Interhemispheric cyst of neuroepithelial origin in association with partial agenesis of the corpus callosum

Case report and review of the literature

LILLIAN C. SOLT, M.D., JOHN H. N. DECK, M.D., ROGER SCOTT BAIM, M.D., AND KARL TERBRUGGE, M.D.

Departments of Pathology and Radiology, The Toronto Western Hospital, Toronto, Ontario, Canada

A total of 10 interhemispheric cysts have been described, in association with complete or partial agenesis of the corpus callosum.2,3,7,8,12,15,19 The histological features of the cyst wall in these cases have been variable. Tissues identified as components of the reported cysts include arachnoid,12 glial tissue,19 choroid plexus epithelium, and ependyma.9 Most of the cases of interhemispheric cyst and partial or total agenesis of the corpus callosum were found in young children,7,9,19 and were reported before the advent of computerized tomography (CT).

This report describes a symptomatic adult in whom a large interhemispheric cyst was demonstrated by CT scan to be immediately adjacent to the posterior third ventricle, which was enlarged and higher in position than normal. The cyst was partially lined by cuboidal epithelium, and tufts of choroid plexus were seen projecting into it. The CT features indicated, and the operative findings were consistent with, partial agenesis of the corpus callosum.

Case Report

This 64-year-old man was admitted to the hospital on March 22, 1979, for investigation of a severe throbbing frontal headache of 6 months' duration. Weakness of both legs was first noticed in 1974. Dizziness and occasional true vertigo, diagnosed as Ménière's disease in 1978, were also documented. He had noticed a 25-lb weight loss over a period of 5 months before his admission. He had been a heavy smoker for 40 years, and consumed up to 24 bottles of beer a week.

Examination. On admission, his vital signs were normal. The general examination was unremarkable. Neurological examination revealed normal cranial nerves and normal-appearing optic discs. He had a positive Romberg's sign and an abnormal wide-based gait, but finger-to-nose test was normal. Long-term memory was normal, but he had slight impairment of short-term memory. Laboratory tests, including blood counts, serum electrolytes, creatinine, and alkaline phosphatase, tests for venereal disease, and urinalysis were normal. Four serial sputum cytology specimens were negative for malignant cells. A chest film was reported normal.

Plain radiographs of the skull were unremarkable. A CT scan was obtained following intravenous injection of a 90-cc bolus of Hypaque. Axial views (Fig. 1) revealed moderate atrophy of the cerebral cortex and cerebellum, and a homogeneous cystic mass of the
Fic. 1. Contrast-enhanced computerized tomography scans, axial projection, showing generalized cerebral atrophy. Note the cystic mass lesion arising in the midline above the posterior third ventricle, and the lack of any intervening corpus callosum. An incomplete rim of increased density is seen (arrow). The mass itself is of the same density as cerebrospinal fluid.

FIG. 2. Contrast-enhanced computerized tomography, coronal projection. The curvilinear density surrounding the mass within the interhemispheric fissure shows apparent attachment to the falx (arrow). Note midline contiguity of the cystic lesion with the roof of the lateral ventricles (two center scans).

density of cerebrospinal fluid (CSF) within the interhemispheric fissure arising just above the third ventricle posteriorly. The cyst was essentially midline, but extended to the left as it approached the falx cerebri. It was incompletely outlined by a thin curved line of increased density. Since the study was performed only after contrast enhancement, it was not clear from the CT pictures whether the curvilinear density was due to vessel enhancement (of the arachnoid vessels surrounding the mass or of the cyst wall itself), or to calcification of the cyst wall outlined externally and internally by low-density CSF.

Coronal views (Fig. 2) showed splaying of the bodies of the lateral ventricles by the interhemispheric cyst. The third ventricle was enlarged and higher than normal in position. The corpus callosum appeared to be absent at the level of the posterior third ventricle, but it was of normal thickness anteriorly. In the abnormal area, the interhemispheric fissure was noted to come in contact with the roofs of both lateral ventricles.

Bilateral common carotid arteriograms were obtained via the femoral approach (Fig. 3). Lateral views in the early and late venous stages of the left carotid artery injection showed evidence of an avascular mass in the frontoparietal region. No tumor vascularity of the cyst wall was demonstrated. The anteroposterior projection showed similar findings, with displacement of both anterior cerebral arteries around the mass.

**Operation.** On the 9th hospital day the patient underwent a left frontoparietal craniotomy. Adhesions between the cyst and dura were coagulated; this exposed the entire cyst, which measured \(4.5 \times 4.0\) cm. A plane of cleavage was easily established between the cyst and the adjacent parietal lobe, and small arterial branches to the cyst wall were interrupted. The cyst was located at the level of the Rolando fissure and contained a crystal-clear fluid similar to CSF. The cyst extended across the midline to the right side, from which it was removed readily. The corpus callosum was not identified. Due to the accidental rupture of the cyst during excision, examination of the fluid was not performed.
Neuroepithelial interhemispheric cyst

Postoperative Course. In the immediate postoperative period, the patient developed a right hemiplegia and expressive dysphasia, both of which gradually improved. The frontal headache disappeared in the immediate postoperative period. Since there was no evidence of raised intracranial pressure and the severe frontal headache was relieved after surgery, the headache is best attributed to traction or pressure on dural structures by the interhemispheric cyst.

