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Neuroradiology of Selected Disorders of the Meninges, Calvarium, and Venous Sinuses

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Mesenchymal cells develop from ectoderm invaginating into the primitive streak during the third fetal week as the embryo is transformed into a trilaminar disk. The mesenchymal cells form loosely woven tissue called mesenchyme that forms the supporting tissue of the embryo. Eventually, mesenchymal cells form smooth and striate muscle tissue, vessels, connective tissue, and skeleton. Mesenchymal tissue surrounds, protects, and supplies the brain, inasmuch as the meninges, calvarium, skull base, and the vascular supply to brain are all derived from mesenchyme. Congenital disorders of mesenchymal tissue include a wide variety of entities. For the purpose of this presentation, the discussion will be limited to true arachnoid cysts of the meninges, abnormal endochondral bone as in achondroplasia, and abnormal formation of the venous sinuses as expressed in vascular malformations involving ectasia of the vein of Galen.

Meninges

Intracranial True Arachnoid Cysts

True arachnoid cysts are fluid-filled cavities situated entirely within the arachnoid membrane. The cysts are lined by collagen and cells originating from the arachnoid mater. They do not have a glial limiting membrane or an epithelial lining in the wall (1). True arachnoid cysts can either communicate freely with or be more or less separate from the subarachnoid space. They are found most commonly in specific locations such as the temporal fossa (Figs. 1 and 2) and the posterior fossa. True arachnoid cysts are thought to be of developmental origin but are first encountered in all age groups. In many patients, the detection of an arachnoid cyst represents an obvious incidental finding. In others, however, the cyst appears to cause elevated intracranial pressure and clinical symptoms such as headaches or seizures.

Secondary arachnoid cysts may form as a response to inflammatory reactions in the subarachnoid space. They represent encystment of the subarachnoid space by arachnoiditis.

Pathogenesis

The arachnoid space forms as an expansion of the extracellular space in the mesenchyme surrounding the neural tube (2). An outer (arachnoid) and an inner (pial) layer of cells is formed, delimiting a subarachnoid space between, which will contain cerebrospinal fluid (CSF). This space is traversed by numerous cells and strands of connective tissue, forming the arachnoid trabeculae. Arachnoid cysts represent congenital malformations of the arachnoid and are thought to represent minor aberrations in the way the space is formed between the arachnoid and pia (3). The common association between middle cranial fossa arachnoid cysts and venous anomalies would date the origin of the malformation to between the sixth and eighth fetal week (4). Cysts in the suprasellar cistern could possibly be caused by an upward diverticulation of the membrane of Liliequist (5). Although the theory that arachnoid cysts could form as a passive distension of the arachnoid space secondary to primary hypoplasia of the brain has been withdrawn by its author, the common association between middle cranial

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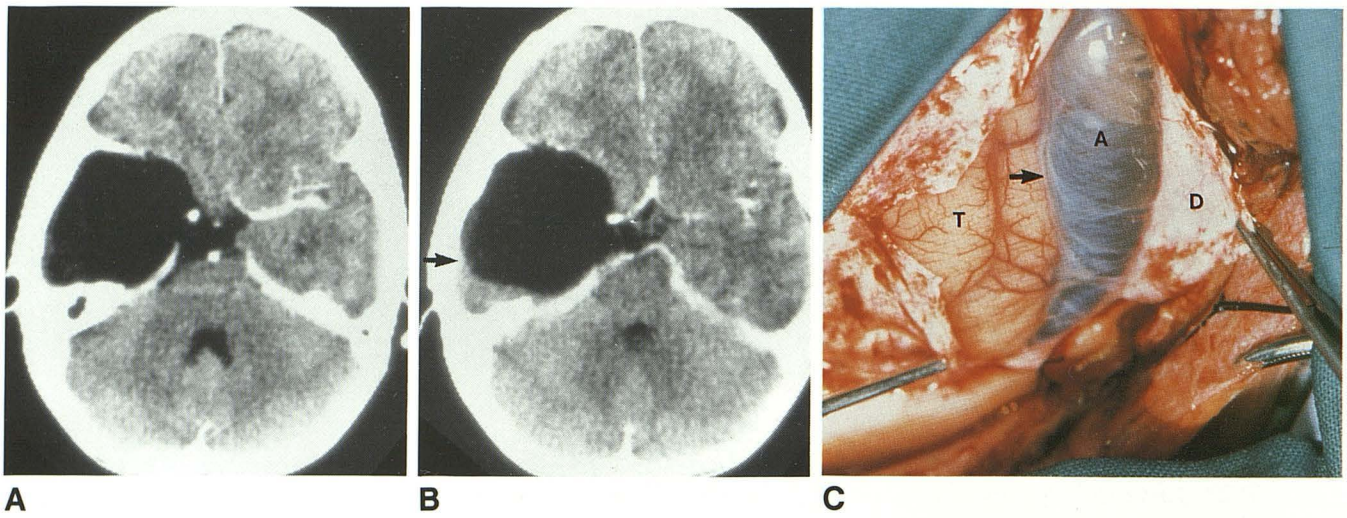


Fig. 1. Temporal fossa arachnoid cyst; 10-year-old boy.

