Supratentorial arachnoidal cyst associated with hydrocephalus

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Supratentorial arachnoidal cysts are usually associated with signs of fracture of the skull but hydrocephalus has not been reported in connexion with such cysts. In none of the 18 cases reviewed by Tiberin and Gruszkiewicz (1961) was hydrocephalus found. A 3-year-old child was recently seen with signs of hydrocephalus and a large arachnoidal cyst of the right cerebral hemisphere. The rarity of such an association warrants this report.

CASE HISTORY

A 3-year-old girl was admitted to the Department of Neurosurgery on 14 August 1962 with a presumptive diagnosis of progressive hydrocephalus. Delivery had been at term and without complications. When she was 6 weeks old she fell out of bed, injured her head, and was rendered unconscious for a few minutes. She was admitted to another hospital, but no clinical details are available. It was noticed that subsequently her head grew larger out of all proportion until the age of 9 months, when this abnormal growth ceased.

On admission, the child appeared underdeveloped both physically and mentally. She was unable to walk, sit, or hold her head up without support. There was a marked spastic right hemiparesis. The shape of the head appeared hydrocephalic. The skull was asymmetrical, the right half bulging and wider than the left. Exophthalmos was noticed on the right and a positive 'sunset phenomenon' in both eyes. The circumference of the head was 63 cm. The anterior fontanelle was still widely open, tight and bulging. Both optic nerve disks were pale. Visual acuity and visual fields on confrontation appeared normal. The range of eye movements was full. The tendon reflexes were slightly exaggerated on the right.

Radiographs of the skull showed tremendous enlargement of the head with bulging of its right half. The electroencephalogram showed low-voltage slow waves in the right parieto-occipital region and a minor disturbance in the left temporal region. It was thought that this record was compatible with a collection of fluid over the right parietal region of the brain.

A lumbar puncture was performed and clear cerebrospinal fluid under a pressure of 180 mm. water was obtained. It contained 30 mg/100 ml. protein, 120 mEq./l. chlorides, and 10 lymphocytes and 17 polymorphonuclear cells per c.mm. The mastix test and Wassermann reaction were negative. An attempt at pneumoencephalography was unsuccessful; ventriculo-

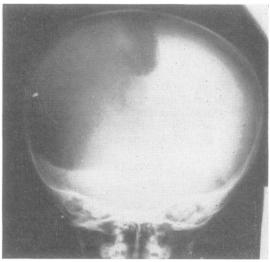
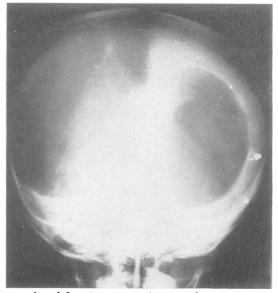


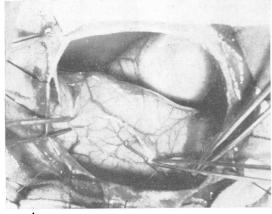
FIG. 1.



FIGS. 1 and 2. Antero-posterior view showing enormous cyst covering the right hemisphere (above), and below air-filled cyst and enlarged left ventricle.



FIG. 3.





FIGS. 3 and 4. Operative field after opening of the dura (above), showing the lateral cyst wall and displaced right hemisphere, and (below) after excision of cyst wall and evacuation of fluid. The displaced hemisphere is raised.

graphy was, therefore, deemed advisable. Xanthochromic fluid was obtained by a puncture through the right side of the anterior fontanelle, containing 200 mg. of protein per 100 ml.; 160 ml. of fluid was exchanged with air. Radiographs showed air only on the right side (Fig. 1). Therefore, puncture through the left side of the anterior fontanelle was accomplished. Clear fluid was obtained with a protein content of 30 mg. per 100 ml.; 80 ml. was exchanged with air. Radiographs showed tremendous enlargement of the left lateral ventricle. On the right side,

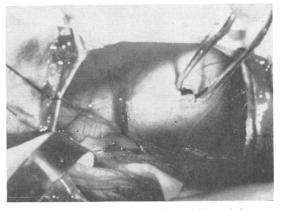


FIG. 5. Opening of third ventricle through distended corpus callosum.

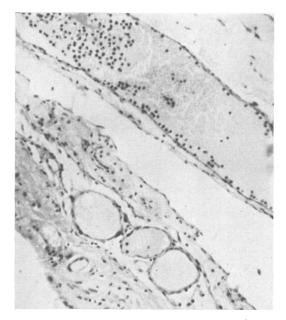


FIG. 6. Two parts of the cyst wall consisting of loose connective tissue with many thin-walled blood vessels (arachnoides). Haematoxylin and eosin \times 320.

however, the air extended toward the internal table of the skull laterally, and up to the midsagittal plane medially (Fig. 2).

