Intraosseous Schwannoma of the Petrous Apex

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Abstract

Background and Importance Intraosseous schwannoma is a relatively rare clinical entity that typically arises in vertebral and mandibular bone. Intraosseous schwannoma located entirely within the petrous bone is exceedingly rare, and only two cases have been reported to date.

Clinical Presentation A 47-year-old Asian man was referred to our hospital with a chief complaint of double vision. Neurologic examination revealed left abducens nerve palsy. Radiologic imaging showed a 35-mm osteolytic expansive lesion located in the left petrous apex. We made a preoperative diagnosis of chondrosarcoma and performed surgical resection. Surgery was performed via a left subtemporal epidural approach with anterior petrosectomy. The histopathologic diagnosis of the tumor was schwannoma. Schwannoma arising from cranial nerves was excluded from intraoperative findings in conjunction with the results for cranial nerves, and intraosseous schwannoma was diagnosed. Postoperative course was uneventful, and abducens nerve palsy resolved immediately after surgery.

Conclusion The differential diagnosis of intraosseous schwannoma should be considered for an osteolytic mass lesion within the petrous apex. Subcapsular tumor removal was considered ideal in terms of preservation of the cranial nerves and vessels around the tumor.

Keywords
- schwannoma
- intraosseous
- petrous apex
- neurinoma

Background

Intraosseous schwannoma is a relatively rare benign neoplasm that arises from Schwann cells of the nerve sheath and involves the substance of bone.[1,2] These tumors usually show a predilection for the mandible and the vertebrae, particularly in the head and neck region.[1,2] We provide just the third report of intraosseous schwannoma of the petrous apex in which abducens nerve palsy developed.[3,4] We also review the two previous reports to identify the common clinical features.

Clinical Presentation

Onset

A 47-year-old Asian man was referred to our hospital with a 1-month history of double vision. Neurologic examination at the time of initial presentation revealed left abducens nerve palsy.

Preoperative Radiologic Findings

Computed tomography (CT) showed an osteolytic expansive lesion, 35 mm in diameter, in the left petrous apex

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adjacent to the posterior wall of the carotid canal, mastoid air cell, and tympanic cavity (►**Fig. 1A**). The lesion showed no apparent calcification and inhomogeneous enhancement after intravenous administration of contrast media (►**Fig. 1B**). High-resolution CT of the left temporal bone revealed a scalloped tumor margin with a thin intact rim (►**Fig. 1C, D**).

Magnetic resonance imaging (MRI) showed the mass lesion demonstrating low signal intensity on T1-weighted imaging (T1-WI), and slightly high signal intensity on T2-weighted imaging (T2-WI) in a patchy fashion (►**Fig. 2A**). Diffusion-weighted imaging (DWI) showed a slight high-intensity lesion (►**Fig. 2B**). According to magnetic resonance angiography, the left internal carotid artery did not seem to have been invaded by tumor. Contrast-enhanced MRI showed heterogeneous enhancement commensurate with the dappled T2-WI intensity (►**Fig. 2C**).

Fast imaging using steady-state acquisition imaging showed that the facial and auditory nerves were located dorsally and isolated from the lesion at the cerebellopontine angle (►**Fig. 2D**). In addition, the trigeminal nerve was seen located cranial to the tumor.

Based on these radiologic findings, a preoperative diagnosis of chondrosarcoma was made, and a surgical treatment of the lesion was planned.

Operation

Tumor removal was performed via a left subtemporal epidural approach with anterior petrosectomy. We used a surgical navigation system (Medtronic, Tokyo, Japan) and monitored auditory brainstem response (ABR), facial electromyography (EMG), and electrooculography (EOG).

Briefly, the temporal dura was detached from the middle cranial base, and the middle meningeal artery passing the foramen ovale was identified, coagulated, and cut. The greater and lesser superficial petrosal nerve was then identified and preserved with the periosteal dura. Dissection was continued toward the petrous apex. At this point, the internal carotid artery was exposed just beneath the greater superficial petrosal nerve.

