Neuroepithelial cysts of the posterior fossa

Case report

SAMUEL F. CIRICILLO, M.D., RICHARD L. DAVIS, M.D., AND CHARLES B. WILSON, M.D.

Departments of Neurological Surgery and Pathology (Neuropathology), School of Medicine, University of California, San Francisco, California

The authors report the case of a patient harboring a posterior fossa neuroepithelial cyst who presented with positional facial weakness and syncope. The patient recovered rapidly after cyst fenestration and placement of an internal cyst-cisternal shunt. The pathogenesis and principles of diagnosis and management of these rare lesions are reviewed.

KEY WORDS: neuroepithelial cyst, posterior fossa, cyst, cerebellopontine angle

Symptomatic intracranial neuroepithelial cysts are rare lesions thought to be of developmental origin that are seen most commonly in the anterior third ventricle. These lesions have been described as colloid, epithelial, ependymal, paraphysial, choroid plexus, and choroidal ependymal cysts. Most patients present with symptoms of increased intracranial pressure caused by obstruction of cerebrospinal fluid (CSF) flow.

While they may become symptomatic at any age, most neuroepithelial cysts are thought to form during embryogenesis from simple folding of neuroepithelium into or out of the primitive ventricular system. Neuroepithelial cysts can occur anywhere along the neuraxis. Most cysts arise from choroid plexus or ependyma along the anterior portion of the third ventricle adjacent to the foramen of Monro, although lesions involving the sella, fourth ventricle, pineal region, sylvian fissure, cervical spine, and conus medullaris have been reported. True posterior fossa neuroepithelial cysts appear to be quite rare; to our knowledge, only five reports of this entity have been published. In most instances, these lesions were treated by surgical excision at craniotomy. We report the case of a patient harboring a neuroepithelial cyst of the posterior fossa who presented with positional facial weakness and syncope. Principles of diagnosis and management of this unusual lesion are discussed.

Case Report

This 33-year-old woman was admitted to our service with a 1-year history of fainting spells precipitated abruptly by bending forward and downward. On the rare occasions that fainting did not occur, she noted a lag in the right eyelid that lasted for several minutes. She also noted mild chronic neck stiffness, headaches, and right eye pain. During the month before admission, she experienced episodes of right thumb numbness and tingling. A tentative diagnosis of basilar migraine was made; a course of propranolol, 100 mg/day, was begun, but her condition did not improve significantly.

Examination. The general physical examination was unremarkable. On neurological examination, the patient had intact cranial nerve, sensory, and motor function, and deep-tendon reflexes and cerebellar function were within normal ranges. A magnetic resonance (MR) image showed the presence of an extra-axial mass with smooth borders centered at the right cerebellopontine angle. The lesion was isointense with CSF on both short- and long-repetition time (TR) images. A mass effect displaced the seventh and eighth nerve complex superiorly and the ninth, 10th, and 11th nerve complex inferiorly (Fig. 1).

Operation. A right suboccipital craniectomy was performed through a vertical retromastoid incision. After incision of the dura and gentle displacement of the cerebellar hemisphere, a gray-yellow cyst was apparent in the interval between the internal auditory canal and the jugular foramen. The seventh and eighth cranial nerves were stretched tightly over the rostral pole of the cyst, and the ninth and 10th nerves were displaced caudally around its lower pole. The cyst wall resembled dense arachnoid except in one area, where it was obvi-
Neuroepithelial cysts of the posterior fossa

FIG. 1. Coronal and axial T₁-weighted magnetic resonance images demonstrating a 3-cm cyst in the region of the right cerebellopontine angle. The cyst displaces the seventh-eighth cranial nerve complex superiorly (single arrow) and the jugular foramen complex inferiorly (arrowhead). The extra-axial fluid collection is isointense with cerebrospinal fluid on both T₁- and T₂-weighted images.

FIG. 2. Follow-up magnetic resonance images 4 months postoperatively showing interval decrease in cyst size (arrow). Brain-stem compression is no longer present.