A and *B*, Contrast-enhanced axial CT scans reveal a well-defined homogeneously lucent CSF density mass that displaces the margin (*arrow*) of the temporal lobe posteriorly. The calvarium is thicker adjacent to the cyst than on the contralateral side.

C, Surgical exposure. Opening the dura (*D*) and reflecting it forward exposes the margin (*arrow*) of the temporal lobe (*T*) displaced by the bluish arachnoid cyst (*A*). The bulging cyst was lightly adherent to the deep surface of the dura. The pial vessels are displaced with the brain surface. No pial vasculature crosses the surface of the cyst.

(Reprinted from Naidich et al (1). Case courtesy of Thomas P. Naidich, MD, Miami.)

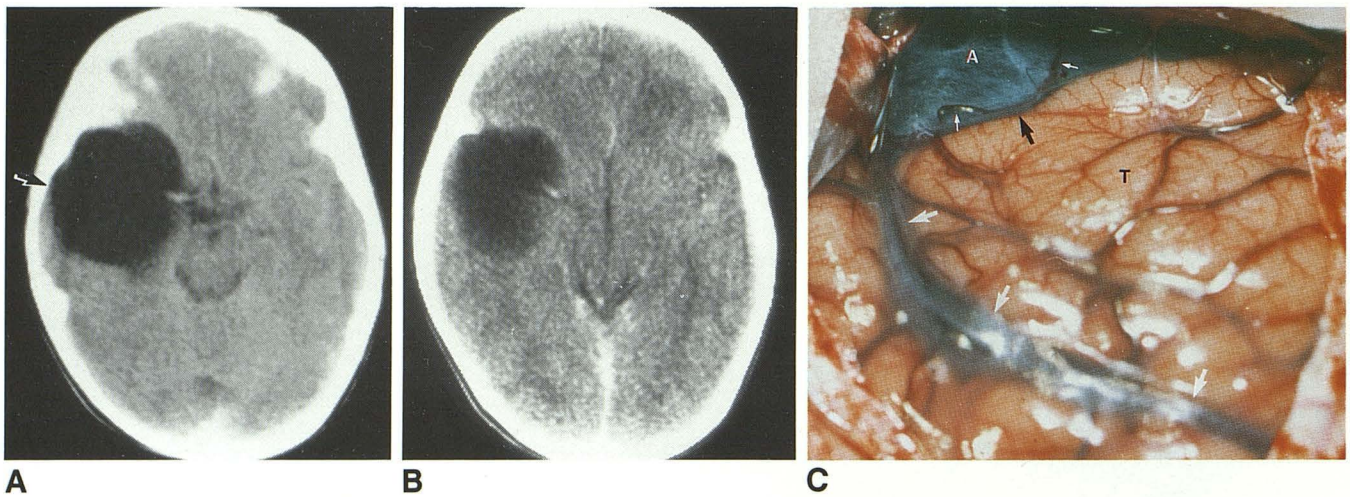


Fig. 2. Temporal fossa arachnoid cyst; 5½-year-old boy.

A and *B*, Contrast-enhanced axial CT scans reveal the well-defined, homogeneously lucent, CSF density cyst at the anterior temporal fossa. The margin (*arrow*) of the temporal lobe is displaced posteriorly by the cyst.

C, Surgical exposure; the dura has been opened widely. The bluish arachnoid cyst (*A*) displaces the margin (*black arrow*) of the temporal lobe (*T*) and the associated pial vessels posteriorly. Only veins (*small white arrows*) cross the external surface of the cyst. The superficial middle cerebral vein (*large white arrows*) passes along the sylvian fissure toward the sphenoparietal sinus.

(Case courtesy of Thomas P. Naidich, MD, Miami.)

fossa arachnoid cysts and hypogenesis of the temporal lobe has been demonstrated using magnetic resonance (MR) imaging (6).

The natural history of arachnoid cysts is poorly understood. Few studies have demonstrated changes in cyst volume on serial studies. It is well known that some arachnoid cysts communicate freely with the subarachnoid space, whereas, in others, contrast or isotope introduced into the subarachnoid space enters the cyst more or less slowly or never at all. Serial imaging has demonstrated that some cysts increase in size with time, while others disappear spontaneously, often in association with hemorrhage. Several theories have been entertained in explaining cyst growth. Perhaps the cyst membrane has a secretory ca-

