Neurenteric cyst with xanthomatous changes in the prepontine area: unusual radiological findings

Case report

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Preoperative diagnosis of neurenteric cysts can be difficult because the imaging findings of a neurenteric cyst may be similar to those of an arachnoid cyst. The authors report a case of a neurenteric cyst with xanthomatous changes in the prepontine area. This 4-year-old girl was admitted to their institution with intermittent neck pain and vomiting. Computed tomography showed a hypodense mass in the prepontine area. Magnetic resonance imaging showed a cystic lesion measuring ~ 4 × 3 cm. The brainstem was displaced posteriorly, and the cisterns in both cerebellopontine angles were widened. The signal intensity of the cyst was similar to that of cerebrospinal fluid. Adjacent to the basilar artery there was a solid component of the mass that enhanced after administration of Gd. Intraoperatively, the authors found a cystic mass containing clear fluid with a yellowish solid nodule. On the basis of histopathological findings, the lesion was diagnosed as a neurenteric cyst with xanthomatous changes. (DOI: 10.3171/PED.2008.2.11.351)

Key Words • arachnoid cyst • neurenteric cyst • radiology • xanthoma

The term neurenteric cyst is generally used to describe a type of epithelium-lined cyst of uncertain histogenesis, but probably of endodermal or neuroepithelial derivation.¹³¹² Neurenteric cysts usually develop in the subdural space on the anterior spinal cord and rarely develop intracranially. When they do develop intracranially, they are usually located in the posterior fossa.²⁸¹⁰ The incidence of neurenteric cysts is higher in adults than in children.² When neurenteric cysts are associated with clinical signs or symptoms, the most common are related to increased intracranial pressure, such as headache, nausea and vomiting. Lower cranial nerve palsy and hydrocephalus are occasionally reported in patients with neurenteric cysts.

We report a case of a neurenteric cyst with xanthomatous changes and describe the neuroimaging and pathological findings in our patient as well as discussing the pathogenesis, differential diagnosis, and treatment of this lesion.

Case Report

History and Presentation. This 4-year-old girl was admitted to our institution for the evaluation of an intra-

cranial cystic mass diagnosed by CT performed at a local hospital because of frequent vomiting. She suffered from intermittent neck pain, morning headaches, and vomiting with a progressive course. Her medical history was otherwise unremarkable.

Examination. Neurological examination performed at admission showed no evidence of mental status impairment, gait difficulties, sensory or motor deficits, or cranial nerve palsy. The findings of routine laboratory tests were within the normal limits. Computed tomography showed a hypodense mass in the prepontine area. Magnetic resonance imaging revealed a cystic lesion measuring ~ 4 × 3 cm located in the prepontine area. The brainstem was displaced posteriorly, and the cisterns in the prepontine area were widened. The signal intensity of the cyst was exactly comparable to that of cerebrospinal fluid. The lesion was diagnosed as a neurenteric cyst with xanthomatous changes.
Operation. Right lateral suboccipital craniotomy was performed. After the lateral cerebellomedullary cistern was opened, CSF ran over. We found a cystic mass containing clear fluid like CSF in front of the prepontine and premedullary cisterns. We dissected and traced the cyst and found a yellowish solid nodule (Fig. 2A and B). The cranial nerves VIII and XI were adherent to the cyst and carefully dissected. The cyst was completely removed.

Pathological Findings. Light microscopy revealed a strong mucinous component within goblet cells along the epithelium (Fig. 3A). The immunoreactive markers for cytokeratin, epithelial membrane antigen (Fig. 3B), and carcinoembryonic antigen were positive, but staining for S100 protein was negative. Periodic-acid Schiff staining, which is mainly used to stain structures containing a high proportion of carbohydrate macromolecules such as mucus, was positive (Fig. 3C). The yellowish solid nodule was composed of foamy histiocytes and chronic inflammatory cells on light microscopy (Fig. 3D). On the basis of these findings, this cystic lesion was diagnosed as a neurenteric cyst with xanthomatous changes.