FIG. 3. Photomicrograph of the cyst wall showing connective-tissue stroma lined by cuboidal epithelium resembling ependyma. H & E, × 100.

Discussion

While neuroepithelial inclusions develop during embryogenesis, neuroepithelial cysts are usually seen in older patients. It is thought that the size increases gradually as secretory and breakdown products from the epithelial lining accumulate within the cyst. Cysts have been shown to contain a proteinaceous fluid comprised of CSF, lipid, and mucicarmine and periodic acid-Schiff reactive material. Hemosiderin-laden macrophages are sometimes seen. Electron microscopic studies have shown the presence of pinocytotic vesicles within the cells lining the cyst, which suggests that they may also expand by transcellular fluid transportation. Because neuroepithelial cysts do not communicate with the subarachnoid space, progressive enlargement cannot be the result of unidirectional CSF flow.

There are several explanations for the origin of neuroepithelial cysts. Some authors have proposed that they are derived from choroid plexus, ependyma, or the paraphysis, a vestigial evagination of third ventricular choroid plexus. Based on a review of the developmental and comparative anatomy of these cysts, Shuangshoti, et al., have shown that they may develop anywhere along the neuraxis lined by primitive ependymal epithelium.
neuroepithelium. During embryological development, aberrant evagination or invagination of the neuroepithelium lining the primitive ventricular system leads to inclusions of ependymal or choroidal rest cells. A review of autopsy specimens confirmed the frequent presence of neuroepithelial inclusions throughout the neuraxis in all age groups; most inclusions are small and asymptomatic.\(^5\) Extra-axial neuroepithelial cysts may also develop from heterotopic neuroglial rests. Several investigators have found heterotopic neuroglial islands in the subarachnoid space that contain choroid plexus and central canals lined by ependymal cells. These heterotopic neuroepithelial structures may be formed by the same processes of invagination and evagination that produce neuroepithelial cysts within the neuraxis.\(^20\) Recently, however, identification by electron microscopy of three distinct cell types and a surface protein coat that have an endodermal origin\(^6,7,10,12,21\) suggests that these cysts may not have a neuroepithelial origin.\(^8,10,11\) Clearly, more work will be necessary to differentiate among the alternative mechanisms proposed for cyst formation.

Most symptomatic neuroepithelial cysts occur in the anterior portion of the third ventricle and frequently obstruct CSP outflow through the foramen of Monro; these comprise a distinct clinical entity known as “colloid” cysts of the third ventricle. Cysts with similar histological appearance, however, have been identified in the sella turcica,\(^18\) sylvian fissure,\(^21\) interhemispheric fissure,\(^19\) quadrigeminal cistern,\(^9\) posterior fossa,\(^1,6,9,14\) and spinal canal.\(^11,13\) In general, there has been a reluctance to use the term “colloid” cyst for these extraventricular cysts.\(^4\)

While asymptomatic neuroepithelial cysts occur frequently in myelencephalic choroid plexus, there are only a few reported cases of histologically verified symptomatic neuroepithelial cysts of the posterior fossa; this low incidence may be related to a failure to recognize the nature of these cysts. Indeed, lesions reported as “arachnoid cysts” lined by cuboidal epithelium may represent neuroepithelial cysts.\(^9\) Parkinson and Childs\(^14\) reported a patient with two fourth ventricular colloid cysts lined by ciliated columnar epithelium; this may have been a case of neurocysticercosis. Harrison\(^9\) described three cases of posterior fossa arachnoid cysts that consisted of glial tissue lined by cuboidal epithelium resembling ependyma. Gardner, et al.,\(^6\) reported three cases of diverticula and one case of a cyst of the foramen of Luschka. The patients with diverticula presented with headache and signs of cerebellar dysfunction; one patient’s symptoms of vertigo were exacerbated by changes in position, as in the patient reported here. The patient with a true cyst of the foramen of Luschka presented with headache, signs of brain-stem compression, and deficits of the sixth to 10th cranial nerves. Microscopic analysis of biopsy material from each patient showed a thin-walled cyst composed of a single row of low cuboidal cells resembling ependyma and an outer surface of flattened arachnoidal cells. The authors suggested that these results represent mild forms of embryonal atresia of the fourth ventricle caused by the relative impermeability of the rhombic roof in the 2nd month of fetal development. Separation and stretch of the embryonal double layer across the foramen of Luschka produces a cyst and diverticulum, respectively.\(^8\)