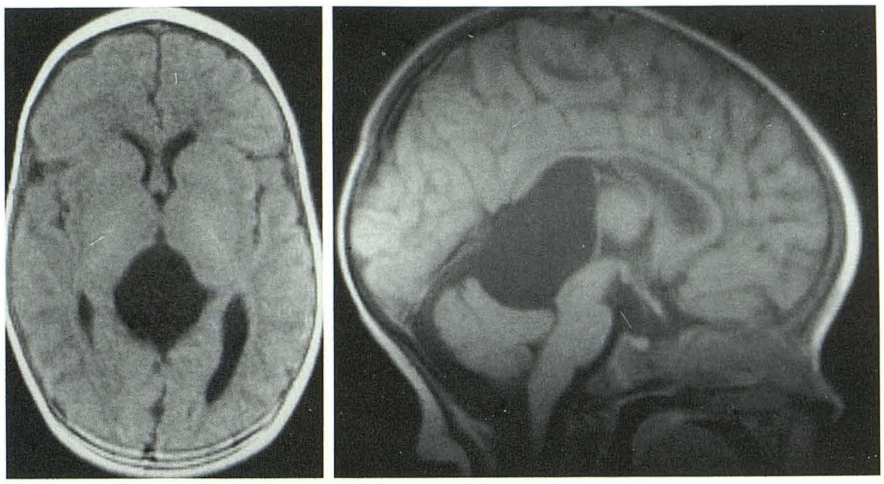
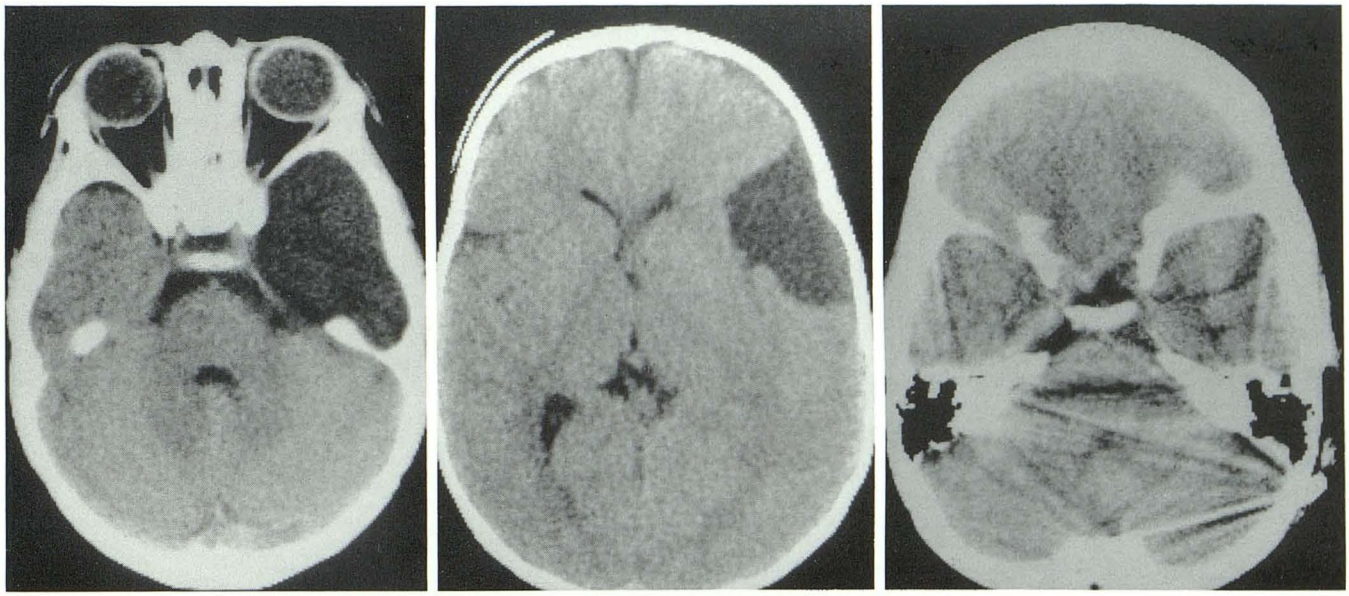


Fig. 3. Quadrigeminal plate arachnoid cyst; 2-month-old girl. *A*, T1 axial MR; *B*, sagittal T1 MR. The features of this arachnoid cyst are most clearly shown in the sagittal plane MR. Although the cyst is expansile, hydrocephalus has not developed. The cyst was initially detected by routine neurosonography.



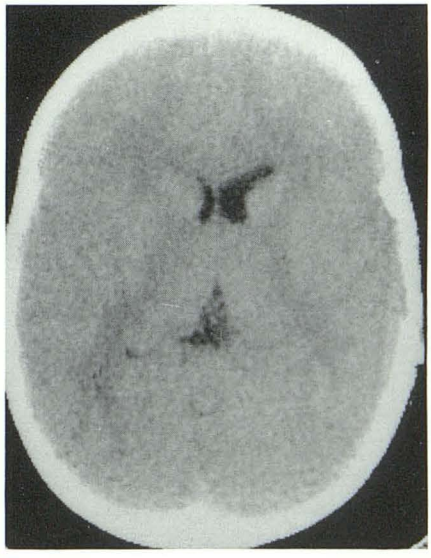


Fig. 4. Temporal fossa arachnoid cyst.

A and *B*, 8-month-old boy; noncontrast axial CT. The left middle cranial fossa arachnoid cyst was found incidentally on CT performed following head injury. The left middle cranial fossa is expanded and the greater wing of sphenoid is pushed forward. The sylvian fissure is widely opened. The lateral ventricle is compressed and there is minor shift of midline structures. The child underwent surgery with placement of a cystoperitoneal shunt.

C and *D*, Repeat noncontrast axial CT, 4 years later. The left middle cranial fossa remains larger. It is filled with brain tissue. There is minimal, probably compensatory, dilatation of the left lateral ventricle but no trace of arachnoid cyst. The tip of the cystoperitoneal shunt is just barely seen.

