

## RADIOLOGICAL AND CLINICAL FEATURES OF AN ARACHNOID CYST OF THE QUADRIGEMINAL CISTERN

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Cummins, Taveras, and Schlesinger (1960) have emphasized again the high mortality rate associated with surgical exploration of lesions in the region of the posterior third ventricle. However, there are certain benign processes which occur in this area which require definitive surgical treatment rather than shunting procedures and/or radiation therapy, and the successful management of obstructing mass lesions in this critical area is dependent upon correct pre-operative diagnosis.

This report records an example of an arachnoid cyst of the quadrigeminal cistern. Nine similar cases have been recorded previously. Hamby and Gardner (1935) described a 16-year-old girl who complained of deafness and suffered from increased intracranial pressure resulting from an ependymal cyst in the supracollicular area. Their patient recovered after surgical evacuation of the cyst through the anterior medullary velum. Although the cyst wall was composed of ependymal cells there was neither radiological nor surgical evidence of a communication between the cyst and the ventricular system. Alexander (1953) reported an 18-month-old patient with an arachnoid cyst of the quadrigeminal cistern. Severe obstructive hydrocephalus was relieved by the operative establishment of a communication between the posterior end of the third ventricle and the cyst. This child, when evaluated three years post-operatively, showed residual spasticity and moderate retardation on psychological testing. Dott and Gillingham (1958) described an infant suffering from spasticity and an enlarging head. A large arachnoid cyst of the supracollicular region was drained through the right ventricle, but the patient died of a post-operative haematoma. Katagiri (1960) recorded two cases of arachnoid cyst of the cisterna ambiens, but the illustrations indicated that these lesions were in fact in the quadrigeminal cistern. Both patients were adults. One died after surgery; the other was

'cured' by ventricular taps. Noetzel (1940) reported three patients with cysts in the cisterna ambiens in whom the symptoms resembled the clinical picture of pineal tumour. Hoshino, quoted by Katagiri (1960), recorded one case in which a cyst in this area produced ventriculographic findings suggestive of a tumour of the quadrigeminal plate. Dyke (1959), in 'Diagnostic Roentgenology', illustrated a lateral film from a ventriculogram which was labelled a 'cyst of dura'. His case, in our opinion, would seem to represent an arachnoid or ependymal cyst of the quadrigeminal cistern.

### Case Report

A 13-month-old boy was admitted to the Syracuse Memorial Hospital on March 29, 1960, because of regression in neuromuscular function and a rapidly enlarging head. The baby had developed normally to the age of 10 months at which time he could stand with help and spoke a few simple words. He then became irritable, slowly lost the ability to stand or sit, and at 12 months developed an internal squint of the right eye.

The mother's pregnancy and delivery were uneventful. There was no history of infectious disease in childhood.

Physical examination revealed a very irritable child. The skull was enlarged (52.5 cm. maximum circumference), but the scalp veins were flat and no cranial bruit was heard. Neurological examination demonstrated a right abducens palsy, bilateral papilloedema, a positive MacEwen's sign, and severe generalized spasticity and clonus. The blood count and urine analysis were indeterminate. Plain skull films showed widened sutures, an enlarged vault, and a simple linear fracture in the right parietal bone.

On the second hospital day subdural taps were performed through the enlarged anterior fontanelle, but no fluid was obtained.

Two days later contrast radiographs were obtained. (A detailed description of the pertinent films made during this and subsequent studies is given beneath Figs. 1 and 2.)