We have seen two other neuroepithelial cysts in the posterior fossa.\(^4\) One patient presented with a cerebellopontine angle mass causing hearing loss and facial numbness; the other had a pontine mass adjacent to the fourth ventricle that caused vertigo, ataxia, and ophthalmoplegia. Microscopic evaluation of material obtained at operation showed a cyst wall composed of low cuboidal epithelium that was lined by an intact basement membrane and connective tissue.

In early reports, posterior fossa developmental cysts were confirmed by means of ventriculography and pneumoencephalography, which showed dilatation of the lateral and third ventricles with evidence of obstruction of CSF flow from the fourth ventricle. On computerized tomography (CT), neuroepithelial cysts resemble other developmental cysts and appear as noncalcified, low-density extra-axial masses that do not enhance with contrast medium. Borders are smooth and clearly defined. If CSF pathways are obstructed, ventricles are enlarged. It is not possible on CT scans to differentiate among the many cystic lesions of the posterior fossa, including arachnoid cysts and cysts associated with isodense tumors such as ganglioglioma and cerebellar hemangioblastoma. Radioisotopic or CT-metrizamide cisternograms or ventriculograms may be necessary to differentiate neuroepithelial cysts from abnormal collections of CSF such as communicating arachnoid cysts, porencephalic cysts, an enlarged cisterna magna, or the posterior fossa cyst associated with a Dandy-Walker malformation.

For these lesions, MR imaging is superior to conventional CT and is the radiographic study of choice in patients suspected of harboring a posterior fossa cyst. Coronal and sagittal views allow better visualization of the underlying pathology and its anatomical relationship to surrounding neural structures. In the case presented here, displacement of the seventh-eighth cranial nerve complex and jugular foramen complex were clearly seen on MR images. Long- and short-TR sequences showed cyst fluid with signal characteristics equal to CSF. Results obtained with steady-state free-processing imaging techniques were consistent with moving fluid. These newer MR techniques designed to assess pulsatile CSF motion cannot distinguish bulk flow of CSF from arterial systolic pulsations transmitted from the surrounding brain tissue.\(^3\) Thus, CT-metrizamide cisternography or ventriculography remains the only reliable technique that can confirm the presence or absence of communication with the subarachnoid space.

Craniotomy is the surgical treatment of choice for neuroepithelial cysts of the posterior fossa and allows
Neuroepithelial cysts of the posterior fossa

direct inspection of the cyst wall, confirmation of the diagnosis by biopsy, and fenestration of the cyst. In the patient reported here, it was not possible to differentiate a neuroepithelial cyst from an arachnoid cyst based on the preoperative evaluation. Intraoperative inspection of the cyst wall and sampling of the gray-colored plaques containing neuroepithelium allowed the correct diagnosis to be made. Wide fenestration of the cyst wall into the ventricular system or subarachnoid space allows free drainage of the proteinaceous fluid. Although recurrence of a neuroepithelial cyst after fenestration alone is rare, it has been reported. An internal shunt with a Pudenz ventricular catheter was placed between the lateral cistern rostrally and the cisterna magna caudally to ensure communication of the cyst contents with the subarachnoid space. This surgical approach was successful, and the patient’s neurological condition improved rapidly.

References

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Address reprint requests to: Samuel F. Ciricillo, M.D., c/o The Editorial Office, 1360 Ninth Avenue, Suite 210, San Francisco, California 94122.