Extradural neurenteric cyst of the cerebellopontine angle

Case report

TOMOHIRO INOUE, M.D., NOBUTAKA KAWAHARA, M.D., PH.D., JUNJI SHIBAHARA, M.D., PH.D., TOMOHIKO MASUMOTO, M.D., KENICHI USAMI, M.D., AND TAKAAKI KIRINO, M.D., PH.D.

Departments of Neurosurgery, Pathology, and Radiology, Faculty of Medicine, The University of Tokyo, Japan

Neurenteric cyst is a developmental malformation found mainly in the spinal canal. The authors report on a 47-year-old man with a neurenteric cyst of the cerebellopontine angle (CPA) who presented with progressive hearing disturbance and facial palsy. The tumor was located extradurally with marked destruction of the petrous bone around the internal auditory canal and demonstrated irregular and heterogeneous high-intensity signals on T1- and T2-weighted MR images, which is atypical for neurenteric cysts. The pathological findings in samples obtained after resection disclosed a single epithelial layer (a feature of neurenteric cyst), which was accompanied by marked xanthogranulomatous changes. Although several neurenteric cysts have been reported in the CPA, extradural lesions with unusual imaging features and marked bone destruction have not been reported previously. This benign developmental lesion should be considered, although it is extremely rare, in patients harboring an extradural temporal bone tumor around the CPA.

Key Words • neurenteric cyst • extradural tumor • cerebellopontine angle

Abbreviations used in this paper: CPA = cerebellopontine angle; CT = computerized tomography; MR = magnetic resonance.
granulomatous reaction and hemosiderin deposition, which gave the cyst its solid appearance.

**Discussion**

Neurenteric cysts are rare developmental lesions derived from endodermal inclusions occurring mainly in the spinal canal with or without associated spinal abnormalities. Intracranial neurenteric cysts are even more rare; fewer than 10 cases have been reported. The majority of such lesions are found in the CPA and are located in intradural and extramedullary space, as are spinal lesions. In this regard, an intracranial extradural neurenteric cyst in the CPA, as described here, seems extremely rare, although an intraosseous neurenteric cyst within the clivus has been reported. Developmentally, neurenteric cysts are presumed to derive from isolated inclusion of the endodermal tissue after incomplete separation of the notochord and foregut during embryogenesis. According to this hypothesis, these lesions could occur anywhere between the gut and spinal axis and may even be connected with the prevertebral space through a dysraphic spinal column. Indeed, one such case featuring a cervical spine neurenteric cyst with mediastinal extension has recently been reported, thus substantiating this theory. The intracranial extradural location in our case would be explained similarly, although why these lesions preferentially involve the CPA remains unknown.

The diagnostic information is mainly obtained by using MR imaging. Neurenteric cysts are usually delineated as nonenhancing homogeneous cysts with isointense to slightly high intensity signals on T1-weighted imaging and high-intensity signals on T2 images, although the T1-weighted signal intensity may differ depending on the protein concentration or hemorrhage within the cyst. In view of these characteristic features, MR imaging findings in our case were atypical because the lesion manifested as a heterogeneous and irregular mass with high-intensity signals in both T1- and T2-weighted images, reflecting multicystic components with marked stromal xanthogranulomatous changes. In addition, associated petrous bone destruction, as demonstrated on a CT scan, has not been reported previously in CPA neurenteric cysts. Because of this atypical imaging characteristic not previously documented, it would be extremely difficult to make a correct diagnosis preoperatively.

Although a variety of tumors may arise in the CPA, a nonenhancing extradural lesion in the CPA with destruction of the petrous apparatus should be differentiated in particular from epidermoid and cholesterol granuloma. Among these tumor types, only cholesterol granuloma is hyperintense on T1-weighted imaging, although they are all hyperintense on T2-weighted imaging. In addition, all of these tumors are depicted as rather homogeneous masses in these conditions. In that sense, a dermoid tumor has a similar MR imaging appearance in our case, although it predominantly affects the midline structures. For the differential diagnosis of these lesions, diffusion-weighted imaging offers valuable information because epidermoid as well as dermoid lesions display high-intensity signals. In our case, the diffusion-weighted image depicted a hypointense mass and may serve as a diagnostic tool of choice, as reported elsewhere.

The final diagnosis should be made based on the histopathological findings, which have been well characterized. Microscopically, neurenteric cysts have cuboidal columnar epithelium with or without cilia and mucus globules. Well-formed intestinal or bronchial epithelium as well as squamous metaplasia are occasionally observed. Immunohistochemical studies have been used to identify cellular origin; in these tests neurenteric cysts are positive for carcinoembryonic antigen, a useful marker for endodermal cells in the embryonic gastrointestinal tract, for cytokeratins (AE1 and AE3), and for epithelial membrane antigen. They are usually negative for S100 protein and glial fibrillar acidic protein, two markers for glial–schwannian and astrocytic–oligodendrocytic differentiation, respectively. The findings in our case were consistent with these typical features except...
for focal positive immunostaining for S100 protein, which has also been documented previously.3

**Conclusions**

We report on an extradural neurenteric cyst of the CPA that was destructive of petrous apparatus and had unusual imaging characteristics that have not been described previously in this location. Histologically, the tumor was accompanied by strong xanthogranulomatous stromal change that might have given rise to the atypical imaging features. We hope that this case presentation will contribute to awareness of this rare developmental tumor with its atypical imaging characteristics when neurosurgeons are making a differential diagnosis of CPA lesions and further understanding of the tumor’s pathogenesis.

**References**


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Address reprint requests to: Nobutaka Kawahara, M.D., Ph.D., Department of Neurosurgery, Faculty of Medicine, the University of Tokyo, 7–3–1 Hongo, Bunkyo-ku Tokyo, Japan, 113–8655. email: kawahara-tky@umin.ac.jp.