Infantile retrocerebellar cyst with immature neural tissue

Case report

JOHN WALSH, M.D., FLOYD H. GILLES, M.D., AND KEASLEY WELCH, M.D.

Departments of Neurosurgery and Pathology (Neuropathology), The Children's Hospital Medical Center, and Department of Surgery and Neurology-Neuropathology, Harvard Medical School, Boston, Massachusetts

A retrocerebellar cyst was excised from a 17-day-old infant with hydrocephalus. The cyst wall contained not only ependyma-like epithelium, choroid plexus tufts, and glial rests, but also prominent areas of immature neural tissue.

KEY WORDS • retrocerebellar cyst • hydrocephalus • posterior cranial fossa • arachnoid cyst

The retrocerebellar "arachnoidal" cyst has been described, with its potential as a surgically treatable cause of infantile hydrocephalus. Some cyst walls surgically removed from infants have contained an ependyma-like epithelium resting on an astroglial membrane or connective tissue, choroid plexus-like fronds, or glial rests.2,3 This case of a neonate with hydrocephalus and a retrocerebellar cyst is reported because, in addition to the above cellular structures, immature neural tissue was found in the wall.

Case Report

This baby girl was the product of an uneventful pregnancy in a primagravida mother. At birth she had a head circumference of 41 cm. Separation of the cranial sutures was observed and later confirmed with radiographs of the skull.

Examination. At 3 days of age the head circumference had decreased to 37.5 cm, a measurement still at the 90th percentile. The anterior fontanel was tense and bulging. When the infant was in the upright position, the fontanel was sunken and there was overriding of the parietal bones along the sagittal suture.

Reexamination at 17 days of age disclosed that the head circumference was again 41 cm, indicating an abnormal rate of growth. Chest circumference and body weight were near the 50th percentile. Cranial sutures were again widely separated and the anterior fontanel was bulging. Poor head control, nystagmus on lateral gaze, greater on the left than the right, and a "setting sun" sign were present. The caudal portions of the posterior fossa transilluminated. The remainder of the neurological examination was normal.

At 17 days ventricular pressure in the right frontal horn was 300 mm H2O, and the ventricular fluid protein was 28 mg/dl. Subsequent air ventriculography demonstrated marked dilatation of the lateral ventricles and
Retrocerebellar cyst with neural tissue

an anterosuperior displacement of the third ventricle. Air was also observed in the upper aqueduct and in the cervical subarachnoid space, but not in the lower aqueduct, the fourth ventricle, the basilar cisterns, or over the cerebral hemispheres. Four-vessel angiography further delineated an avascular retrocerebellar mass.

Operation. At posterior fossa exploration, a large retrocerebellar cyst was present. The cyst appeared to be attached superiorly to the posterior inferior surface of the cerebellar hemisphere and extended caudally to the C-1 level. A portion of the cyst wall was circumferentially excised. The remainder of the cyst was fenestrated into the cervical subarachnoid space. The protein content of the cyst fluid was 1.08 gm/dl.

Postoperative Course. After surgery the intracranial pressure was markedly reduced. It gradually increased over several days and on the ninth day a sudden outpouring of spinal fluid occurred from the midline incision. The infant was given ampicillin, and placed on constant ventricular drainage for 6 days until the incision healed. Subsequently, mild increased pressure was managed with Decadron (dexamethasone) and Diamox (acetazolamide). Angiography at 5 months of age showed no evidence of the retrocerebellar cyst and satisfactory control of the hydrocephalus.

At one year, the head circumference again increased and was greater than the 97th percentile. A ventriculoperitoneal shunt with a medium-pressure Holter valve was placed. When the patient was aged 5 years, the distal portion of the shunt was lengthened. Her head circumference has stabilized at the 97th percentile. She has been without clinical symptoms and, at 6 years of age, her motor, language, and social development was excellent.

Pathological Examination. The membrane was serially sectioned at 10 μ and alternate sections were stained with hematoxylin and eosin or phosphotungstic acid hematoxylin.

The membrane was composed of an outer mesenchymal surface and an inner neural surface. The outer surface of the cyst was comprised of a thin wall of fibrous tissue with small blood vessels. The cellular lining of the inner surface varied from region to region. A large portion consisted of a low cuboidal epithelium resting directly upon the connective tissue of the outer surface. No cilia or blepharoplasts could be discovered in these cells. In some regions this cuboidal epithelium was pushed up into papillary fronds indistinguishable from choroid plexus (Fig. 1). Elsewhere, the cuboidal epithelium gave way to a pseudostratified epithelium resting upon the connective tissue base (Fig. 2). In other regions the epithelium was markedly attenuated. A small amount of hemosiderin was present.

In many regions, both epithelia rested upon a band of glial tissue containing astrocytes and glial fibrils that, in turn, rested upon the connective tissue of the outer surface of the cyst. In many sections, the glial fibrils were oriented obliquely or perpendicularly to the connective tissue.

The most striking feature of this cyst wall was the presence of large patches of immature neural tissue (Fig. 3). The inner surface of these patches did not contain a recognizable cuboidal or pseudostratified epithelium; the outer surface rested upon the connective tissue of the outer surface of the cyst wall. The immature neural tissue consisted of masses of small nuclei densely packed together. No organization of this tissue could be seen. No rosettes were found. The immature neural tissue was sometimes separated from the outer connective tissue by a thin layer of glial tissue. Mitoses were scattered throughout the immature component with no predilection for those cells adja-
cent to the inner surface. The immature neural tissue was almost avascular and contained a small number of karyorrhexic nuclei.

Discussion

The presence of an ependyma-like epithelium on an astroglial or connective tissue base, of choroid plexus-like tissue, and of glial rests in some retrocerebellar cyst walls can readily be accounted for as remnants of a persistent outpouching of the fourth ventricular roof. This normally transient pouch was described by Blake in 1900, in his studies on the embryogenesis of the foramen of Magendie. The histological characteristics of these cystic structures have since been described. In contrast, what is more difficult to account for by this hypothesis is the immature neural tissue noted in this case. Since anatomical orientation of the surgically excised cyst wall specimen was not preserved, it is difficult to determine from what portion of the abnormal fourth ventricular roof the immature neural tissue arose. Perhaps in this infant there was not only failure of regression of Blake's diverticulum, but also abnormal persistence of the fetal neural tissue of the roof of the fourth ventricle. While the mitotic activity indicates a proliferative process, the fact that the patient is alive and well 6 years after incomplete removal suggests that it does not imply neoplastic activity.

Acknowledgment

The authors are indebted to Miss Donna Potash for assistance in manuscript preparation.

References

1. Blake JA: The roof and lateral recesses of the fourth ventricle, considered morphologi-
Retrocerebellar cyst with neural tissue


This study was supported in part by funds provided by the United Cerebral Palsy Research and Educational Foundation (R-224-74), The Children’s Hospital Medical Center Mental Retardation and Human Development Research Program (5 P30 HDO 6276) and NINCDS Grant 5 P01 NS09704.

Address reprint requests to: John Walsh, M.D., Division of Neurosurgery, University of Kentucky Medical Center, Lexington, Kentucky 40506.