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AJNR Am J Neuroradiol 1986, 7 (4) 725-728

<http://www.ajnr.org/content/7/4/725.citation>

This information is current as
of April 20, 2024.

Arachnoid Cyst of the Quadrigeminal Plate Cistern: Report of Two Cases

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Arachnoid cysts of the quadrigeminal plate cistern are rare, probably congenital, cystic infratentorial lesions that are located between the collicular plates and the incisural notch of the tentorium. Twenty-nine cases have been reported in the literature since 1940 [1–18], the great majority before the advent of CT.

The current report describes two cases in which the location and precise extent of an arachnoid cyst of the quadrigeminal plate cistern were clearly shown by CT in conjunction with reformatted imaging in the sagittal plane. Additional data concerning the CSF communications were provided by radionuclide or metrizamide CT ventriculography or concomitant metrizamide CT and radionuclide cisternography. Both cases also showed significant supratentorial extension of the cyst. The finding of ectopic choroid plexus in the wall of the cyst in one patient was decidedly rare.

Case Reports

Case 1

A 10-year-old boy was seen at the emergency room of Milwaukee Children's Hospital because of vomiting, irritability, and increasing obtundation. He had had a ventriculoperitoneal shunt placed at age 22 months for progressive hydrocephalus related to a posterior fossa cyst that was thought to be a Dandy-Walker cyst. He had been doing well except for mild developmental delay until the time of the present admission.

Cranial CT showed dilated lateral and third ventricles. There was a large midline posterior fossa cyst (Fig. 1A). A revision of the cranial end of the diversionary shunt resolved the vomiting and obtundation. The following morning he was alert and oriented. Cranial CT now showed a significant decrease in the size of the ventricles, but the size of the posterior fossa cyst was unchanged. A reformatted image in the sagittal plane showed that the cyst bowed the tentorium upward and extended through the incisura for 2.5 cm (Fig. 1B).

The CSF communications of this cystic structure, if any, were uncertain. A radionuclide evaluation of the diversionary shunt system revealed no communication between the shunted ventricular system and the cyst. Subsequently, simultaneous radionuclide and metrizamide CT cisternography showed that the cyst did not communicate

with the fourth ventricle or the subarachnoid spaces of the posterior fossa. The cyst abutted against the quadrigeminal plate cistern (Fig. 1C).

The cyst was explored through a suboccipital craniotomy and was found to contain fluid that was grossly identical to CSF. A small linearly oriented tuft of tissue was found on the margin of the rostral inferior aspect of the cyst. This tissue was histologically normal choroid plexus (Fig. 1D), whereas the cyst was lined with arachnoid.

Fenestrations were created between the cyst and the left lateral ventricle as well as the cisterna magna. An attempt to create a window into the collicular plate cistern was abandoned because of multiple layers of thickened arachnoid. Postoperatively, the child has done well and is currently asymptomatic. A follow-up CT showed that the cyst markedly decreased in size.

Case 2

A 10-month-old boy had been in good health until age 6 months, when his head began to enlarge. Between age 6 and 9 months his head continued to grow at a disproportionate rate, and his acquisition of developmental skills was slow. Cranial CT (Fig. 2A) showed moderate dilatation of both lateral ventricles and the third ventricle. In addition, a midline cyst was noted that appeared to arise from either the posterior aspect of the third ventricle or from the region of the quadrigeminal plate. A ventriculoperitoneal shunt was placed. Three days later, a metrizamide ventriculogram showed a decrease in the size of the lateral ventricles but no change in the size of the midline cyst (Fig. 2B).

The cyst did not communicate with the ventricles. A reformatted image in the sagittal plane showed that the tentorium was elevated and that the cyst extended through the tentorial notch. The third ventricle was displaced anteriorly; the cerebellum was displaced inferiorly (Fig. 2C). A CT obtained 2 months later revealed an increase in the size of the cyst. The lateral and third ventricles remained decompressed. A cyst-peritoneal shunt was performed. The fluid in the cyst was grossly identical to CSF. Since this shunt was placed, the child has been asymptomatic.

Discussion

The majority of intracranial arachnoid cysts lie above the tentorium; infratentorial arachnoid cysts are rarer. Five loca-

Received September 7, 1984; accepted after revision December 22, 1984.

