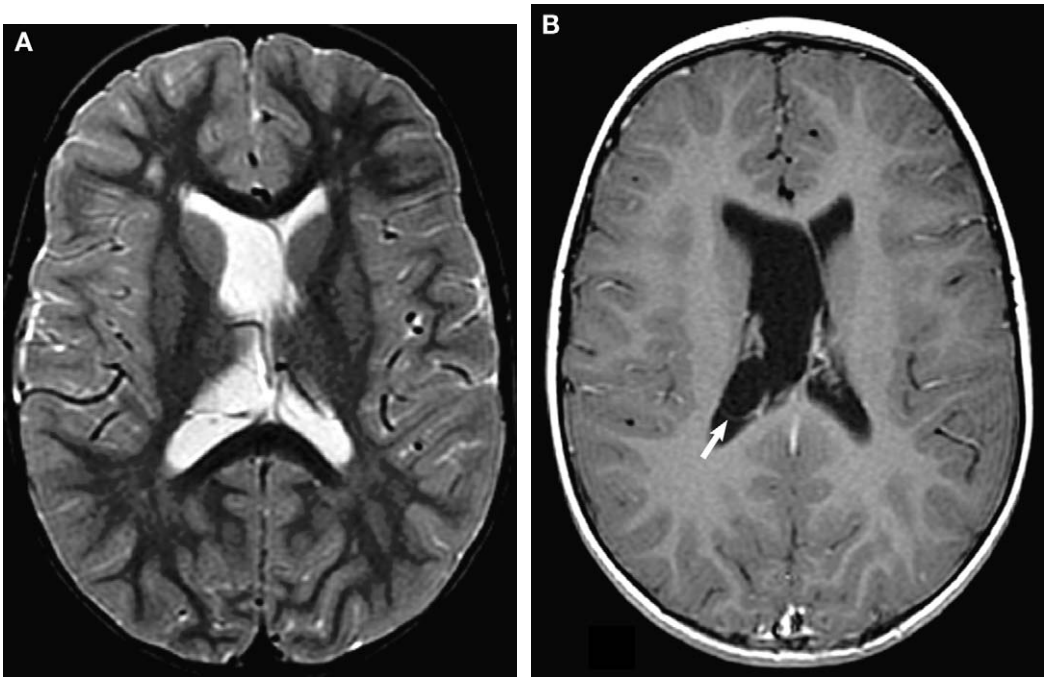

Fig. 5. Ependymal cyst. (*A*) Axial T2-weighted image demonstrates asymmetric lateral ventricles with deviation of the septum pellucidum. (*B*) Axial T1-weighted postcontrast image confirms the presence of an intraventricular cyst with cerebrospinal fluid intensity with visualization of the posterior margin of the cyst wall (*arrow*). Imaging findings are consistent with an ependymal or arachnoid cyst.

contain CSF-like fluid. They rarely become symptomatic. If they do, it is from enlargement with obstruction of the aqueduct of Sylvius. Typical clinical symptoms include headache, gaze palsy, and papilledema [36]. Headaches may be either chronic or intermittent, with intermittent obstruction of the aqueduct responsible for the latter. As described by Miyatake and colleagues [37], compression of the veins in the pineal region, including the precentral cerebellar vein, the internal cerebral veins, and the great vein of Galen, may also contribute to headaches. A sudden onset of headache can be caused by hemorrhage into the cyst [36,38]. Parinaud syndrome with upward gaze palsy is caused by focal compression of the superior colliculus of the tectum mesencephali. On MRI scans, pineal cysts appear hyperintense on T2-weighted images and can have a higher signal than CSF on T1-weighted images because of their higher protein content (Fig. 6). Typically, there is no or mild enhancement of the cyst wall [39,40]. The cyst wall can be calcified, and this is best visualized on CT scans (see Fig. 6).

The current opinion about the ideal treatment of symptomatic pineal cysts is controversial. Even after adequate decompression, symptoms like headache may persist. The aqueduct may not reopen if secondary adhesions have occurred. Operative indications should be restricted to large cysts with compression of nervous structures causing Parinaud syndrome, visual field defects, or disturbances of motor or sensory long-tract fibers. Open surgery via either the infratentorial-supracerebellar or interhemispheric-transcallosal approach has been advocated by some authors [41–46]. Surgery has the advantage of direct visualization of the surrounding veins. Stereotactic cyst aspiration was proposed by others [47–50]. The associated risk of hemorrhagic complications after stereotactic aspiration seems to be lower than formerly estimated. Nevertheless, improper tissue sampling as well as the high probability of cyst re-expansion makes the stereotactic approach less favorable. Michielsen and colleagues [51] have clearly stated that the endoscopic or endoscopic-assisted microneurosurgical approach in experienced hands is the best form of treatment for pineal cysts.

