INFANTILE HYDROCEPHALUS CAUSED BY ARACHNOID CYST

CASE REPORT

A. J. LEWIS, M.B.

Department of Pathology, Children’s Hospital, Winnipeg, Manitoba, Canada

(Received for publication November 6, 1961)

This report describes a congenital arachnoid cyst. These cysts rarely are reported in infancy, although Oliver17 described 1 in a child of 10 months, and Klein and Brogly11 reported 1 in an infant of 1 month. They usually are not considered in the differential diagnosis of hydrocephalus. The patient in the case reported here died at the age of 71/2 months shortly after operation, the provisional diagnosis having been an arachnoid cyst, on radiological grounds. It is considered of interest because of the early age of the patient, and the presence of choroid plexus-like tissue in the wall of the cyst.

CASE HISTORY

Clinical Summary. The patient was a boy who weighed 8 lbs, 5 oz. (3800 gm.) at birth. In spite of hyperemesis, gestation continued to term, and labor was uneventful. His mother was aged 32 years and his father 44; they and a sibling aged 5 are alive and well. Condition at birth was good and the circumference of the head was thought to be normal.

He was admitted to hospital at age of 7 months with hydrocephalus, the circumference of the head being 21 1/2 in. He had fallen 18 in. off a couch at age 3-4 months, but apparently was uninjured and did not lose consciousness. Examination on admission revealed no abnormalities except for the large head. Count of cells in cerebrospinal fluid was 1 lymphocyte on first examination and 48 (0/66 per cent neutrophile) 4 days later. Protein was 11 mg. per cent on both occasions.

Roentgenogram of the skull confirmed hydrocephalus, and ventriculography (Dr. A. E. Childe) suggested a space-occupying lesion in the posterior fossa, possibly herniating through the apex of the tentorium. This was thought to be a congenital cystic lesion. Operation on the 7th hospital day (Dr. D. Parkinson) disclosed a cyst bulging up through the tentorial notch; when the tentorium was opened, this was seen to occupy the upper part of the posterior fossa, compressing the cerebellar hemispheres downwards and laterally. An opening was made in the bulging posterior wall of the 3rd ventricle. The wall of the cyst was stripped until excessive bleeding was encountered over the roof of the 4th ventricle. The cystic fluid was clear, but none was obtained for analysis. A biopsy of the wall of the cyst was taken. About 1 1/2 hours after operation, the infant collapsed with a rapid weak pulse and cyanosis, and died.

The biopsy showed delicate, almost avascular tissue lined on at least one side by plump mesothelial cells. There was no inflammatory infiltrate.

Autopsy Report. The body was that of a well-developed male infant. Circumference of the head was 21 in. (53.5 cm.). The head was symmetrical, and the sutures were separated. The base of the skull was of normal shape. The dura mater was normal and not adherent to the arachnoid. The pia-arachnoid was not abnormally adherent to the underlying brain and showed no thickening.

The superior aspect of the cerebellum was distorted by a mid-line depression measuring 4×1×1 cm. (Fig. 1). The inferomedial aspect of the right occipital lobe was distorted by a depression measuring 6×3×2.5 cm. (Fig. 2). Subject to the disturbance created by surgery, the cysts filling these depressions appeared to be continuous at the tentorial edge. They were roofed and lined by translucent membrane which was continuous peripherally with the pia-arachnoid. The roof contained only a few delicate vessels. The cerebral hemispheres collapsed, and weight of the empty brain was 585 gm. The lateral and 3rd ventricles were markedly dilated, but the aqueduct and 4th ventricles were compressed and small. The gyral pattern underlying the cysts was normal.

The lungs showed moderate edema. No other abnormalities were noted.

Histological Report. Those parts of the walls of the cyst applied to the brain were normal pia-arachnoid, except for showing recent, slight, subpial and subarachnoid hemorrhage. Inflammatory cells and iron pigment were absent. In just one area of the supratentorial cyst, normal pia was overlain by a vascular, collagenous membrane, lined on the cystic surface by cubical cells. These were thrown up into villi resembling choroid plexus (Fig. 3). The roofs of the cysts were of almost avascular delicate membrane lined by flattened cells, with no inflammatory features. Underlying brain showed distortion but no obvious defect.

The cerebral hemispheres showed atrophy of white matter. The lungs showed early bronchopneumonia and edema. No other significant histopathology was found.

DISCUSSION

Cases of arachnoid cyst or cystic arachnoiditis reported in the literature appear to fall into three pathological categories. Those described by Horrax5 and by Craig4 were associated with gross thickening of the meninges extending beyond the confines of the cyst, and with arachnoid adhesions. They are assumed to be inflammatory in
origin, and Horrax noted their frequency in years following high incidence of influenza and encephalitis lethargica. A different pathological group is made up of cysts following fractures of the parietal region with unhealed dural tears, noted by Taveras and Ransohoff and by Peyser and Weissberg. These are associated with bony overgrowths within the cavity of the skull, and the cysts are often multilocular. The third group are those not associated with arachnoid adhesions or with gross pathological evidence of trauma (though there may be a history of trauma at some time in the past). Twenty-seven such cases in the literature have been reviewed, in addition to the remarkable series of Klein and Brogley, who saw 25 cases of cisterna-magna cysts within a short period at one centre. Histology in their cases showed “thickening of the cyst-wall” but no inflammatory exudate, and there was no mention of adhesions or trauma. Ages of patients ranged from 1 month to 40 years. Whatever the etiology in these cases (and such a series may well indicate an epidemic factor), the cysts appear to differ from those of “sporadic” cases in that marked thickening is present.

In 18 of the 27 “sporadic” cases the cysts were

Fig. 1 (left). Superior aspect of cerebellum, showing mid-line depression. Arachnoid has been stripped from the right side.