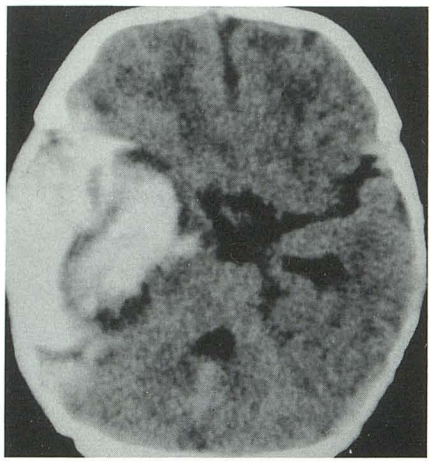


Fig. 5. Hemorrhage into an arachnoid cyst; newborn. At surgery the fresh hemorrhage was found to be located mainly within a right middle cranial fossa arachnoid cyst. The presence of this cyst was not suspected prior to surgery.

pacity, but this is controversial. Hydrodynamic effects from arterial and CSF pulsations combined with a ball-valve mechanism has been more widely accepted as the cause of expansile arachnoid cysts. Statistical analysis of radiologic findings in a large number of arachnoid cysts has failed to show an overall correlation between cyst size and patient age. The results further suggested that a large majority of small arachnoid cysts remain constant in size, whereas only a small subgroup of expansile lesions exists. These lesions are more commonly large, suggesting that large arachnoid cysts less commonly communicate with the subarachnoid space and should require particular clinical attention (7).

Incidence

Arachnoid cysts constitute about 1% of intracranial mass lesions. They occur sporadically and affect males more often than females. Most cysts are single lesions found during the first two decades of life. The middle cranial fossa is the most common location (50%) and, in this location, left-sided lesions are about twice as common (7). About one third of the arachnoid cysts are found in the posterior fossa, while about 10% are found in the suprasellar region (1).

Clinical Signs

Signs and symptoms clearly resulting from an arachnoid cyst are usually related to large cysts with raised intracranial pressure. Small cysts are found incidentally. Although patients found to have arachnoid cysts frequently complain of chronic headaches, seizures, psychomotor retardation, and behavioral problems (4), the precise relationship to the arachnoid cyst detected is uncertain since these symptoms also represent the most common indications for neuroradiologic evaluation. Hence the casual relationship between the finding of an arachnoid cyst and these symptoms is rather tenuous, despite reports of virtual total therapeutic success in patients complaining of headaches and seizures (4).

Signs of raised intracranial pressure are usually associated with larger cysts or cysts in the posterior fossa with hydrocephalus. Although drainage of the cyst usually alleviates the hydrocephalus, the mechanism behind the hydrocephalus may be disturbed CSF resorption rather than mechanical obstruction of the CSF pathways. Hemorrhage into an arachnoid cyst can cause rapid onset of focal neurologic symptoms. Subdural hemorrhage has been reported to originate in a preexisting arachnoid cyst, a diagnosis that may be difficult unless the radiologist is aware of this association (8).

Cysts of the suprasellar cisterns have symptoms specific to their location, especially endocrine abnormalities. Large cysts in this location can cause hydrocephalus and damage to the optic chiasm.

Neuroradiology

Arachnoid cysts can be diagnosed correctly with neurosonography, computed tomography (CT), or MR imaging. Although useful only in infants, neurosonography is superior in defining the cyst as a unilocular structure filled with fluid. Both CT and MR are capable of establishing that the content of the cyst has attenuation or signal characteristics indistinguishable from CSF (Fig. 3). On MR, the signal of CSF in the cyst can be similar or identical to that of an epidermoid tumor.

The most common location for an arachnoid cyst is in the *middle cranial fossa*. These cysts are clearly demarcated, usually unilocular cysts with CT attenuation and MR signal identical to CSF in all imaging sequences. Smaller cysts occupy the anterior portion of the middle cranial fossa without obvious mass effect. These lesions may be missed on routine CT imaging, because partial volume averaging of the cyst with bone tends to obscure the lesion in the usual imaging plane. Somewhat larger cysts tend to expand into

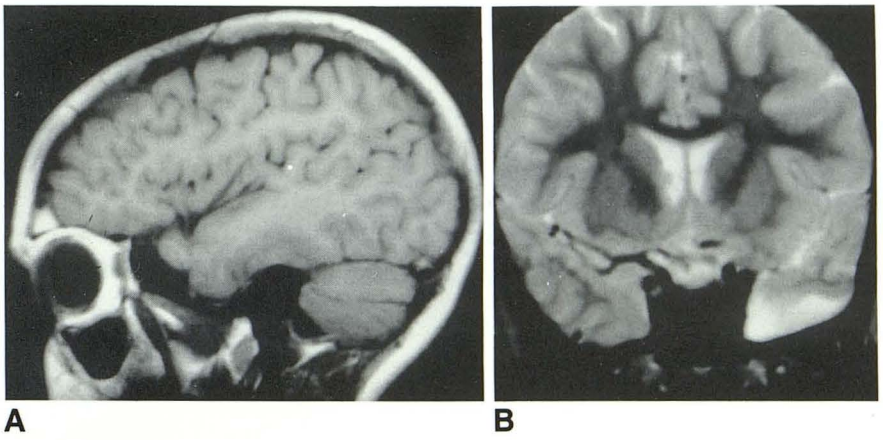


Fig. 6. Temporal fossa arachnoid cyst. *A*, Sagittal T1 MR; *B*, coronal T2 MR. The arachnoid cyst occupies the most anterior part of the left middle cranial fossa. The temporal lobe appears hypogenetic with irregular margins and without obvious mass effect associated with the cyst. The signal in the cyst is the same as the ventricular CSF. The loss of tissue in the left temporal lobe is obvious.