After anterior petrosectomy was completed, a yellowish tumor component covered by white capsule was identified (►**Fig. 3A**). The superior and anterior walls of the internal auditory meatus were also opened, and subcapsular removal

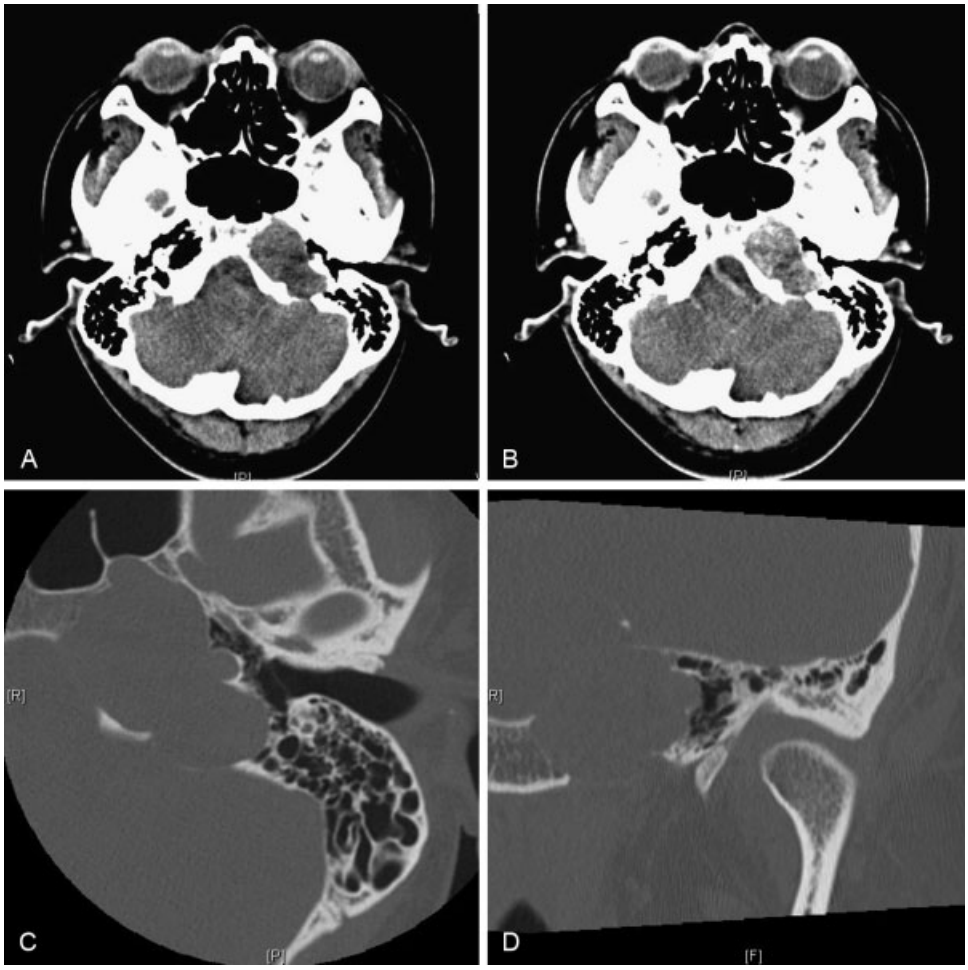

Fig. 1 (A) Axial head computed tomography (CT) shows an isodense area within the left petrous apex, 35 mm in diameter and with no calcification. The tumor has partially expanded into the posterior cranial fossa, especially the internal auditory meatus and jugular foramen. (B) enhanced axial head CT shows patches of slight enhancement. (C) High-resolution axial CT of the left temporal bone shows bony erosion of the posterior wall of the carotid canal and internal wall of the mastoid and tympanic cavities. (D) High-resolution coronal CT of the left temporal bone shows tumor mostly encased in petrous bone. Thinned-out bone cortex was maintained overall.