The cerebellar signs (positive Romberg’s sign and ataxic gait) persisted, and are attributed to atrophy of the cerebellar vermis which was documented in the CT scan.

Pathological Examination. The operative specimen consisted of a round cystic structure, 4.0 × 3.5 cm in diameter. The outer surface showed a few thin-walled blood vessels, while on the inner aspect, the surface had a pearly appearance with some irregular, elevated white-gray patches. The specimen was fixed in 10% buffered formalin. Many sections were taken and stained with hematoxylin and eosin and Mallory’s trichrome. Microscopic examination showed the wall of the cyst to be composed of collagenous tissue arranged in interlacing fascicles; the wall thickness was variable, and minute areas of calcification were not infrequent. Medium- and small-sized vessels with hyaline walls were seen scattered on the outer surface. The inner surface of the cyst for the most part showed no distinctive lining; however, a few areas were lined by a single layer of cuboidal cells with dark, round basal nuclei. Most importantly, a few papillary tufts were present, composed of typical choroid plexus with a central core of collagenous tissue and small blood vessels covered by cuboidal cells with rounded basal nuclei, typical of choroid plexus epithelium. Structures resembling psammoma bodies were seen in some of the choroid plexus projections (Fig. 4).

Discussion

Interhemispheric cysts are rare, and it is of considerable interest that all reported cases have been associated with complete or partial agenesis of the corpus callosum. In 1968, Loeser and Alvord reported the case of a 1-day-old girl with multiple large interhemispheric cysts lined with ependyma and choroid plexus, in whom total agenesis of the corpus callosum was present. A partial posterior agenesis of the corpus callosum was associated with an in-
FIG. 4. Collagenous wall of the cyst showing a tuft of choroid plexus projecting into the lumen. A few thin-walled blood vessels are present in the external surface. H & E, × 5.

terhemispheric lipoma, and a large interhemispheric cyst in a 1-year-old girl with Fallot's tetralogy. The only case of interhemispheric cyst previously reported in an elderly patient was described by Zingesser, et al., as an arachnoid cyst. Their description of glial tissue composed of astrocytes within the cyst wall is inconsistent with their diagnosis of arachnoid cyst, and indicates a neuroepithelial origin of the cyst.

Total or partial agenesis of the corpus callosum, the large commissure that interconnects the neopallial cortex of the hemispheres, has been associated with several etiological factors. Experimentally, agenesis of the corpus callosum can be produced in mice and rats in early gestation by injecting the mothers with trypan blue, by x-ray irradiation, and by maternal riboflavin-deficient diets. Agenesis of the corpus callosum in humans has been reported in association with X-linked recessive genes, trisomy 13 to 15, occasionally trisomy 18, and the Aicardi syndrome. Vascular, traumatic, toxic, or infectious lesions have also been considered as causative. However, the etiology remains unknown in the majority of the human cases.

Agenesis of the corpus callosum is explained on the basis of a defect (genetic, metabolic, or mechanical) confined to the commissural plate early in development. Loeser and Alvord postulated that agenesis of the corpus callosum is a manifestation of dysgenesis of the telencephalic midline structures. These authors suggested that dilatation of the diencephalic roof plate due to hydrocephalus or intrinsic tissue failure could produce a midline cystic structure extending dorsally in the roof of the third ventricle, and this in turn could interfere with callosal growth and midline development. The dilatation of the roof of the third ventricle could contain choroid plexus, and thus explain the presence of choroid plexus epithelium in the interhemispheric region.

Brun and Probst favored an abnormal cortical plate and, therefore, a reduced number of axons as an important factor in the pathogenesis of the callosal defect. These authors described adult cases with hypoplasia and partial absence of the callosal body in a background of severe perinatal anoxic encephalopathy. They emphasized the fact that although the callosal defect could be severe in encephaloclastic conditions such as anoxic encephalopathy, it was never associated with the formation of an interhemispheric cyst.

Although corpus callosum defects can be produced either early in gestation or in the peri- or postnatal period, interhemispheric cysts are always associated with congenital defects of the corpus callosum that occur early in gestation.

Interhemispheric cysts were diagnosed preoperatively by ventriculography as early as 1937. However, it seems likely that the widespread use of CT scanning will probably result in the discovery of these cysts with greater frequency in the future. Agenesis of the corpus callosum usually produces no gross neurological problems unless it is associated with other abnormalities, one of which is an interhemispheric cyst, which, because of its mass effect, is of clinical significance.

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References


L. C. Solt, J. H. N. Deck, R. S. Baim and K. TerBrugge
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Present address for Dr. Solt: Department of Pathology, Childrens Hospital Medical Center, 300 Longwood Avenue, Boston, Massachusetts 02115.
Present address for Dr. Baim: Department of Radiology, Health Sciences Center, State University of New York at Stonybrook, Stonybrook, New York 11794.
Address reprint requests to: John H. N. Deck, M.D., Department of Pathology, Toronto Western Hospital, 399 Bathurst Street, Toronto, Ontario, Canada M5T 2S8.