A gas ventriculogram was performed first because of

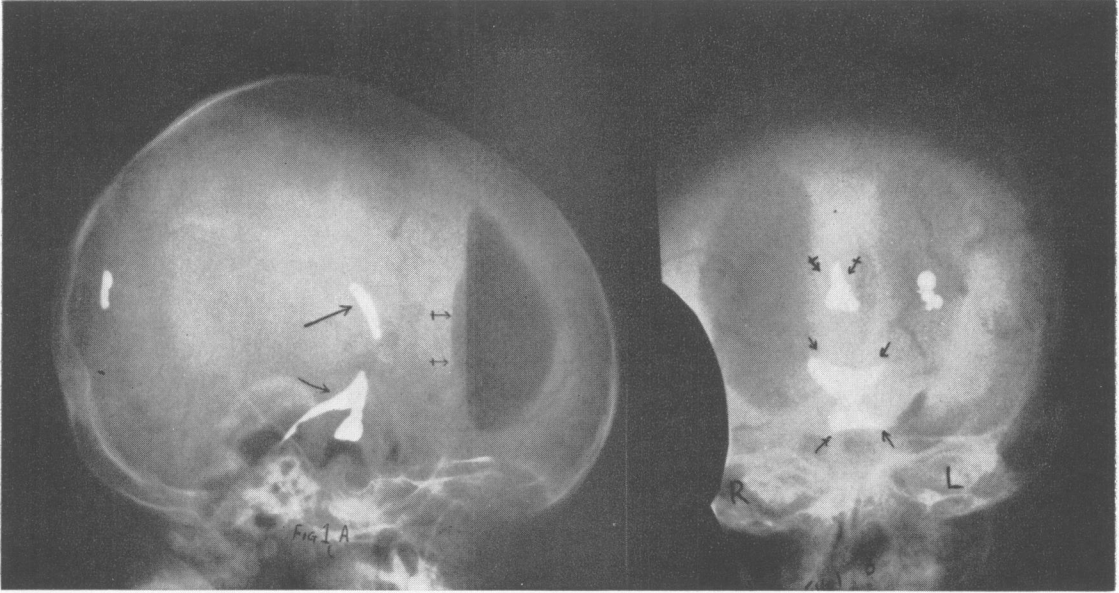


FIG. 1A

FIG. 1B

FIG. 1.—Lateral (A) and frontal (B) films of the skull made with the patient in the brow-up position following air ventriculography, attempted pneumoencephalography, and Pantopaque ventriculography.

The gas outlines the undisplaced anterior portion of a markedly dilated third ventricle (†) and symmetrically dilated frontal horns. The Pantopaque outlines the posterior inferior portions of the dilated third ventricle and the aqueduct of Sylvius (A). The suprapineal recess is bowed and displaced upward and forward, and the junction of the third ventricle and aqueduct and the proximal aqueduct are bowed forward anteriorly and inferiorly in a smooth curve. (A †) The distance from the aqueduct to the dorsum sellae is markedly reduced. The aqueduct is not visualized in (B) because it is obscured by the opaque oil in the posterior inferior third ventricle.

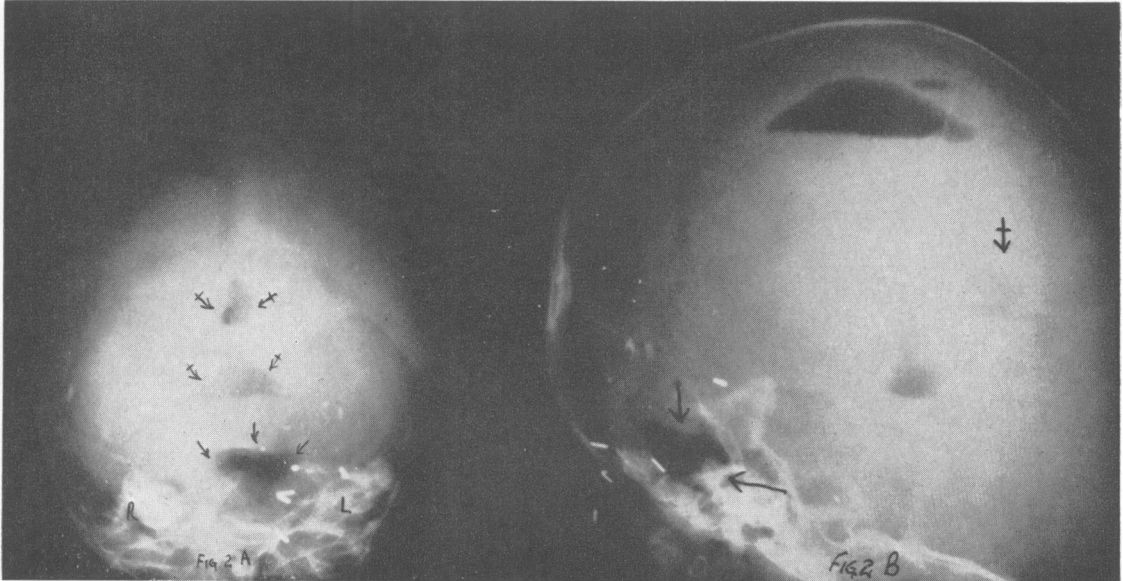


FIG. 2A

FIG. 2B

FIG. 2.—Frontal (A) and lateral (B) skull films made with the patient erect and with the neck semiflexed during the post-operative pneumoencephalogram after the first craniotomy.

