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Interhemispheric Arachnoid Cyst with Agenesis of Corpus Callosum

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Arachnoid cysts are benign developmental cysts that occur throughout the cerebrospinal axis in relation to the arachnoid membrane and the subarachnoid space(l). Intracranial arachnoid cysts usually occur in close proximity to arachnoid cisterns, most often in the sylvian fissure(2) and they become symptomatic in early childhood(3). We report an interhemispheric arachnoid cyst associated with agenesis of corpus callosum in a neonate.

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Manuscript received: February 3,1997
Initial review completed: February 27, 1997
Revision accepted: April 4, 1997

Case Report

A term male baby born vaginally to a grand multigravida mother with Apgar scores of 5 and 7 at 1 and 5 minutes, respectively and weighing 3500 grams had focal clonic seizures of left upper limb with associated orofacial movements on the second day of life. Clinical examination of the neonate was essentially normal with a head circumference of 37 cm and length of 54 cm.

Investigations which included arterial blood gases, serum electrolytes, blood sugar, serum calcium and cerebrospinal fluid examination and culture were normal. Ultrasonography of the skull revealed 70 x 81 x 70 mm anechoic well defined cystic structure in the midline. Computed tomography of the brain revealed a large midline interhemispheric cyst displacing the brain parenchyma suggestive of an arachnoid cyst. The lateral ventricles, third ventricle and fourth ventricle were well visualized. Agenesis of corpus callosum was also seen. No other brain malformation or atrophy was noted (Figs. 1 & 2). Neonatal seizures were controlled with phenobarbitone and phenytoin therapy. The neonate was discharged on day fifteen

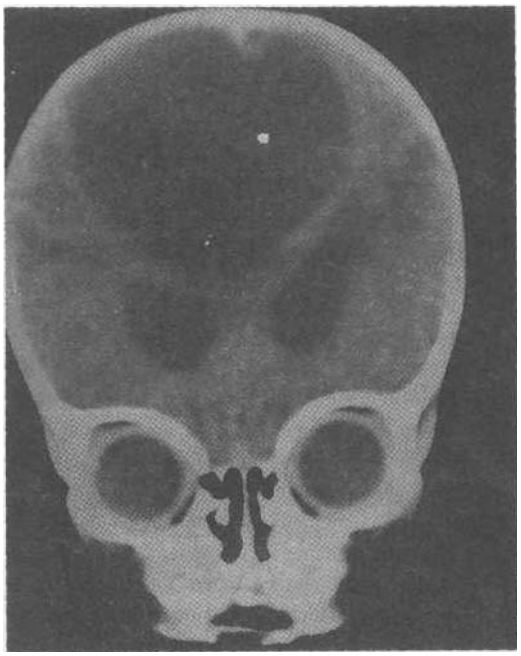

Fig. 1. Computed tomography of the brain showing large midline interhemispheric cyst with displacement of lateral ventricles.

of life on oral phenobarbitone. Since the patient was neurologically normal, no neurosurgical intervention was contemplated.

Discussion

Arachnoid cysts are developmental collections of cerebrospinal fluid contained within a lining leptomeningeal membrane that were first described in 1831(4). They account for 1% of all atraumtic intracranial mass lesions(l).

Though arachnoid cysts can be found intracranially at any location, only 5% of them are located in the interhemispheric fissure(2). Interhemispheric arachnoid cysts tend to be enormous in size because of the accommodation of the cyst by the brain and expanding calvarium. The lining of arachnoid cyst is usually smooth showing hyperplastic arachnoid cells and thick layer of collagen(2) and there is no evidence of choroid plexus, inflammation or tumor. The fluid content is most often clear, colorless and similar to cerebrospinal fluid but the protein content varies.

There is a curious association of interhemispheric arachnoid cyst with partial or complete agenesis of the corpus callosum(5,6). Embryologically, arachnoid cysts probably arise as a result of anomalous splitting and duplication of the endomeninx during neural tube fold(5). It may be postulated that the presence of a large arachnoid cyst in the interhemispheric tissue may mechanically impede the development of interhemispheric association fibres(5) but such a possibility is negated by the fact that a very small arachnoid cyst is sometimes found in the interhemispheric area associated with complete agenesis of corpus callosum(7). Embryologically, the corpus callosum developes from the commissural plate that lies in close proximity to the anterior neuropore. The first fibres forming the corpus callosum appear anteriorly near the lamina terminalis at around twelve weeks and crossing of the fibres is complete by twenty-two weeks.

Clinical manifestations of an arachnoid cyst are often mild relative to its large size(8). As they are very slow growing cysts, they generally become symptomatic in early childhood(3). In the absence of associated brain anomalies, agenesis of corpus callosum is usually diagnosed in childhood during investigation of mental retardation or hydrocephalus(8).

Computerised tomography is essential for definite diagnosis of arachnoid cysts. The cyst appears as a non calcific low density extraparenchymal mass with smooth and clearly defined borders and rounded shape when situated in the midline(3). Agenesis of corpus callosum is visualized as separation of the lateral ventricles(9).

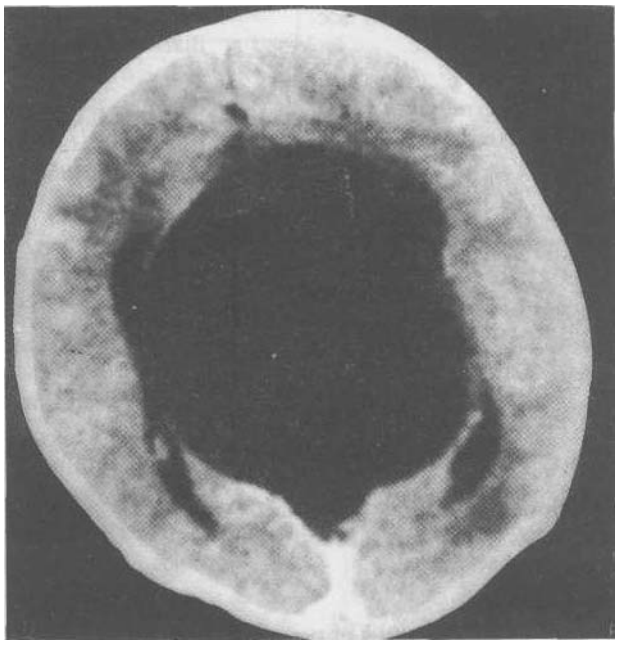

Fig. 2. *Computed tomography of the brain (coronal section) showing the interhemispheric cyst with agenesis of corpus callosum.*

Accidental discovery or minor treatable symptoms resulting from arachnoid cysts usually do not warrant surgery. When symptoms are severe or significant, surgical intervention to decompress the cyst including shunting procedure is required. Cyst-peritoneal shunting is usually the best initial procedure(10).

Acknowledgement

Authors thank Dr. (Mrs.) P.M. Pai, Dean, Seth G.S. Medical College and K.E.M. Hospital for permitting them to publish this case.

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