Neurenteric cysts

Abnormal separation of germ cell layers in the third week of gestation is thought to be responsible for persistence of the endodermal tissue elements that can form neurenteric cysts in the central nervous system (CNS). In most reported cases, the cysts are spinal in location [52,53] often with associated vertebral anomalies caused by abnormal development of the adjacent mesoderm. Fewer than 20 cases of intracranial neurenteric cysts have been reported thus far, with most of these located anterior to the brain stem [54,55] or at the craniovertebral junction [56]. Histologically, neurenteric cysts are composed of a thin epithelial wall, which may contain cuboidal or columnar cells with or without ciliation that lie on a basement membrane and connective tissue of varying vascularity [54,57]. If other germ cell elements are present within the cyst, the diagnostic trend is toward a teratoma. Afshar and Scholtz [58] reported on an enterogenous cyst of the fourth ventricle. Because of their expanding nature, surgical cyst excision is the treatment of choice. Aspiration of the cyst content alone is considered insufficient, because the cyst may re-expand by an osmotic gradient or by active mucin secretion of the epithelium. These cysts are known to be attached firmly to surrounding structures, however, making them difficult to remove completely without further neurologic deficits. If cyst removal is not safely possible, opening of the cyst and partial removal may be effective.

Epidermoid cysts

These intracranial cysts are rare and are thought to arise from ectopic ectoderm that was misplaced during the time of neural tube closure. Favorite sites are the cerebellopontine angle and the chiasmal region, but they also occur within the cerebral hemispheres and the ventricles [59]. They may lie within the brain parenchyma adjacent to or in communication with the ventricles [59,60]. There are reports of intrathecal seeding along the CSF pathways as a result of rupture of the cyst into the ventricle and subsequent scattering of oily contents into the CSF space [61,62].

Dandy-Walker malformation

The typical triad of Dandy-Walker syndrome is agenesis or hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ventricle, and supratentorial hydrocephalus (Fig. 7). Most cases are sporadic, but there is evidence of a chromosomal disorder in some reports involving chromosomes 9p and 12p as well as the X-chromosome [63,64] Associated findings are cranial and anterior rotation of the cerebellar vermis as