At this time the aqueduct was not obstructed and gas passed into the third ventricle and right lateral ventricle from below. The fourth ventricle is seen (†) widened from side to side and diminished in its cephalo-caudad dimension. There is no lateral displacement of the fourth ventricle but it is displaced caudad.

The roof of the dilated third ventricle and the displaced and dilated suprapineal recess are outlined with gas. (†) The two air-fluid levels visible are probably due to the indentation of the posterior portion of the roof of the third ventricle by kinking forward of the corpus callosum.

the presence of brain-stem signs and increased intracranial pressure. The posterior fossa structures were not well visualized. Therefore, a lumbar pneumoencephalogram was attempted. Finally, in order to define the aqueduct 1.5 ml. of Pantopaque was introduced into the right lateral ventricle.

On March 4, 1960, a craniotomy was performed to permit infra- and supratentorial exploration. The cerebellar tonsils were herniated through the foramen magnum, more marked on the left. Both cerebellar hemispheres were bulging, more so on the left. There was no evidence of arachnoiditis and the fourth ventricle was not obstructed. A large thin-walled cyst, containing clear fluid, was found between the anterior surface of the cerebellum and the tentorium. The cyst had stretched the tentorial incisura markedly and the major portion of the cyst lay supratentorially. The cyst contents were aspirated and the infratentorial dome of the cyst was removed, thereby establishing communication posteriorly with the superior cerebellar cistern. Indigo-carmin injected at this time into the lateral ventricles quickly appeared in the fourth ventricle, but did not enter the

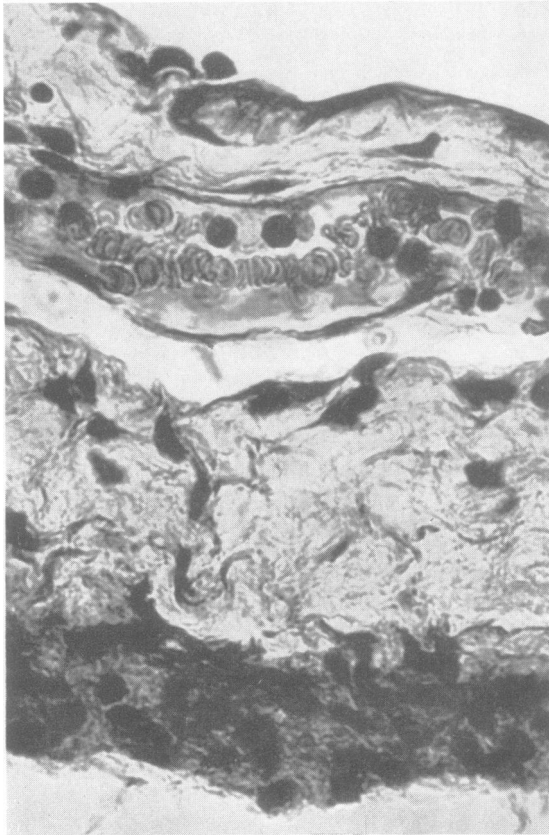


FIG. 3.—Photomicrograph of cyst wall. The inner surface is at left. The wall consists of loose connective tissue cells with an occasional capillary. Haematoxylin and eosin  $\times 650$ .

cyst. Microscopic examination of the wall of the cyst revealed a 'benign cyst composed of mesodermal structures of arachnoidal origin'. No inflammatory cells were seen in the cyst wall and there was no pigmentation to suggest that the cyst had resulted from haemorrhage (Fig. 3).