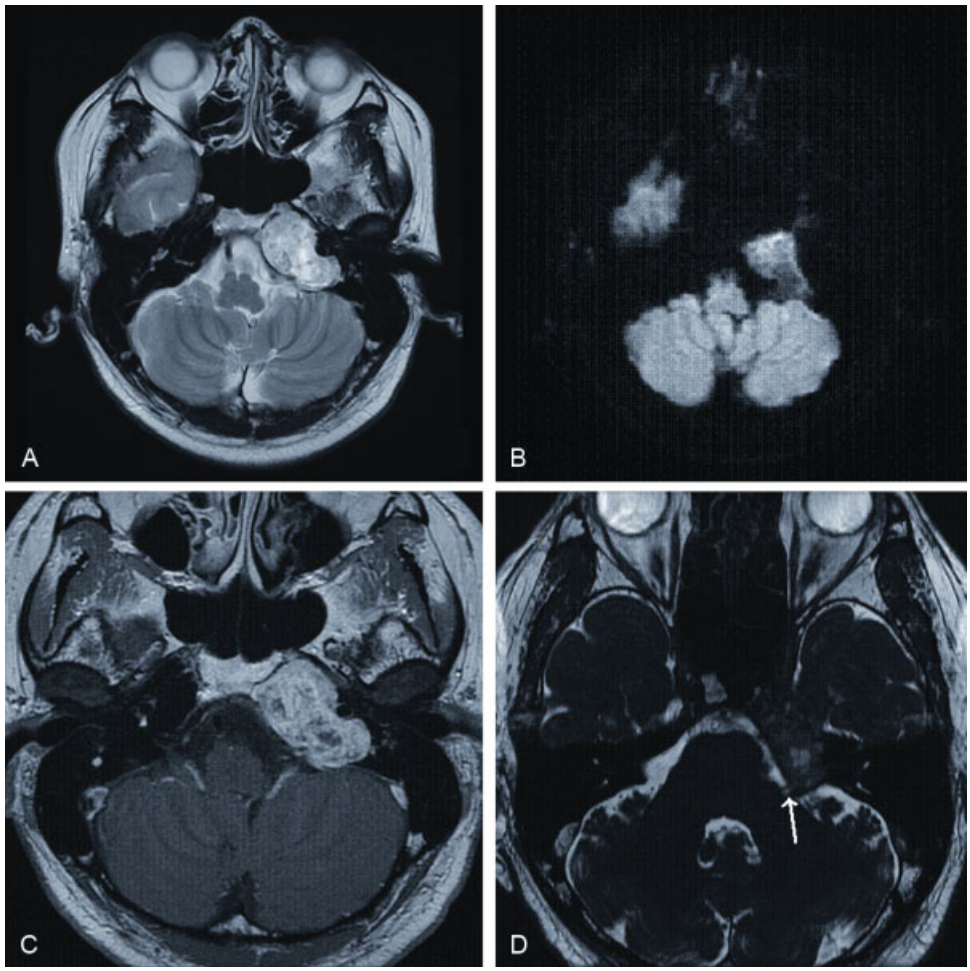

Fig. 2 (A) Head axial T2-weighted image shows a mixed-intensity lesion and no edematous effect around the tumor. (B) Head axial diffusion-weighted imaging shows a slightly hyperintense lesion with partial isointensity. (C) Contrast-enhanced magnetic resonance imaging shows heterogeneous enhancement commensurate with the dappled T2-weighted imaging intensity. (D) Fast imaging using steady-state acquisition shows facial and auditory nerves located dorsally and isolated from the lesion in the cerebellopontine angle (arrow, auditory nerve).

of the tumor was performed with the assistance of a neuroendoscope (Olympus, Tokyo, Japan). Partial defect of the posterior cranial fossa dura was identified (►**Fig. 3B**). The abducens nerve could not be identified during surgery. Intraoperative monitoring of ABR, EMG, and EOG showed no marked changes at any time during the procedure.

Tumor Pathology

Histopathologically, spindle-shaped cells that showed a palisading pattern as well as hyalinized vessels, fibrous formations, lymphocyte invasion, and hemorrhage were identified (►**Fig. 3C, D**). The histopathologic diagnosis of the tumor was schwannoma.

Postoperative Course

CT performed 1 day postoperatively showed no sign of apparent residual tumor. The postoperative course of the patient was uneventful. Abducens nerve palsy resolved immediately after surgery. The patient was discharged from our hospital 8 days postoperatively without neurologic sequelae.