Fig. 2 (right). Coronal section of cerebral hemispheres behind the splenium, viewed from anterior aspect. The severe hydrocephalus is apparent, and the distortion produced by the supratentorial part of the cyst is shown by the right hemisphere.

Fig. 3. Section of choroid plexus-like tissue in wall of cyst. The photomicrograph does not include underlying pia. Note the collagenization and large vessel, but absence of core in the villous processes.
HYDROCEPHALUS CAUSED BY ARACHNOID CYST

subtentorial, with ages of patients at diagnosis ranging from 10 months to 51 years. They presented signs of increased intracranial pressure, as cases of hydrocephalus and others as cases of acoustic neurinomas (found to be in the cerebellopontine angle). In 9 cases the cysts were above the tentorium, in or near the Sylvian fissure. They usually presented signs of space-occupying lesions, 2 with epilepsy, but 2 of them were accidental autopsy discoveries. Ages at diagnosis ranged from 10 to 79 years. Such lesions are variously attributed to injury, infection or malformation.

A history of trauma was elicited in 7 cases. Thompson reported the case of a man who injured his leg, but not his head, in a parachute jump. Two months later he was found to have a posterior-fossa cyst 5×3×4 cm in size, thin-walled and infiltrated with round cells and occasional granulocytes. Trowbridge and French described a man with a posterior-fossa cyst discovered 5 years after he had struck his head without losing consciousness; this showed no inflammatory infiltrate. The authors considered this to have been probably congenital. In a case discussed by Aring et al. a girl had been struck by a car and was unconscious for 24–48 hours; 26 years later she was found to have a posterior-fossa cyst with a thickened roof and a few inflammatory cells. Oliver reported a man who, shortly after a chest injury sustained at football, was found to have a large thin-walled cyst of the Sylvian fissure. This showed recent hemorrhage in the wall, but no inflammatory change of long standing, and Oliver considered, on the basis of size and microscopic findings, that the cyst antedated the injury. Nichols and Manganelli noted a cyst of the cerebellopontine angle discovered 3 years after the patient fell from a wagon and was unconscious for 20–30 minutes. The cyst was described as blue-domed and opaque, without report of histology. Tiberin and Gruszkiewicz reported a boy who fell from bed, and was briefly unconscious at the age of 3; 3 months later a swelling developed in the left temporal area. At age 10 he was found to have a middle-fossa cyst, lined by arachnoid with psammoma bodies. Montricq described a posterior-fossa cyst with a subdural hematoma found 2 weeks after a head injury. The size of the cyst indicated its presence for more than 2 weeks. Thus in 3 of these 7 cases the authors felt that the cyst probably antedated injury, and it is apparent that in Thompson's case, too, the cyst was very large for the length of history. In 2 of these 4 cases the patients were not struck on the head.

Only 2 cases had any history of infection, and in 1 (Case 4 of Starkman et al.) the authors considered the meningitis independent of the presence of a middle-fossa cyst. García Bengochea and Blanco reported a cyst presenting as a tumor of the cerebellopontine angle, with no gross or microscopic evidence of inflammation. There was a history of possible otitis externa 31 years previously.

There remain 18 cases with no history of infection or trauma; 5 had no histological report, and 9 cysts showed no evidence of inflammation. In Case 2 of Maneke there was inflammatory infiltrate, attributed by the author to a possible intrauterine infection although there was no history suggestive of this; the child was aged 5½ years and also had congenital heart disease. In 2 other cases the cysts showed occasional lymphocytes, and 1 a trace of iron pigment. Klein and Brogley's cases also belong in this group, and the cysts showed thickening, presumably collagenous.

It is apparent that there are no cases with even a suggestive history of past meningeal infection. This does not disprove the role of occult infection, but prompts careful consideration of alternative etiological factors. The significance of trauma is obvious, but whether it is a causative or precipitating factor is less evident. When injury precedes the discovery of the cyst by a long period, it may not be possible to assess which came first on pathological grounds. In all 7 cases with a history of trauma, symptoms closely followed the injury, but in 4 of these there may well have been a cyst present at the time of injury. In Oliver's case, fresh hemorrhage into the wall of the cyst was seen microscopically.

A few of these cases showed features suggestive of malformation. Starkman et al. described 3 cases of a cyst within a split arachnoid; in the present case the cyst may have been intra-arachnoid, but this is difficult to assess after surgical intervention. Bucy reported a case of posterior-fossa cyst, which contained a tuft of choroid plexus in the wall. The author considered this a developmental error. Hamby and Gardner described an ependymal cyst of the supracallosal region, possibly formed in the same way. In the present case, a part of the cyst lining mimics choroid plexus; however, correspondence is not exact, and there is no real collag enous stroma or significant vascularization of the villous tufts. It is not possible to exclude the formation of these structures by metaplasia. Other reports did not note the presence of choroid plexus, and Schreiber and Heppner suggested that most of the cysts are caused by incomplete canalization of the subarachnoid space. The resulting pouch-like spaces are thought to be distended by cerebrospinal fluid as a result of arterial pulsation.

Etiologically, it seems that these cysts are a mixed group. Some, as described by Horrax and Craig, showed clear evidence of more widespread inflammation (not necessarily infection) and others showed manifest injury. In many, when
definite evidence of neither factor is forthcoming from the gross appearance of the lesion, the cyst, on present evidence, was probably congenital. It is not possible to exclude traumatic origin, though apparently injury need not be severe or even directly to the head. At the least, trauma often is important as a precipitating factor.

SUMMARY

A case of arachnoid cyst with choroid plexus-like tissue in the lining is reported. Literature of arachnoid cysts is reviewed briefly, and a group is separated off in which there is neither adhesive arachnoiditis nor gross evidence of injury. Many of these cysts probably are malformations.

REFERENCES