Considering the result of the air studies and the different protein contents in the fluids obtained, it was suspected that a cystic cavity was present on the right side of the brain.

OPERATION On 29 August under general anaesthesia, a right fronto-parieto-temporal bone flap was elevated. The dura was transparent and very tense, and fluid and

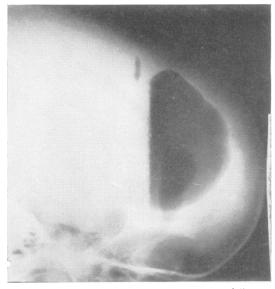


FIG. 7. Brow-up pneumoencephalogram at follow-up, showing filling of both lateral ventricles.

bubbles of air could be seen through it. After opening the dura, it was noted that the major part of the right hemisphere was covered by an enormous cyst with the arachnoid forming its walls. The temporal lobe was atrophic, with small and flattened gyri. In the walls of the cyst there were several blood vessels continuous with those of the pia mater covering the brain (Figs. 3 and 4).

The cyst wall was excised and part of it removed for examination. Clear xanthochromic fluid escaped, and about 750 ml. was evacuated from within the cyst. After the cyst had been emptied a white structure was seen, which was punctured with a brain cannula and clear fluid obtained, probably from the right lateral ventricle. Ventriculostomy was therefore performed (Fig. 5), and the dura closed in the usual way.

The histological examination of the wall of the cyst showed a membrane similar in structure to arachnoid (Fig. 6).

POST-OPERATIVE COURSE This was uneventful except for some elevation of temperature during the first fortnight. Repeated lumbar punctures following the operation showed that pressure and composition of the cerebrospinal fluid had returned to normal by the end of three weeks.

The patient was discharged from hospital 25 days after the operation, much improved. She was able to hold up her head and sit without support. The weakness of the right extremities had almost disappeared, and the fontanelle was no longer bulging. The child was readmitted for follow-up investigation two and a half months later. Control pneumoencephalography showed free communication within the ventricular system with filling of both lateral ventricles. No air was seen in the cranial subarachnoid spaces, obviously showing that the opening made in the roof of the third ventricle at operation had sealed off (Fig. 7).

DISCUSSION

The aetiology and pathogenesis of arachnoid cysts are not well understood. They have been divided into four groups according to their origin.

1 Developmental malformation and localized brain atrophy, a condition for which Robinson (1958) suggested the term 'external hydrocephalus' which, in his opinion, was due to localized agenesis of the brain.

2 Inflammatory, as described by Horrax (1924), de Martel and Guillaume (1930), and others, in which otitis media, nasal sinus infections, meningitis have been found.

3 Traumatic, with fracture of the skull as reported by Taveras and Ransohoff (1953), Peyser and Weissberg (1961), and others, in which cases localized post-traumatic brain damage was found with laceration of the dura and the formation of porencephalic and leptomeningeal cysts.

4 Traumatic, usually after minor injury, but without fracture of the skull: Cohen (1927), Childe (1953), Tiberin and Gruszkiewicz (1961), and others have reported such cysts, usually located in the middle cranial cavity.

After even minor trauma without skull fracture, localized subarachnoid bleeding may occur with a haematoma forming. Adhesions develop because of difference in osmotic pressure, the haematoma increases in size, and a collection of fluid rich in protein is formed.

Supratentorial arachnoid cysts are usually located in the middle cranial cavity (Sylvian or Rolandic fissure) or between the hemispheres. Clinically they may remain silent and will only be detected on postmortem examination. If, however, they cause focal neurological signs, these are usually of a minor degree. Air studies and arteriography may be helpful in making the diagnosis, but ultimately only surgical exploration will establish the nature of the lesion.

Arachnoid cysts which are the result of a developmental anomaly were shown by gross and microscopic examination to lie between two arachnoid membranes. They are, therefore, in fact intraarachnoid cysts (Starkman, Brown, and Linell, 1958). Other 'cysts', which are the result of brain atrophy, are not true cysts but localized enlargement of the subarachnoid space. It may be extremely difficult to differentiate between the two conditions.

Differential diagnosis will have to take into consideration subdural hygromas and subdural haematomas, which are characterized by the formation of neo-membranes around the fluid collection. Histological examination of the cyst wall will help in arriving at a correct diagnosis.

The present case seems to be one of congenital hydrocephalus associated with a huge arachnoid cyst. Though a developmental anomaly cannot entirely be excluded, the traumatic origin of the cyst should seriously be considered.

SUMMARY

The case of a 3-year-old girl with hydrocephalus and an arachnoid cyst is presented. It resulted in pressure on the right cerebral hemisphere and local bulging of the skull. Surgical treatment resulted in remarkable clinical improvement.

The origin of such cysts is discussed.

Our thanks are due to Mrs. B. Kulka for the photo-graphs.

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