Chordoma of Petrosal Mastoid-region: a case report and review of the literature.

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Abstract
BACKGROUND
Chordoma is a rare low-grade malignant tumor originating from embryonic notochordal tissue mainly occurring in the axial bone, mostly in the sphenoccipital junction and sacrococcyx, that accounts for approximately 1% of all malignant bone tumors and 0.1% to 0.2% of intracranial tumors. Chordoma in the petrous mastoid region is rare.

CASE SUMMARY
We describe a 36-year-old male patient with chordoma in the left petrous mastoid region. The main clinical manifestations were pain and discomfort, which lasted for 2 years. Magnetic resonance imaging (MRI) showed a lobulated mass in the left petrous mastoid with an unclear boundary and obvious enhancement. The tumor was completely removed after surgical treatment, and a histological examination confirmed that the tumor was a chordoma. During 5 years of follow-up, no clinical or radiological evidence of recurrence or metastasis was found.

CONCLUSION
Although chordoma in the petrosal mastoid region is rare and may be misdiagnosed as a jugular glomus tumor or middle ear cancer, the disease should be included in the
differential diagnoses of petrosal mastoid-region tumors. In addition, an imaging examination can sufficiently show the extent of the focus and surrounding involvement, which can provide useful information for clinical treatment.

**Key Words:** chordoma, petrous mastoid, rare disease, bone tumor; magnetic resonance imaging


**Core Tip:** Chordoma is a relatively rare disease, especially in the petrous mastoid region. Its imaging findings are rarely reported, and an understanding of its MRI findings is lacking. However, in the differential diagnosis of petrous mastoid tumors, chordoma should be considered, especially when lobulated masses are found.

**INTRODUCTION**

Chordoma is a rare low-grade malignant tumor originating from embryonic notochordal tissue with an incidence of less than 0.1/100,000 individuals per year [1]. Chordoma is locally destructive and easily invades important structures, such as bones, nerves, and large blood vessels. Chordoma is likely to relapse after resection, but metastasis is rare. The pathogenesis of chordoma is unclear, and the disease is speculated to originate from residual cells of the notochord that developed during fetal development. The tumor usually occurs in the axial bone, especially at both ends of the axial bone. Most chordomas in the head and neck occur in the sphenooroccipital junction, accounting for almost 35% of all chordomas [2]. A few cases occurring in the nasopharynx, paranasal sinuses, oropharynx, and jugular foramen have been reported [2, 3], and approximately 0.2% of the cases reported in the literature occurred in the jugular foramen region [4] but not the petrosal mastoid region. Here, we report a case of
chordoma in the left petrosal mastoid region with intracranial expansive growth resulting in cerebellar compression and deformation. The main clinical manifestation was a 2-year history of temporal pain and discomfort.

CASE PRESENTATION

Chief complaints
A 36-year-old man visited our hospital in December 2014 because of left temporal pain.

History of present illness
The patient had a history of hepatitis B infection for more than 30 years, and his liver function was normal.

History of past illness
In 2012, no obvious cause for his left temporal pain and left-sided headache could be identified, and the patient had no symptoms of hearing loss, dizziness, nausea, or vomiting and no obvious redness, swelling, heat, pain, or structural aberrations in the adjacent skin.

Personal and family history
No specific genetic or family history of disease was identified.

Physical examination
Neurologic exam revealed House Brackmann (HB) Grade II, gait disturbance, tinnitus. The patient denied hearing loss, diplopia.

Laboratory examinations
No abnormality was found in the patient's laboratory examination.

Imaging examinations
A magnetic resonance imaging (MRI) examination of the brain showed a lobulated mass in the left petrous mastoid region with a maximum interface of approximately 3.2 cm×3.9 cm×5.0 cm, surrounding bone absorption and destruction, uneven signal intensity on T2-weighted images (Figure 1a), isointensity and hypointensity on T1-weighted images (Figure 1b), hyperintensity on fluid-attenuated inversion recovery (FLAIR) (Figure 1c), small cystic degeneration in the interior of the tumor (Figure 1a-b), and obvious enhancement of the solid components of the lesion on the contrast-enhanced scan (Figure 1d). No sign of involvement of the left jugular vein and left sigmoid sinus was observed. Left cerebellar compression and displacement were evident, but no brain parenchymal edema was noted.

FINAL DIAGNOSIS
The gross specimen of the tumor was grayish-red and tough with a rich blood supply and had an unclear boundary. Microscopically, the tumor consisted of a dual cell population embedded in an abundant myxoid background. The cells were epithelioid polygonal in appearance with clear or eosinophilic cytoplasm, and a few mitotic cells were observed (Figure 2a). The immunohistochemical staining showed that vimentin, soluble protein-100 (S-100), epithelial membrane antigen (EMA) (Figure 2b), and CK19 were positive, and the cytokeratin (CK) staining was negative. The Ki67 antigen (Ki-67) fraction was approximately 1%. Ultimately, a diagnosis of chordoma was made.

TREATMENT
The patient underwent microscopic tumor resection through the upper and posterior windows of the left ear. During the operation, the tumor was found to be located in the epidura; the petrous part of the temporal bone and the surface of the occipital bone were eroded by the tumor; the left transverse sinus was involved; and the lower part of the tumor surrounded the nerves and vessels.