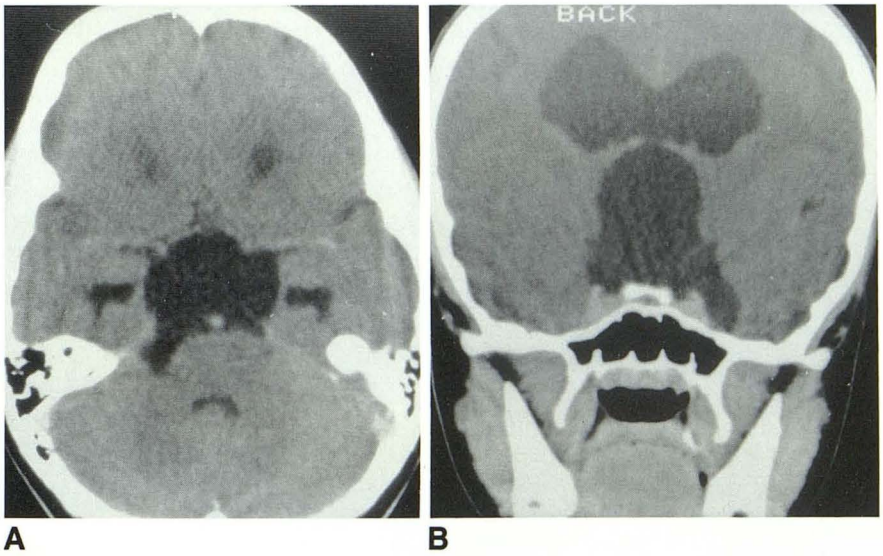


Fig. 7. Suprasellar arachnoid cyst; 9-year-old boy with precocious puberty.

A, Axial contrast-enhanced CT; the nonenhancing suprasellar cyst expands in all directions.

B, Coronal contrast-enhanced CT. The cyst has invaginated into the third ventricle. Peroperative neurosonography (data not shown) demonstrated that attempts to shunt the cyst failed, because the cyst wall gave way to the needle. The shunt was eventually placed in the third ventricule outside the cyst.

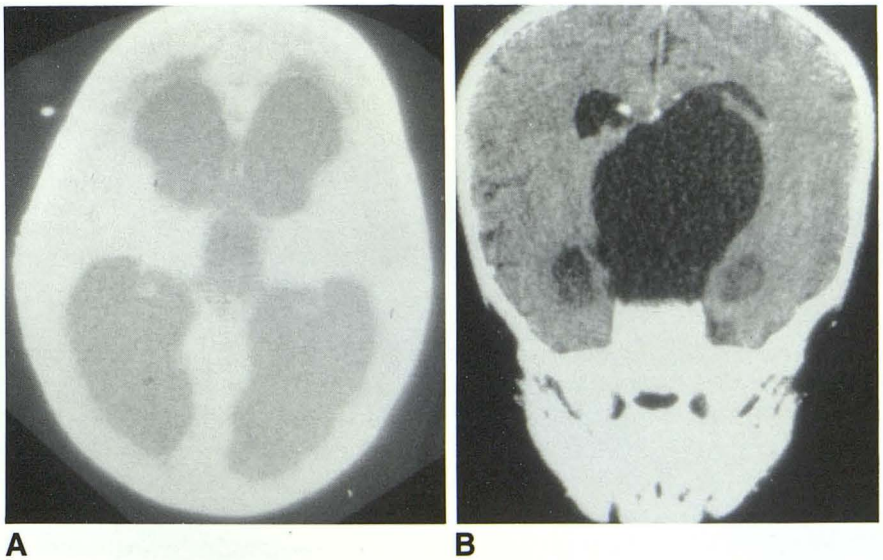


Fig. 8. Suprasellar arachnoid cyst; 5-month-old girl.

A, The initial CT scan was interpreted as representing aqueductal stenosis, so the infant underwent ventriculoperitoneal shunting.

B, The patient developed blindness. Repeat CT 6 months later showed massive expansion of a suprasellar arachnoid cyst.

the sylvian fissure and open up this space. Large lesions commonly expand the bony middle cranial fossa anteriorly and inferiorly and may cause thinning and bulging of the temporal squama (Fig. 4). Uncommonly, they are associated with paradoxical thickening of the adjacent calvarium (Fig.

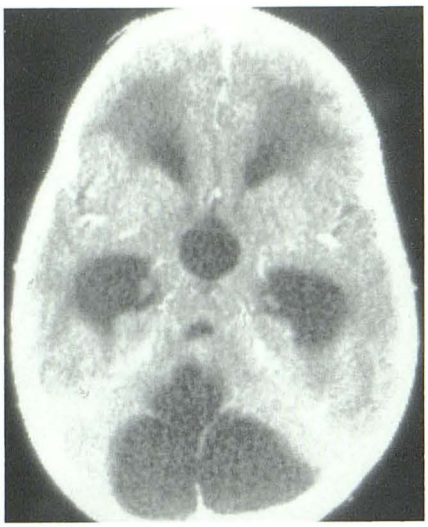


Fig. 9. Posterior fossa arachnoid cyst; 2½-year-old boy with macrocephaly and clinical evidence of raised intracranial pressure. CT shows hydrocephalus due to a multilocular arachnoid cyst in the inferior midline of the posterior fossa. The fourth ventricle is displaced and deformed.

