Unusual CT-dense posterior fossa epidermoid cyst

Case report

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A posterior fossa epidermoid cyst with high computerized tomographic attenuation is reported. The pathological and radiological features of this unusual case are presented.

KEY WORDS • epidermoid tumor • pearly tumor • computerized tomography • posterior fossa tumor

Intracranial epidermoid cysts are variously estimated to comprise from 0.2% to 1% of all intracranial tumors.10 Computerized tomographic (CT) studies of these lesions have indicated their characteristic low attenuation values.2-4,7,8 We report a patient with a large, sharply demarcated posterior fossa epidermoid cyst that appeared homogeneously dense (66 to 99 Hounsfield units) on preoperative CT scanning. Review of the English language literature revealed only one other publication reporting such dense epidermoid tumors.1 Our case emphasizes that these lesions, although rare, should be included in the preoperative differential diagnosis of high-attenuation posterior fossa masses.

Case Report

This 16-year-old Chinese girl presented to the University of Malaya Hospital with a 5-month history of bifrontal headaches. Four months before admission, the patient developed severe vertigo on awakening in the morning; this was often associated with vomiting. The headaches, which precluded reading, persisted throughout the day, and eventually prompted hospitalization. Past medical history included chronic otitis media throughout childhood, with frequent episodes of purulent ear drainage. There was no history of meningitis.

Examination. The patient was alert and oriented. Her neck was moderately stiff. Both tympanic membranes were perforated, but no drainage was seen. There was bilateral papilledema. Her sensorimotor examination was intact, but she was unable to tandem walk and had bilateral upper extremity dysmetria. No pathological reflexes were elicited.

A CT scan (Fig. 1) showed a homogeneously dense midline lesion in the posterior fossa that did not enhance with contrast infusion. The fourth ventricle was not visualized, but the third and lateral ventricles were grossly enlarged. Inferiorly, the mass extended laterally to the right petrous bone. A right vertebral angiogram showed an avascular mass lesion displacing the posterior inferior cerebellar artery inferiorly and compressing the basal artery against the clivus.

Operation. A suboccipital craniectomy was performed. The cerebellar tonsils were found to be displaced into the cervical spinal canal, and when the widened cerebellar vermis was split, a soft dark cyst was encountered. This extended from the incisura to the foramen magnum and contained viscous green-brown fluid. Laterally, the cyst extended to the right petrous bone, where it was adherent to the dura. There was no detectable abnormality of the petrous bone itself. The fourth ventricle was displaced to the left but was not invaded by the tumor.
CT-dense epidermoid cyst

Fig. 1. Noncontrast-enhanced computerized tomographic scan demonstrating a high-density (99 Hounsfield units) midline cerebellar lesion. There is dilatation of the anterior recesses of the third ventricle and temporal horns. The fourth ventricle is not seen.

Microscopic examination of the cyst wall showed loose connective tissue lined by stratified squamous epithelium. The contents of the cyst were made up of eosinophilic acellular colloid-like material, with collections of hemosiderin-laden macrophages. Giant cells and lamellar keratotic material were also noted. Cultures of this material for bacteria, fungi, and mycobacteria were sterile.

Postoperative Course. The patient had transient worsening of her right upper extremity ataxia, and a right fourth cranial nerve paresis. Her hydrocephalus resolved without ventricular shunting. Headaches and vertigo both improved postoperatively.

Discussion

Intracranial epidermoid cysts are thought to result from developmental abnormalities in which epithelium is misplaced during embryogenesis,5,11 but their exact pathogenesis remains obscure. Since many of these tumors are both extra-axial and not midline, it seems unlikely that they result from simple entrapment of epithelium during neurulation. Lepoire and Pertuiset6 presented the hypothesis that epithelial inclusions are carried intracranially by the developing cerebral vasculature and that these rests are responsible for subsequent cyst formation.

On CT scanning, epidermoid cysts usually appear as low-density (−8 to +32 Hounsfield units) space-occupying lesions, although one previous report by Braun, et al.,1 documents the occurrence of dense epidermoid cysts in three patients (80 to 120 Hounsfield units). The reason for the high CT attenuation values in these patients is unknown. In our case, the gross appearance of the cyst fluid, as well as the presence of hemosiderin-laden macrophages in the cyst wall, suggested the possibility of prior bleeding into the cyst, but such an occurrence must be distinctly uncommon given the avascular nature of these lesions. The usual cyst content in epidermoid tumors is a white, waxy material which is desquamated keratin from the epithelial cyst lining. However, viscous brown cyst fluid similar to that found in our case has previously been described within intracranial epidermoid tumors,10 and also within epidermoid cysts in other locations, such as the spleen.9 It is unfortunate that no cyst fluid has been chemically analyzed in reported cases of dense epidermoid tumors.

Curiously, all reported cases of epidermoid cysts with high CT attenuation have occurred in the posterior fossa of females. As noted by Braun, et al.,1 this concurrence may simply reflect the higher incidence of these lesions in females, and their increased frequency in the posterior fossa.

Other posterior fossa lesions with a CT appearance similar to that in our patient include meningiomas, acoustic neuromas, and choroid plexus papillomas. Generally, these lesions have lower attenuation values on noncontrast scanning and, unlike our case, they usually exhibit marked contrast enhancement. Metastases, especially from colonic carcinoma, may be homogeneously dense and not display contrast enhancement. We report this patient because epidermoid cysts, lesions that are benign and curable by surgical excision, should also be included in the differential diagnosis of posterior fossa lesions with high CT attenuation values.

References


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