Postoperative Course. Postoperative MR imaging scans revealed complete tumor removal, and there were no postoperative complications. The patient’s preoperative symptoms completely disappeared after tumor removal.

Discussion

Neurenteric cysts occur in patients of all ages, but the average age at presentation is 34 years. The age distribution has 2 distinct peaks: a large peak occurring in the third and fourth decades and a smaller peak observed in the first decade. In 61% of the reported cases the patients were male.

An epithelial cyst of endodermal origin is thought to develop from ectopic remnants of the endoderm. This hypothesis is supported by various theories. Most authors favor the theory of partial regression of the neurenteric canal, which leaves endodermal cells in and around the neural tube. Shin et al., citing Brooks et al., Lonjon et al., and Simon et al., state: ”Neurenteric cysts arise as a result of persistence of the neurenteric canal, which is the temporary connection between the amniotic and yolk sacs during the third week of embryogenesis. Persistent endodermal adhesions or a persistent adhesion between the notochord and endoderm may produce a notochordal dysgenesis, such as diastematomyelia, and a neurenteric cyst may result.” This explains how endodermal cysts can be distributed from the posterior fossa to the sacral area.

The most difficult aspect of diagnosis of a neurenteric cyst is differentiation from an arachnoid cyst. Most arach-
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Variable MR imaging signal characteristics of cysts have been identified. Cysts can be filled with mucus or blood and exhibit iso-, hyper-, or mixed intensity relative to CSF on T1- and T2-weighted images. This difference from CSF in signal intensity on ≥ 1 pulse sequences allows for distinction between CSF and arachnoid cysts. This difference in signal intensity between arachnoid cysts and CSF is the most important clue for distinguishing between the 2. In the case presented in this paper, the finding of the same signal intensity in the background of CSF on T1- and T2-weighted images made the preoperative differential diagnosis of an arachnoid cyst difficult. Moreover, the small-sized lesion with solid enhancement suggested another pathology different from arachnoid cyst. The solid enhancing lesion was pathologically confirmed to have xanthomatous changes.

Xanthomatous changes are a nonspecific chronic cellular reaction characterized by the presence of foam or xanthoma cells. Xanthogranulomatous reactions are known to occur in benign, noninflammatory cystic or cyst-like lesions, such as a neurenteric cyst, colloid cyst, epidermoid cyst, cholesterol granuloma, choroid plexus cyst, and even a renal or pancreatic cyst, but these are very rare. Shin et al., reported on a case similar to ours, although theirs involved a middle-aged woman rather than a child. The lesion showed iso-intensity on enhanced T1-weighted images and low signal intensity on T2-weighted images. The latter finding, the authors observed, reflects the high cellularity that results from the presence of compact foamy histiocytes and chronic inflammatory cells. They also noted that the solid nodule showing the xanthogranulomatous reaction adhered to the adjacent normal tissue in the surgical field, making complete dissection difficult. In our case, the cyst also adhered to the cranial nerves VIII and XI, and the tumor was carefully dissected.

Neurenteric cysts can be confused with epidermoid cysts. The presence of lobulated contours on each tumor would be helpful for distinguishing between endodermal and epidermoid cysts, but it may not be possible to distinguish between the 2 types using imaging studies alone in cases of smoothly contoured epidermoid cysts.
Preoperative diagnosis is sometimes difficult as the imaging findings of a neurenteric cyst may be similar to those of an arachnoid cyst. The treatment of choice for a neurenteric cyst is surgical removal of the cyst wall rather than fenestration because the cell lining can secrete fluid, which is not the case in arachnoid cysts. Recurrence of neurenteric cysts is rarely reported, and the clinical course is usually benign, even after subtotal removal of the lesion. Accurate and prompt diagnosis on MR images and successful operation preserving the cranial nerve might be important for treating neurenteric cysts in the prepontine area.

Disclaimer

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References


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