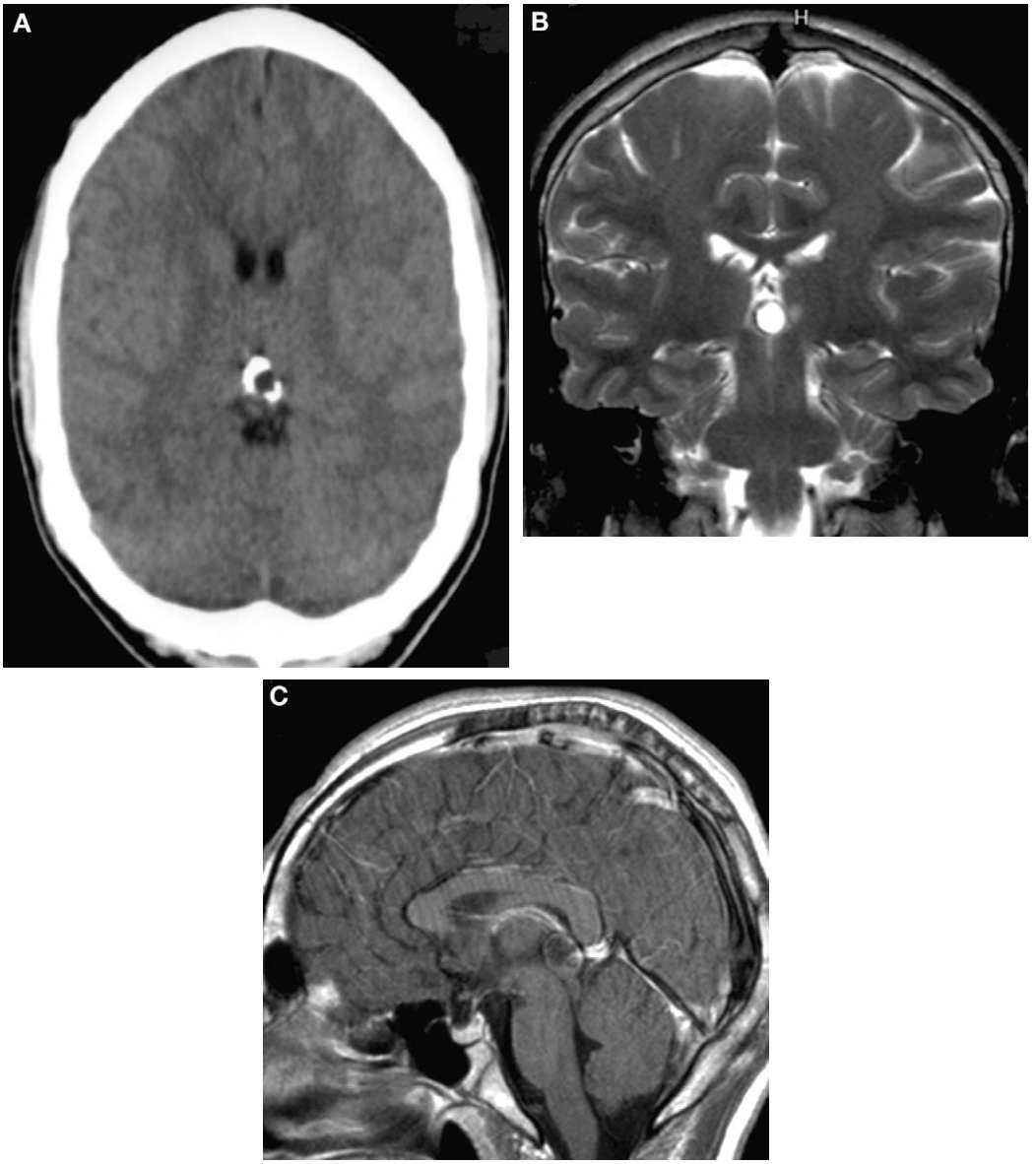

Fig. 6. Pineal cyst. (*A*) Unenhanced CT scan shows a low-density lesion with thick rim calcifications in the region of the pineal gland. Coronal T2-weighted image (*B*) and sagittal T1-weighted postcontrast image (*C*) confirm the presence of a nonenhancing cyst within the pineal gland. Note normal appearance of the third ventricle and lack of calcification visualization on the MRI scan.

well as cranial displacement of the tentorium cerebelli, falx cerebelli, torcular herophili, and transverse sinuses. There is no communication of the cystic CSF collection with the basal subarachnoid space.

Most pediatric patients present early within the first year of life with increased head circumference or symptoms caused by hydrocephalus. The incidence of cerebellar signs or cranial nerve dysfunction is unexpectedly low. Delayed psychomotor development may be present when the syndrome is diagnosed after the first year of life. In up to 68% of patients, other CNS abnormalities can be found, including agenesis of the corpus callosum, occipital encephalocele or meningocele, aqueductal stenosis, CSP, choroid

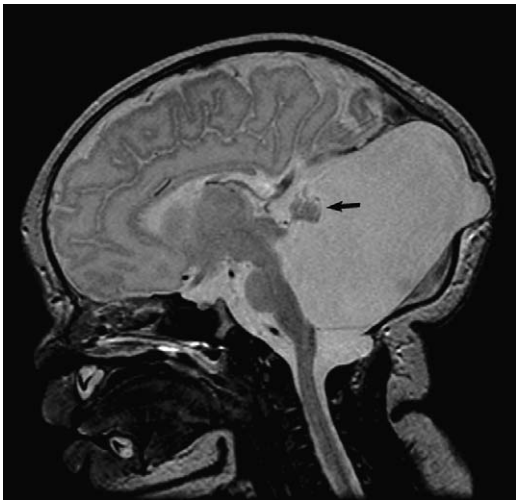

Fig. 7. Dandy-Walker malformation. Sagittal T2-weighted image demonstrates the classic features of a Dandy-Walker malformation. The vermis is severely hypogenetic (*arrow*). The fourth ventricle is "open," communicating with a large retrocerebellar cyst. The posterior fossa is enlarged with torcular-lambdoidal inversion. Note the small occipital meningocele, a common association with Dandy-Walker malformations.

plexus cysts, infundibular hamartomas, macrogyria, microcephaly, syringomyelia, posterior fossa lipoma, and tuberous sclerosis. In addition, systemic abnormalities are found in one fourth of the cases, especially cardiac defects (eg, atrial and ventricular septal defects, patent ductus arteriosus, tetralogy of Fallot), but facial, skeletal, gastrointestinal, and urogenital defects can also be present.