The post-operative course was complicated by a subcutaneous collection of fluid which necessitated a second surgical procedure to close a pinpoint dural fistula overlying the cisterna magna. Thereafter the patient did well and was discharged with much less spasticity. Improvement continued for the next six months. On November 30, 1960, the patient was re-admitted to the hospital because of vomiting of two weeks' duration. Physical examination revealed an irritable child with a soft bulge in the suboccipital area. The head circumference was now 54.5 cm. and the papilloedema and spasticity had recurred. It was thought safe to perform a lumbar pneumoencephalogram because of the previous suboccipital decompression. The encephalographic findings were identical with those demonstrated originally.

On December 2, 1960, the cyst was approached through an occipital osteoplastic flap. At this operation a medical artist was present and his concept of the lesion is illustrated in Fig. 4. On this occasion a 1 cm. communication was established between the posterior end of the third ventricle and the anterior wall of the cyst. A second and larger communication was made by removing a portion of the medial wall of the left occipital horn and fixing the ependyma of the horn to the cyst wall with two silver clips. Fluid aspirated from the cyst contained 93 mg. % protein compared with 4 mg. % in the ventricular fluid and 8.5 mg. % in the lumbar spinal fluid. The post-operative course was uneventful. All signs of increased intracranial pressure subsided. Since the time of writing, the child has been seen for his 16 months' follow-up examinations. He continues to show motor and intellectual improvement.

#### Radiological Features

Cysts of the quadrigeminal cistern can be classified radiologically as posterior incisural lesions as described by Taveras (1960). This lesion produces obstructive hydrocephalus by stretching the aqueduct forward and inferiorly, compromising its lumen. The posterior margin of the third ventricle is encroached upon and is displaced forward in a very smooth, rounded fashion. The suprapineal recess is elevated and bowed around the cyst. When the cyst extends infratentorially it will depress, flatten and widen the fourth ventricle. Obviously the quadrigeminal cistern cannot be filled with gas. The pontine and crural cisterns may be smaller than usual due to brain-stem displacement but they are not obliterated. In our case the posterior portion of the roof of the third ventricle was indented by a soft tissue mass, probably caused by forward buckling of the splenium of the corpus callosum.

The important point in diagnosis would appear

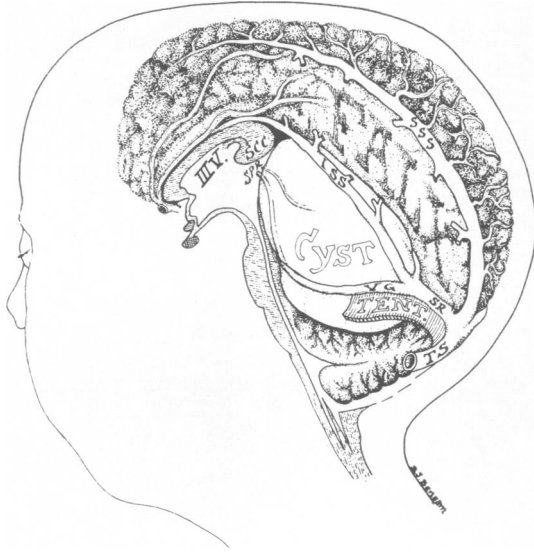


FIG. 4.—Artist's concept of the operative findings.

Note the marked forward displacement of the third ventricle, and the downward displacement of the anterior cerebellum. The lesser veins of Galen are separated and they and the great vein are displaced downward by the floor of the cyst.

- III V = third ventricle
- SCC = splenium of corpus callosum
- SPR = suprapineal recess
- SSS = superior sagittal sinus
- ISS = inferior sagittal sinus
- Tent = tentorium
- VG = vein of Galen
- SR = sinus rectus
- TS = transverse sinus

to rest upon recognition of the marked stretching and bowing of the structures in the vicinity of the posterior tentorial notch, suggesting the presence of a relatively soft mass such as a cyst, and permitting the development of marked internal hydrocephalus. Lateral displacement of the midline structures, *i.e.*, the third ventricle, aqueduct, and fourth ventricle, is not present because the cyst originates from the centrally situated quadrigeminal cistern.

Absolute radiological diagnosis would be possible only if contrast material entered the cyst, but this is unlikely unless the cyst were tapped intentionally or inadvertently during gas ventriculography. However, this gas-filled cavity should not be confused with the enlarged but not displaced suprapineal recess occasionally seen in association with hydrocephalus, *i.e.*, 'suprapineal diverticulum'.