1). Very large cysts in this location have striking mass effect and compress the brain. Arachnoid cysts in the middle cranial fossa have been reported to be associated with hemorrhage much more frequently than cysts in any other location. This is thought to be due to frequently associated vascular, mainly venous, anomalies in this location (1). The radiologist must be aware of this association as the original arachnoid cyst may be very difficult to detect in the presence of fresh hemorrhage (Fig. 5). Careful evaluation of the bony structures may suggest a long-standing mass lesion in the middle cranial fossa (8).

Recent experience with MR imaging of middle cranial fossa arachnoid cysts has revived the discussion about associated hypogenesis of the temporal lobe. Although the arachnoid cyst and the hypogenesis of the temporal lobe occur together, a causative relationship may not be present. The two lesions may be caused by the same insult rather than one being the cause of the other. Early treatment of an arachnoid cyst may result in complete disappearance of the cyst despite apparent hypogenesis of the temporal lobe (Figs. 4C and 4D). It is possible that treatment early in infancy may have a greater chance to reduce the volume of the cyst since the brain maintains some growth potential (4). Imaging in the sagittal plane through the temporal lobe depicts clearly the anatomy of the temporal lobe (Fig. 6). Eight of eight patients studied with MR showed evidence of associated hypogenesis of the temporal lobe, in three associated with compression (6). The authors suggest that if MR shows absence of compression, surgery would not be indicated.

Cysts of the *suprasellar cisterns* have a more definite relationship with clinical signs and symptoms as they expand in all directions (Fig. 7). These cysts may be difficult to diagnose correctly, because small cysts may cause variations in the shape of the suprasellar cisterns without much change in volume, whereas very large cysts can distort the anatomy so that it is difficult to recognize a large CSF space as a cyst. Although the pressure in the cyst may be just slightly higher than in the ventricular system, the arachnoid cyst may expand superiorly and invaginate into the third ventricle. It can completely replace this structure (Fig. 7B). The appearance may then closely simulate the usual picture of aqueductal stenosis with dilated third and lateral ventricles. If unrecognized, this situation is dangerous, since surgical decompression of the lateral ventricles may change the balance of pressure in the intracranial cavity and allow the cyst to expand freely (Fig. 8). An epidermoid in the suprasellar cistern may have signal characteristics indistinguishable from an arachnoid cyst on MR. However, epidermoid tumors are usually lobulated with slightly inhomogeneous signal and less distinct margins.

Cysts in the *posterior fossa* are usually large and more frequently associated with hydrocephalus. The most common locations include the cerebellopontine angle and the inferior midline (Fig. 9). Arachnoid cysts in the latter location must be separated from the Dandy-Walker complex. This is usually possible using CT. In difficult cases, MR imaging, particularly in the sagittal plane, will show the cyst to be separate from the fourth ventricle and vallecula (9).

Calvarium-Skull Base

Abnormal Endochondral Bone

Achondroplasia is a bone dysplasia characterized by failure of normal endochondral bone formation. The base of the skull from the mendosal suture posteriorly to the frontosphenoid synchondrosis anteriorly is formed from cartilage. Abnormal formation of endochondral bone will result in a small skull base, with reduction in the area of all foramina through the base. Macrocranium is a well-known phenomenon in patients with achondroplasia. Although many reasons for the large head have been proposed in the past, ven-

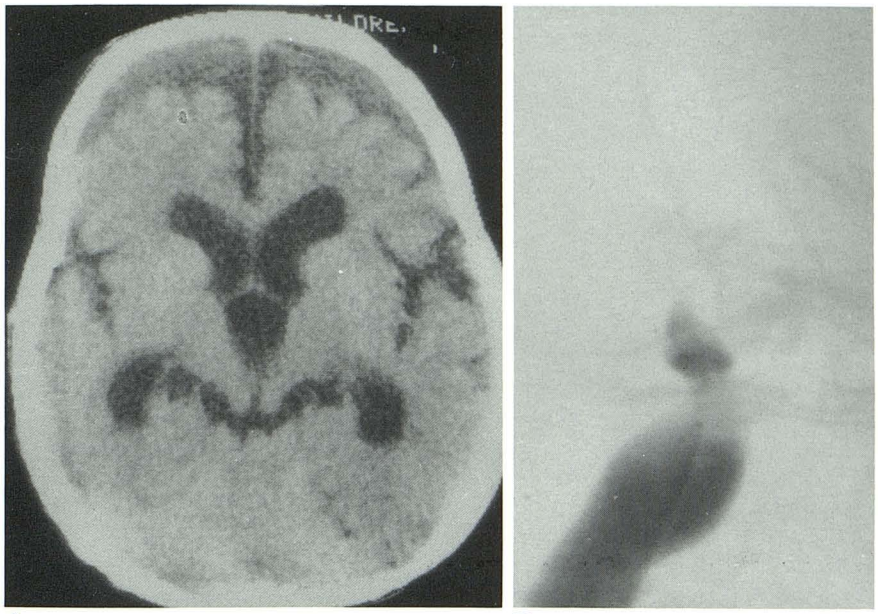


Fig. 10. Achondroplasia; 2-year-old boy with macrocrania.