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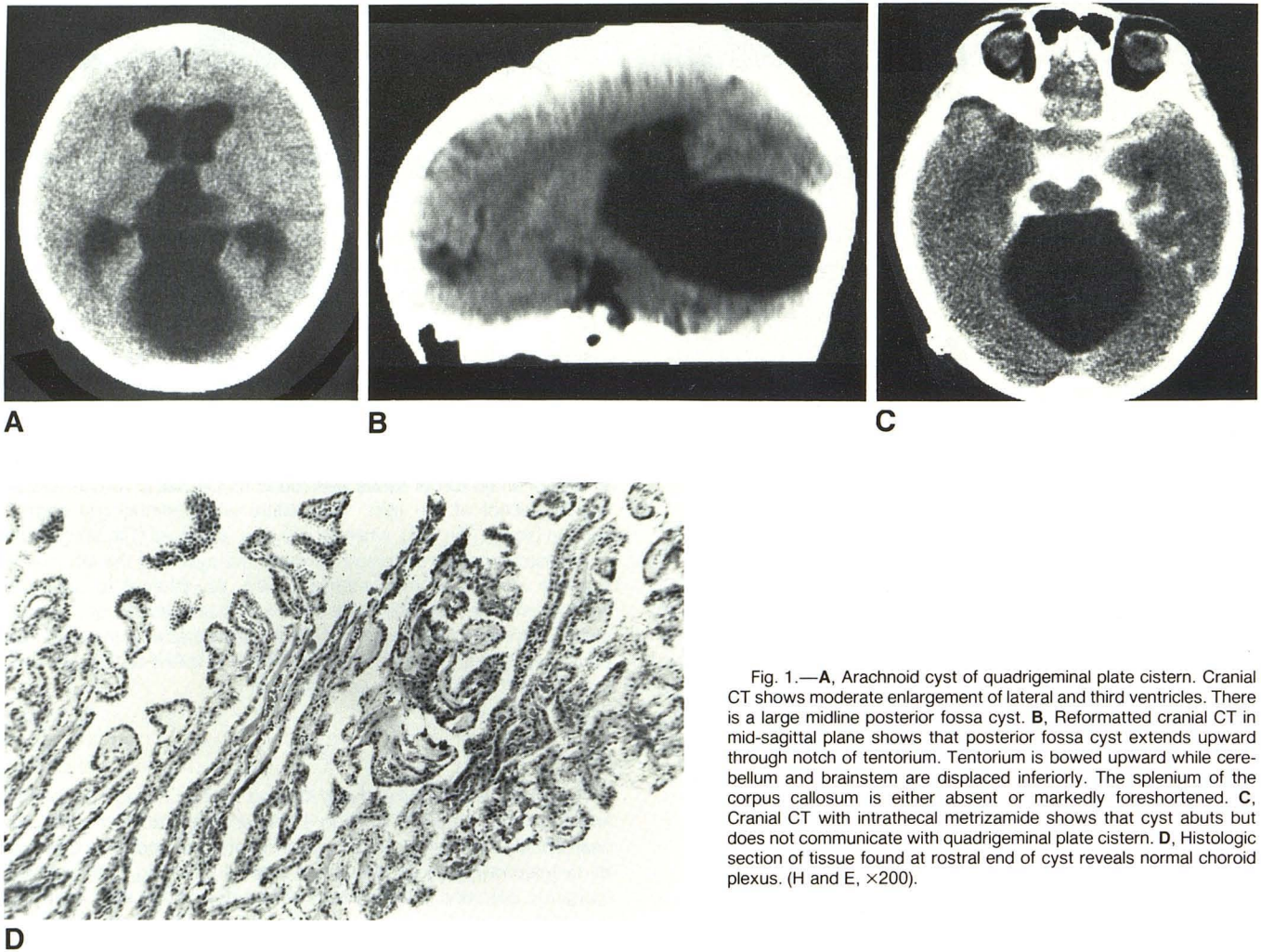