Angiography was not performed in this instance. However, observations made at surgery indicated a forward bowing and separation of the lesser veins of Galen as they passed around the cyst before joining to form the great vein of Galen (Fig. 4).

#### Radiological Differential Diagnosis

The first step in differential diagnosis should be the elimination of lesions other than incisural masses which cause symmetrical dilatation of the lateral and third ventricles.

Infratentorial lesions such as supra- or infrafastigial cerebellar masses can be excluded because they kink the aqueduct forward with the apex of the kink at the intercollicular level. Upward herniations through the tentorium may mask these classical findings (Twining, 1939).

Masses anterior to the aqueduct are easily differentiated because they bow the aqueduct posteriorly and/or laterally and may rotate the brain-stem, aqueduct, and fourth ventricle. Complete obstruction of the aqueduct is rare with these anterior lesions, and when the third ventricle is impinged upon it is usually the floor of the third ventricle rather than the posterior portion which is distorted. If the basal cisterns are delineated with gas they are partially obliterated.

Stenosis of the aqueduct characteristically occurs at its most rostral portion and causes no local deformity of the third ventricle.

The four lesions which are most likely to be confused with a cyst of the quadrigeminal cistern are pinealoma, aneurysmal dilatation of the vein of Galen, midline meningiomas arising from the posterior tentorial notch, and central gliomas invading the quadrigeminal plate. Pinealomas encroach most severely upon the posterior end of the third ventricle. The pineal recess is obliterated and the suprapineal recess is displaced upward or obliterated, depending upon the size of the pineal tumour. However, since pinealomas are usually solid tumours, total occlusion of the aqueduct tends to occur early and therefore marked elongation and bowing of this structure would be less likely to occur. Infratentorial extension of pinealomas is rare. At angiography the distal portion of the internal cerebral veins and the vein of Galen should be raised and bowed upward and backward. However, none of these differential points is absolute.

Aneurysm of the vein of Galen may produce all of the radiological signs of the lesion under discussion here when studied by encephalography, but angiography should make the differentiation. In retrospect such a procedure might well have been performed in our patient, although the clinical picture was not suggestive of a vascular lesion (Russell and Nevin, 1940). Posterior incisural meningiomas may produce similar encephalographic findings although the manifestations of a relatively soft mass will be less likely to occur (Taveras, 1960). The displacement of the deep veins at angiography is very similar to that visualized at surgery in our case.

Gliomas invading the quadrigeminal plate will straighten the aqueduct and displace it forward but are unlikely to bow and stretch it as happened with this cystic lesion (Lysholm, 1946). Lateral displacement of the aqueduct would be likely because the gliomas are usually not strictly midline lesions.

### Discussion

The aetiology and mechanisms of cyst formation in the subarachnoid pathways are not clearly understood. It is likely that they result from more than one cause. Hamby and Gardner (1935) thought in their case that the cyst had formed from a ventricular diverticulum, but since their patient had suffered a fall they could not exclude trauma as a causative factor. Alexander's (1953) patient had had severe pertussis at the age of 5 months so the cyst could not necessarily be considered congenital in origin. In Dott and Gillingham's (1958) case there was a history of 'minor birth trauma' so that again trauma could not be excluded as the basic mechanism for the cyst formation.

Our patient had sustained a linear skull fracture but whether the trauma which produced the fracture was the cause of the cyst or the result of the child's ataxia secondary to the presence of the cyst, cannot be ascertained. The absence of pigment or inflammatory changes in the cyst wall seem to make trauma unlikely as the precipitating factor.

Dott and Gillingham (1958) postulated that some inflammatory factor such as blood or exudate might initiate a pathological process which could obstruct

the outlet of any of the subarachnoid cisterns. Cerebrospinal fluid would then continue to enter the partially blocked cistern as it was 'milked' or pumped forward by transmitted pressure waves from nearby pulsating arteries. This mechanism would then gradually result in distention and increased pressure in the obstructed cistern.

From the evidence it is impossible to state whether the cyst in our case represented an acquired or a congenital lesion, although we consider the latter the more likely possibility.

### Summary

An example of obstructive hydrocephalus secondary to a large cyst of the quadrigeminal cistern is reported.

The salient radiological features and the differential diagnosis of this lesion are discussed.

The importance of suspecting and directly treating these benign masses is stressed.

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