A, Axial CT shows ventriculomegaly and prominent subarachnoid space. This constellation of clinical and neuroradiologic findings is consistent with communicating hydrocephalus. Invasive monitoring over 24 hours confirmed raised intracranial pressure.

B, Venography of the jugular veins bilaterally showed stenosis at the level of the jugular foramen and a pressure gradient across the foramen.

triculomegaly has been found in all these patients (Fig. 10A). The cause of the ventriculomegaly has been controversial. It has not been clear whether ventriculomegaly is caused by abnormal CSF dynamics and hydrocephalus and whether treatment is indicated.

The large head and the ventriculomegaly tend to stabilize in the older child. Clinically, overt raised intracranial pressure is rare in these patients. Invasive monitoring of intracranial pressure over 24 hours in children with achondroplasia has shown the intracranial pressure to be elevated (10). These data would indicate that ventriculomegaly does indeed represent hydrocephalus, albeit nonprogressive later in childhood. The cause of the abnormal CSF dynamics was further proved to be unrelated to any obstruction to CSF flow, since contrast and radioisotope instilled in the ventricles rapidly diffused throughout the entire ventricular system and subarachnoid space. This suggested impaired CSF resorption as the most likely cause of hydrocephalus.

CSF is absorbed in the arachnoid villi that provide a direct communication between CSF and the venous sinus. A pressure gradient between the subarachnoid space and the venous sinus maintains this communication. Thus the absorption of CSF is passive, with CSF flowing down the pressure gradient from the subarachnoid space to the venous sinus. The CSF absorption capacity is three to four times the normal production of CSF. Although venous hypertension may decrease absorption of CSF, the causal relationship between this and hydrocephalus is disputed. However, hydrocephalus has been documented in young children with intracranial venous sinus occlusions, whereas the same situation in older children failed to produce hydrocephalus. The presence of open sutures appears to be a prerequisite for the development of hydrocephalus secondary to intracranial venous hypertension. Thus, only congenital venous obstructions may cause hydrocephalus (11).

The jugular foramen is congenitally small in patients with achondroplasia. Invasive studies have shown a pressure gradient across the jugular foramen in children with achondroplasia and hydrocephalus (Fig. 10B). This stenosis will cause intracranial venous hypertension that is unaffected by changes in the intracranial pressure. Thus venous hypertension in patients with achondroplasia is likely the *cause* of impaired CSF absorption and subsequent hydrocephalus and not a consequence of raised intracranial pressure (10). Indeed construction of a venous bypass from the transverse sinus to the jugular vein has resulted in decreased intracranial pressure and reduced ventriculomegaly in a patient with achondroplasia (12).

Veins and Venous Sinuses

Vein of Galen Ectasia

Close anatomical analysis of children and adults found to have ectasia of the vein of Galen associated with an arteriovenous malformation