Fig. 1.—**A**, Arachnoid cyst of quadrigeminal plate cistern. Cranial CT shows moderate enlargement of lateral and third ventricles. There is a large midline posterior fossa cyst. **B**, Reformatted cranial CT in mid-sagittal plane shows that posterior fossa cyst extends upward through notch of tentorium. Tentorium is bowed upward while cerebellum and brainstem are displaced inferiorly. The splenium of the corpus callosum is either absent or markedly foreshortened. **C**, Cranial CT with intrathecal metrizamide shows that cyst abuts but does not communicate with quadrigeminal plate cistern. **D**, Histologic section of tissue found at rostral end of cyst reveals normal choroid plexus. (H and E, $\times 200$).

tions of infratentorial arachnoid cysts are recognized: (1) midline retrocerebellar, (2) lateral (or hemispheric) retrocerebellar, (3) cerebellopontine angle, (4) clival, and (5) quadrigeminal. Of these, the retrocerebellar and cerebellopontine-angle cysts are decidedly more common [11].

A distinction can also be made on the basis of age at clinical presentation, since quadrigeminal cysts are found in infants or young children while the two types of retrocerebellar cysts are seen in adolescents or young adults and cerebellopontine-angle cysts are found almost exclusively in adults [8, 11, 16].

Considerable dispute surrounds the etiology of arachnoid cysts, since both acquired and developmental theories exist as to their formation. Meningeal inflammation and trauma are two major etiologic mechanisms proposed for acquired lesions, while defective duplication of the leptomeninges and growth of ectopic ependymal and arachnoid tissue are postulated as the explanation for developmental lesions. It is generally agreed that most intracranial arachnoid cysts, especially those found beneath the tentorium cerebelli, are developmental in origin. It has also been proposed that some arachnoid cysts develop from embryonic rests that develop into rudimentary secretory organs or into mature choroid

plexus [11, 19, 20]. An ectopic choroid plexus in a posterior fossa cyst is rare. The majority have been retrocerebellar lesions; only one previous case was in a quadrigeminal plate cyst [6]. The presence of ectopic choroid plexus in the wall of the cyst of case 1 may account for the large size of the cyst.

It may not always be clear to the surgeon whether such a cyst lies between sheets of arachnoid (intraarachnoid), between the pia and the arachnoid (subarachnoid), or between the arachnoid and the dura (subdural) [11]. However, it has been shown by electron microscopy that cysts of the quadrigeminal plate cistern are located within the arachnoid and are formed by a splitting or duplication of the arachnoid membrane [19, 21].

Quadrigeminal arachnoid cysts may extend above the tentorial notch, a finding seen in about 25% of cases. If the supratentorial component is exceptionally large, the tentorium cerebelli may be depressed rather than elevated [5].

Since 1940, 29 cases of arachnoid cyst of the quadrigeminal plate cistern have been reported [1-18]. The most common clinical presentation is progressive enlargement of the head of infants between the ages of 2-10 months, seen