OUTCOME AND FOLLOW-UP
The patient recovered well after surgery, no specific discomfort was mentioned, and he was discharged 1 wk after the operation. As chordoma is a low-grade malignant tumor, the patient was reexamined once a year after the operation, and no obvious abnormality was found after a follow-up for 5 years (Figure 1e-f).

**DISCUSSION**

Chordoma is a rare, slow-growing, locally invasive malignant tumor originating from primitive notochordal tissues that develop longitudinally along the axis. Chordomas account for 3% of all primary bone tumors \[^3\]. Approximately 50% of chordomas occur in the sacrococcyx, 30% of chordomas occur in the sphenoccipital region, and 20% of chordomas occur in the mobile spine \[^6\]. Chordoma can occur at any age, and the most common age at diagnosis is between 20 and 40 years. The gross specimen of the tumor showed lobulated and expansive growth with mucus, cystic necrosis, a small bleeding focus, calcification, ossification, and cartilage islands, which easily invaded the surrounding bone and caused extensive bone destruction. Histologically, the tumor was composed of droplet cells; the tumor tissue was divided into lobules by connective tissue; and the tumor cells were arranged into small clusters, flakes, strips, and acini with common degeneration and myxoid degeneration in the stroma. Histologically, chordoma appearance can be divided into the following three subtypes: classical, chondroid and dedifferentiated; classical chordoma is the most common subtype. In addition, chondroid tumor growth is the slowest, and dedifferentiated tumor growth is the fastest.

Typical chordoma CT findings mainly include irregular low-density masses with well-defined boundaries, expansive growth, absorption and destruction of surrounding bone, irregular calcification inside the lesion, and obvious enhancement \[^7\]. CT scans can accurately show bone and internal calcification but are insufficient for soft tissue analyses, and an MRI examination is needed to facilitate further diagnosis.

The MRI findings of chordomas are diverse \[^8\]. The typical MRI findings of chordomas in the clival region show obvious hyperintensity and a "beehive-like" appearance on
T2WI, reflecting the histological characteristics of tumor tissue mainly composed of mucous stroma and droplet tumor cells secreting mucus. In this case, the presence of scattered strips and flakes with a low signal intensity suggested that the tumor was related to bone destruction, calcification, or a fibrous septum, and the high signal area in the tumor was separated and appeared "beehive-like" [9]. The enhancement of the chordoma on imaging was mainly inhomogeneous and obvious, while the dynamic enhancement scan showed continuous and slow enhancement, which is mainly caused by the adsorption of Gd-DTPA molecules to the mucin in the cell or cell interstitium, and the typical enhancement of chordoma shows a "honeycomb sign" [9]. In this case, the MRI findings differed from those of typical chordomas in the clival region. The T2WI results showed that the lesion was mainly slightly hyperintense with only narrow areas of hyperintensity. Given the pathological analysis results, the low signal intensity on T2WI was likely due to the relatively dense arrangement of tumor cells and the relative scarcity of mucous cells. On the contrast-enhanced scan, the focus was not uniformly enhanced, the small cystic area was not enhanced, and the typical "honeycomb sign" was not observed.

Chordoma in the petrous mastoid region should be differentiated from the following tumors. (1) Jugular glomus tumors also show osteolytic destruction with flaky calcification, marginal bone infiltration, and a "salt and pepper sign" on T2WI, while chordomas show a high signal intensity, small cystic degeneration and obvious enhancement on T2WI. (2) Most patients with middle ear cancer have a history of chronic suppurative otitis media, and symptoms, such as earache, tinnitus, and ear canal bleeding, may occur. The main manifestations on MRI are a soft tissue mass with the middle ear as the center, an unclear boundary, surrounding bone absorption and destruction, and enhanced inhomogeneous enhancement; in contrast, although chordomas show expansion and growth, the boundary is relatively clear, and the enhancement is obvious. (3) Chondrosarcoma and chordoma have overlapping ages of onset, both feature calcification and surrounding bone infiltration in the mass, and T2WI can show a significantly high signal intensity; thus, distinguishing between these
two diseases relying solely on imaging features is difficult, and the final diagnosis depends on histopathology. (4) Solitary fibroma tumor (SFT) is a borderline tumor, and most SFTs are considered benign and more common in adults than children. MRI mainly shows well-defined masses mostly in the supratentorial space that are primarily attached to the dura mater with an abundant blood supply and obvious enhancement; a specific sign of intracranial solitary fibroma is a hypointense area (collagen fibers) on T2WI. In most cases, empty blood vessels can be observed, and most adjacent bones show compressive changes, while chordoma has hyperintensity on T2WI, and the surrounding bone is absorbed and destroyed.

CONCLUSION

Chordoma occurring in the petrous mastoid region is relatively rare, its imaging findings are rarely reported, and an understanding of its MRI findings is lacking. However, in the differential diagnosis of petrous mastoid tumors, chordoma should be considered, especially when lobulated masses are found. The T2WI results are mainly characterized by a high signal intensity, obvious enhancement, and surrounding bone absorption and destruction. Therefore, an imaging examination can sufficiently show the extent of the focus and surrounding involvement, which can provide more useful information for clinical treatment.
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