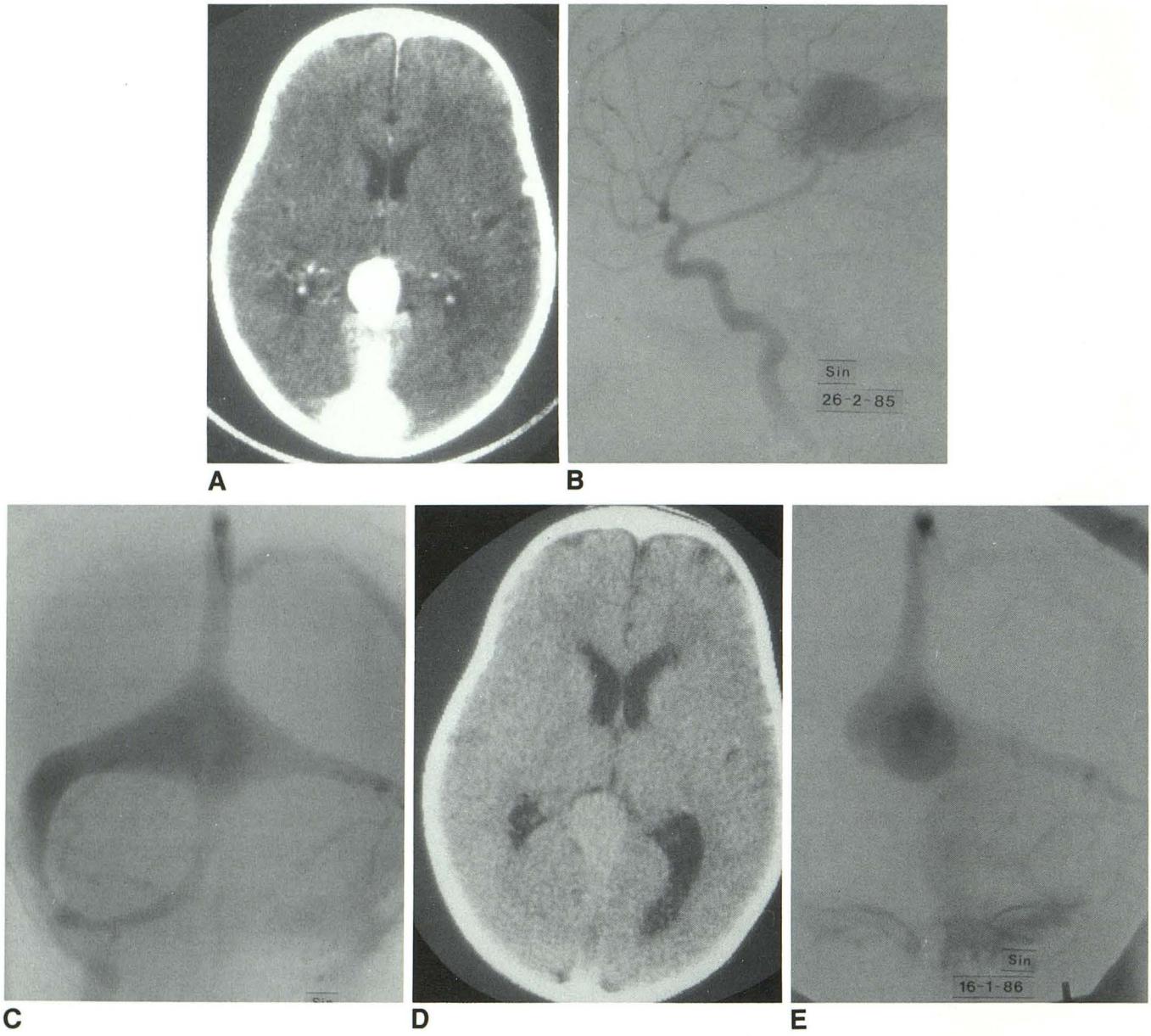


Fig. 11. Vein of Galen ectasia; 18-month-old girl with failure to thrive.

A, Contrast-enhanced axial CT shows a vein of Galen malformation with normal ventricular size.

B, Cerebral angiograms 3 weeks later confirmed the presence of an AVM supplied by vessels from both posterior cerebral arteries and transmesencephalic branches from the basilar artery. The ectatic vein of Galen drained to the straight sinus.

C, Late venous phase showed agenesis of the left sigmoid sinus and marked stenosis at the right jugular foramen.

D, Repeat CT 10 months later, when the child complained of headaches, showed increased ventricular size, successfully treated with a ventriculoperitoneal shunt.

E, Repeat angiogram 2 months later in preparation for endovascular embolization of the AVM. Late venous phase showed spontaneous total thrombosis of the right transverse sinus since the prior angiogram. This was the likely explanation for the onset of hydrocephalus 2 months earlier.

The child was successfully treated with partial endovascular embolization with bucrylate. Stereotactic radiosurgery was then applied to the residual AVM. No remaining AVM could be detected 2 years following radiosurgery.

has raised interesting questions concerning the etiology of this abnormality and its associated clinical consequences.

The presence of an intracerebral arteriovenous shunt with preferential drainage into the vein of Galen is the prerequisite for the formation of ectasia of the draining vein of Galen (13) (Figs. 11A and 11B). However, many deep-seated arteriovenous malformations (AVM) may have a high flow draining into the vein of Galen without

ectasia of this vein. Hence, the ectasia of the vein is a consequence not only of high flow but of a hemodynamic phenomenon, where an obstruction distal to the vein of Galen can create the conditions necessary for the formation of ectasia.

Obstructions of the venous drainage can be due to acquired thrombosis or congenital agenesis or hypogenesis of dural sinuses. Although there are no reports in the literature of thrombosis of the straight sinus producing ectasia of the vein of Galen, a dural venous obstruction of congenital origin distal to the vein of Galen is always present in cases with ectasia of the vein. Agenesis of the straight sinus is most common, but a more distal obstruction can be found at the level of the jugular foramen if the straight sinus is present (Fig. 11C). The closer the obstruction is to the vein of Galen, the more prominent is the ectasia.

The arteriovenous shunt draining into the ectatic vein of Galen will increase the venous pressure. Increased intracranial venous pressure can impair the resorption of CSF and cause communicating hydrocephalus in a child with open sutures. However, the obstruction distal to the ectasia in the venous system will reduce the flow through the shunt and, depending on location, may protect the venous system draining the rest of the brain. The intracranial venous pressure will remain low and not affect the resorption of CSF in the granulations of Pacheoni if the dural sinus obstruction is located close to the ectatic vein of Galen. Ventricular enlargement in this situation is due to direct compression of the aqueduct of Sylvius, ie, noncommunicating hydrocephalus. Conversely, the pressure will be high in all intracranial venous sinuses if the venous obstruction is located more distally, close to the jugular foramen. In this situation, the CSF resorption will be impaired and the hydrocephalus will be of the communicating type (14) (Figs. 11D and 11E).

It has been shown that arteriovenous shunting and venous sinus obstruction are present in every patient with ectasia of the vein of Galen. Hence, it is possible that venous obstruction is the cause of both the ectasia and the shunt. Although the embryology of AVM is unknown, the venous system matures before the arterial system, so it is conceivable that a venous obstruction could maintain the capillary network in a plexiform stage and thus induce the persistence of an arteriovenous shunt. This theory is speculative and has yet to be proved.

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