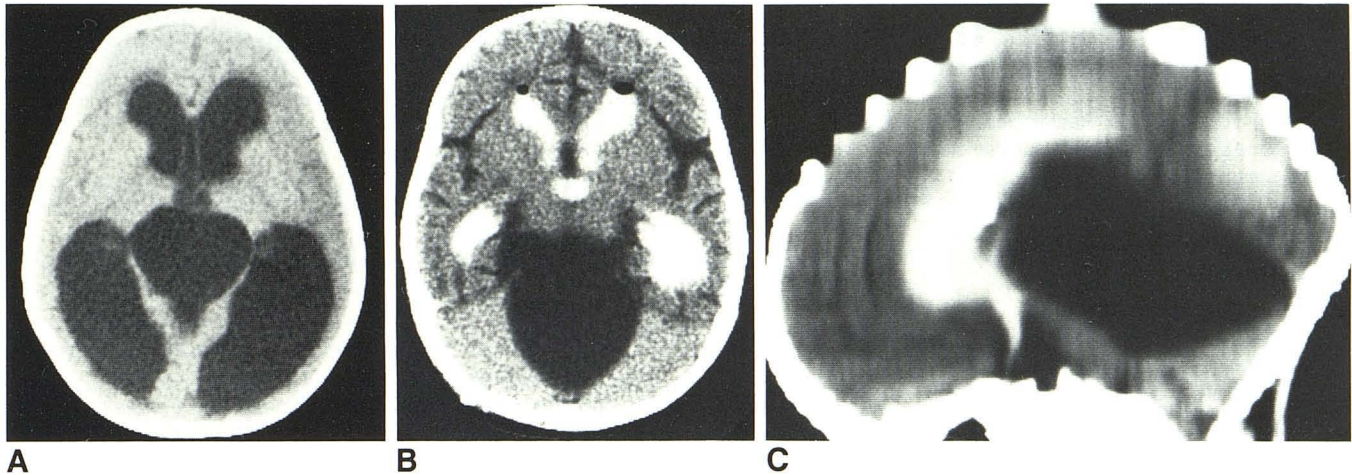


Fig. 2.—**A**, Arachnoid cyst of quadrigeminal plate cistern. Cranial CT shows moderate dilatation of lateral and third ventricles. Third ventricle is displaced anteriorly by large midline cyst that, on lower sections, extended into posterior fossa. There is a cavum septum pellucidum that is also dilated. **B**, Cranial CT after placing metrizamide into right lateral ventricle through diversionary shunt

shows reduction in size of ventricles and enlargement of subarachnoid spaces. Posterior fossa cyst does not communicate with ventricles. **C**, Reformatted CT image in the mid-sagittal plane shows that cyst extends upward through notch of tentorium. Tentorium is elevated while cerebellum is depressed. The splenium of the corpus callosum is either absent or markedly foreshortened.

in about twice as many males as females. Typically, there are no localizing neurologic signs, but signs of increased intracranial pressure are common. Occasionally, a delay in achieving developmental milestones is seen. Surprisingly, focal neurologic deficits from compression of the pineal gland, colliculi, brainstem, or cerebellum are rare. When present, these include Parinaud's syndrome [4, 9], visual disturbances [4, 7, 18], nystagmus [4, 7, 13], or hearing deficits [4, 22]. Motor signs such as hemiparesis [17], lower extremity weakness [16], or generalized spasticity and clonus [5] are very rare. Occasionally, lateral rectus palsy [5, 18] has been reported, probably a "false" localizing sign of increased intracranial pressure.

Infants with cysts of the quadrigeminal plate cistern are normal at birth. Signs of hydrocephalus develop between 4–12 months of age. Six cases have been reported in patients in a distinctly older age group [1, 4, 11]. It is uncertain whether these lesions represent a different type of quadrigeminal plate cyst.

Before CT became available, the diagnosis of posterior fossa cysts was difficult. Pneumoencephalography or ventriculography, either with air or positive contrast material, were the usual diagnostic procedures. Currently, CT without or with metrizamide cisternography can readily differentiate the various types of posterior fossa cysts. Radionuclide ventriculography (if the patient has a diversionary shunt) or cisternography can provide supplementary data about the CSF dynamics.

The CT hallmarks that establish the diagnosis of a quadrigeminal arachnoid cyst are a midline, supracerebellar, infratentorial cyst that abuts the quadrigeminal plate cistern. The contents of the cyst have attenuation characteristics that are identical to CSF. The cyst, while infratentorial, may extend for a variable distance upward through the tentorial notch. This is best seen on images reformatted in the sagittal plane.

Similar data could be obtained with MRI. The cerebellum is displaced inferiorly. The aqueduct is stretched and narrowed, causing obstructive dilatation of the third and lateral ventricles. Metrizamide and/or radionuclide cisternography show that the cyst does not communicate with either the ventricular system or the subarachnoid space.

Patients with arachnoid cysts of the quadrigeminal plate cistern require surgical intervention for shunting of the obstructed ventricular system as well as drainage of the cyst, either externally or internally. A detailed knowledge of the anatomy of the cyst and the CSF spaces is crucial for effective placement of the efferent catheter or catheters of the diversionary shunt system. It is generally agreed that a cyst of the quadrigeminal plate cistern should be drained by establishing a permanent fistula between the cyst and a ventricle and/or the subarachnoid space [23]. Also, if internal cyst drainage (windowing) is elected, strategic placement of the fenestrations, using meticulous surgical technique, is mandatory. When permanent internal drainage is feasible, it is always the preferred technique. If successful, the possibility of neurologic damage from shunt obstruction is obviated. The risk of this is greater with posterior fossa cysts because of the added risks of upward or downward herniation through the incisura of the tentorium and the possibility of localized deformity of the brainstem by the undecompressed cyst.

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