Discussion

The pathologic diagnosis of the tumor in the present case was schwannoma. From the location, schwannoma in the present case could have originated from cranial nerves V, VI, VII, or VIII.

The greater superficial petrosal nerve (GSPN) and lesser superficial petrosal nerve (LSPN) were not thought to have been the origin because of the intraoperative findings. We separated the intact GSPN and LSPN lying on the floor of the middle cranial fossa to retract the temporal lobe. Abducens nerve schwannoma was strongly suspected; however, the chief complaint of abducens nerve palsy showed complete resolution postoperatively. In addition, EOG of the abducens nerve revealed no changes. These findings definitively rule out the abducens nerve as the origin of the tumor. ABR and EMG showed no marked changes throughout the procedure. In addition, the patient showed a good postoperative course with no new neurologic deficits such as facial palsy, hearing disturbance, or facial sensory disturbance. Schwannoma in this case was thus considered intraosseous.

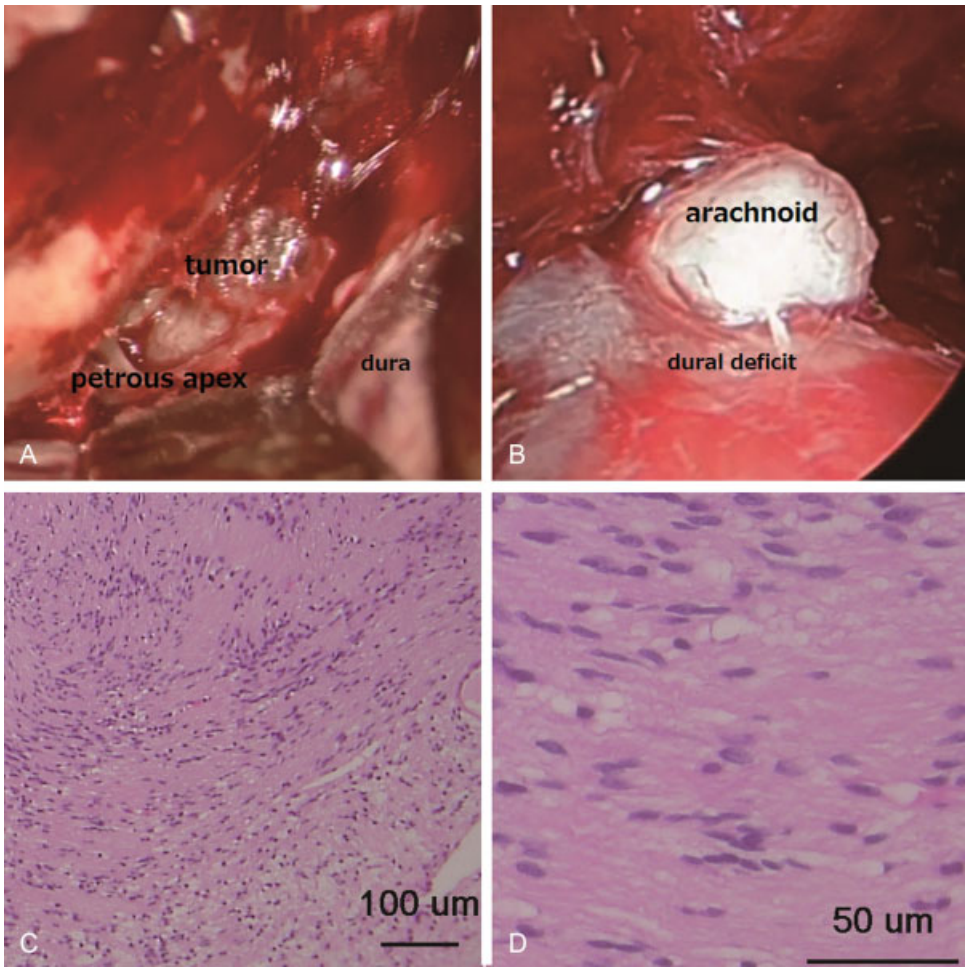

Fig. 3 (A) The tumor was comprised of soft yellow tissue. We removed the surface of the petrous apex and could identify the tumor components within. The greater superficial petrosal nerve and lesser superficial petrosal nerve were dissected free from the dura of the temporal lobe and kept intact. (B) The tumor partially destroyed the bone of the posterior side of petrous apex and dura of the posterior cranial fossa. The tumor was partially adherent to the posterior fossa dura. Neuroendoscopy showed complete resection of the tumor and a small dural deficit. The arachnoid of the posterior cranial fossa was apparent throughout. (C) Photomicrograph showing spindle-shaped cells in a palisading pattern with Antoni A and B patterns. Typical schwannoma is evident. Hematoxylin and eosin (H&E) stain; original magnification ×10. Magnification bar: 100 μm. (D) Photomicrograph showing spindle-shaped cells with hyalinized vessels, fibrous formations, lymphocyte invasion, and hemorrhage. H&E stain, original magnification ×40. Magnification bar: 50 μm.

Intraosseous schwannoma is a rare benign neoplasm that arises from Schwann cells of the nerve sheath and involves the substance of bone.[1,2] We must think about the possible origins of intraosseous schwannoma. Such a tumor could arise from Schwann cells of sensory nerves distributed on the dura, from pial cells, from Schwann cells of the subarachnoid, pial or parenchymal nerve plexuses around vessels, or from embryologic migration of Schwann cells. Intraosseous schwannoma is mostly considered to originate according to this last theory.[5–8]