The terminology of posterior fossa cystic lesions has been confused by the application of different terms and classifications. The Dandy-Walker complex includes the Dandy-Walker malformation, the Dandy-Walker variant, and the mega-cisterna magna. The persistent Blake's pouch cyst has been added by Tortori-Donati and colleagues [65].

In the Dandy-Walker variant, the anterior rotation of the hypoplastic cerebellum is not as prominent and the roof as well as the lateral walls of the fourth ventricle can be recognized. The Dandy-Walker syndrome and the Dandy-Walker variant are thought to originate from a failure of assimilation of the area membranacea anterior within the tela choroidea of the fourth ventricle. The foramen of Magendie is usually patent; however, it may open in a delayed fashion. The persistent Blake's pouch is defined by failed regression of the embryonal pouch secondary to nonperforation of the foramen of Magendie. Physiologically, the lateral foramina of Luschka open later than the foramen of Magendie, and the fourth ventricle tends to enlarge during the first stage of development when the latter is not patent. The cerebellar hemispheres and vermis appear hypoplastic but re-expand after shunting, thus indicating mainly compression of the cerebellar structures and not hypoplasia [66]. The aqueduct is not affected.

The mega-cisterna magna is characterized by an enlargement of the cisterna magna with free communication with the basal subarachnoid space and the fourth ventricle. There has been a considerable debate in the past about the optimal treatment of posterior fossa cystic lesions in cases associated with supratentorial hydrocephalus. The question, whether a ventriculo-peritoneal (VP) shunt or a cysto-peritoneal (CP) shunt, or a combination of both, would be superior to the other techniques remains unanswered. There is evidence that after shunting of the posterior fossa cyst, the normal CSF pathway is not always restored, most likely because of secondary aqueductal stenosis. A study performed by Asai [67] and colleagues demonstrated that 9 of 21 patients with initial VP shunts subsequently required CP shunts, whereas only 1 of 10 patients with a CP shunt as the first treatment needed a VP shunt thereafter. Conversely, CP shunts carry a higher risk for secondary brain stem injury, and the occlusion/malfunction rate is higher [68].

Intraventricular congenital lesions

Lipoma of the corpus callosum

Intracranial lipomas are considered to be benign developmental anomalies; they mainly arise in the interhemispheric fissure and only rarely in the interpeduncular, chiasmatic, Sylvian, quadrigeminal, or ambient cistern. Interhemispheric lipomas can be associated with lipomas of the choroid plexus and aplasia of the corpus callosum, anomalies of the cingulate gyrus and the septum pellucidum, and other dysraphic states (Fig. 8) [69–72]. These lesions rarely become symptomatic. Conservative therapy is recommended.

Tuberous sclerosis

The tuberous sclerosis complex is an autosomal dominant inherited disease, but most cases are sporadic. Lesions occurring in this context involve all three primary germ cell layers and are

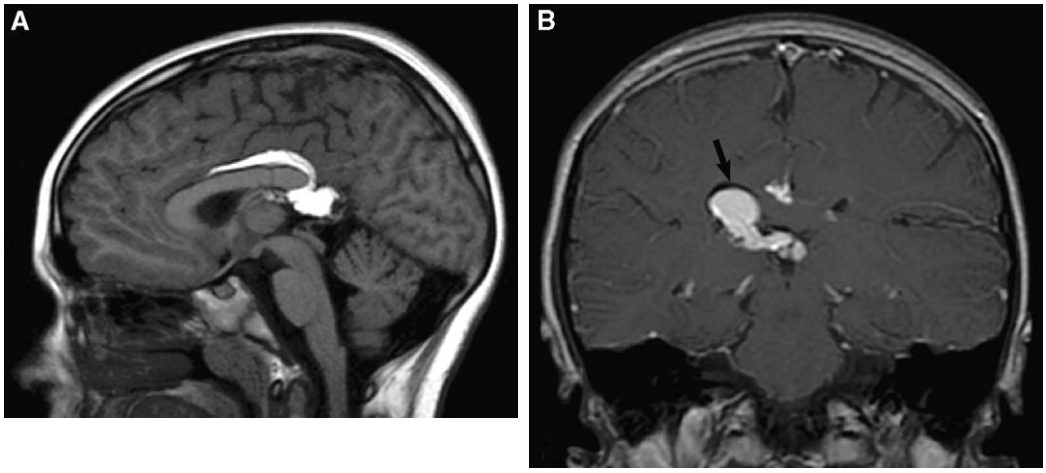

Fig. 8. Interhemispheric lipoma. (*A*) Sagittal T1-weighted image shows a hyperintense lesion surrounding a minimally hypogenetic corpus callosum (absent splenium). (*B*) Coronal T1-weighted postcontrast image demonstrates intraventricular extension (*arrow*), a common finding.

classified as hamartias, hamartomas, or benign tumors. Typical skin lesions are small hypopigmented areas known as ash leaf spots, facial angiofibromas, subungual fibromas, and shagreen patches. Other features include cardiac rhabdomyomas, pulmonary lymphangiomatosis, renal angiomyolipomas, renal cysts, and retinal hamartomas. The most common lesions in the CNS are cortical tubers with seizures, either partial or infantile spasms, as the main presenting symptom. Subependymal nodules along the surface of the lateral ventricles sometimes resembling "candle gutterings" are the lesions that relate to the topic reviewed here (Fig. 9). They are most commonly situated in the caudothalamic groove, have a predilection for the foramen of Monro, and may cause obstructive hydrocephalus. They are often calcified, and their number increases gradually with age until the age of 10 years. The study of Hosoya and colleagues [73] clearly demonstrates that patients with five or more subependymal nodules have a significantly greater number of cortical tubers and white matter lesions as well as a significantly higher percentage of infantile spasms or mental retardation. If these nodules show a tendency to grow, they likely represent subependymal giant cell astrocytomas (SEGAs) (see Fig. 9).

Two distinct genetic loci have been identified in association with the tuberous sclerosis complex. TSC1 is located on chromosome 9q, and TSC2 resides on chromosome 16p. Both loci seem to

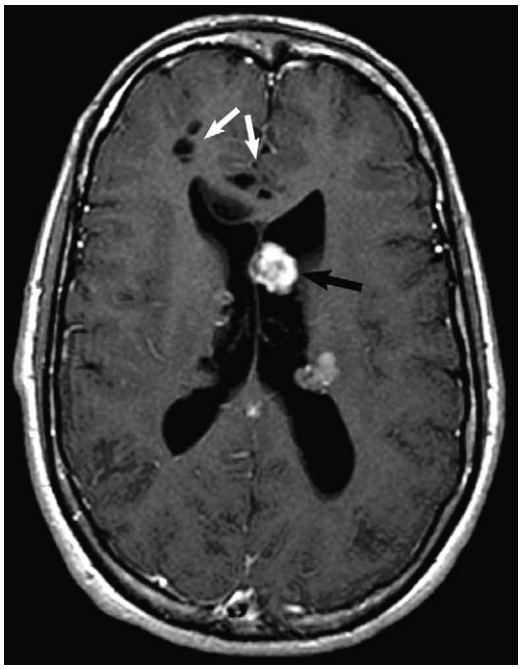

Fig. 9. Tuberous sclerosis. Axial T1-weighted postcontrast image demonstrates multiple enhancing and nonenhancing subependymal tubers. The large enhancing lesion in the vicinity of the foramen of Monro (*black arrow*) is consistent with a giant cell tumor. Mild enlargement of the left lateral ventricle may be related to intermittent obstruction of the foramen of Monro. Note multiple periventricular parenchymal cysts (*white arrows*), not uncommonly identified in tuberous sclerosis.

possess tumor suppressor gene activity. Their exact role in tuber development is not completely understood, but it is speculated that progenitor cells on the germinal layer of the ventricular zone, which are destined for the cortex, undergo inactivation of the TSC1 or TSC2 gene. Those immature neuroepithelial cells that carry the double-hit mutations on either locus are believed to proliferate, migrate, and differentiate abnormally [74]. It is speculated that abnormal cerebral function results from improper synaptic delivery of a neurotransmitter in the mutated cell. There are single case reports of intraventricular hamartomas that required surgical removal or CSF shunting because they caused obstructive hydrocephalus [75,76]. Hamartomas on the floor of the third ventricle are known to cause gelastic epilepsy.

Summary

Intraventricular congenital lesions and colloid cysts comprise a rather large spectrum of different pathologic conditions. In most cases, treatment in not warranted unless there is progressive ventricular obstruction with hydrocephalus or growth of the lesion itself, making tissue biopsy and histopathologic diagnosis necessary. Accordingly, a precise neuroradiologic evaluation is of the utmost importance, because most lesions, if not symptomatic, only require clinical and radiologic follow-up.

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