Some differential diagnoses must be considered for mass lesions arising within the petrous apex as in this case including cholesteatoma, mucocele, chordoma, chondrosarcoma, epidermoid cyst, GSPN schwannoma, schwannoma originated from deep petrous nerve, and intraosseous schwannoma. Retrospectively, these differential diagnoses could be excluded based on radiologic findings. First, cholesteatoma (or epidermoid cyst) usually demonstrates no contrast enhancement and high signal intensity on DWI. Smooth erosion of bone can be seen on bone window setting CT. That is related to mastoid air cells. Second, mucocele shows only peripheral enhancement on contrast-enhanced imaging. Unlike schwannoma, mucocele generally shows destruction of septa on CT. Third, chondrosarcoma also demonstrates a mineralized chondroid matrix with a “ring-and-arc” appearance. This lesion can be seen infiltrating bone with eroded fragments on CT. Chordoma and chondrosarcoma showed avid enhancement or sometimes no enhancement. In addition, they usually display no calcification.[9,10] Fourth, schwannoma originated from deep petrous nerve tend to show the tumor encasement of the internal carotid artery.[11] In this case, CT showed tumor adjacent only to the posterior wall of the internal carotid artery.

In contrast, MRI of usual schwannoma arising from cranial nerves shows signal isointensity on T1-WI, mixed signal intensity on T2-WI, and heterogeneous enhancement. Nonenhancing lesions show high intensity on T2-WI, indicating the Antoni B region, and enhancing lesions show low

intensity on T2-WI, indicating the Antoni A region.[12,13] In addition, DWI usually shows a lesion displaying either slight hyperintensity or isointensity.

Arriving at a differential diagnosis of schwannoma preoperatively was difficult because the lesion was completely located within petrous bone, which is very unusual for schwannoma. However, images in this case were compatible with schwannoma except for the location.

To date, only two cases of intraosseous schwannoma of the petrous apex as in this case have been reported. The present case and the two previously reported cases show some radiographic characteristics similar to usual schwannoma arising from cranial nerves. However, this case also showed some different features from the two reported cases.

First, tumor size was markedly larger than the two reported cases, with the mass occupying most of the petrous apex. In the reviewed reports, the intraosseous schwannomas of the petrous apex were found despite their small size because the petrous apex is an area containing many important cranial nerves, usually resulting in the early appearance of symptoms.[3,4] However, the present case was first identified after the tumor had enlarged and compressed the abducens nerve. Symptoms resulted from nerve compression by the neighboring tumor in this case, rather than the nerve itself being neoplastic; therefore, this may imply that the time from symptom onset is related to the location of the causative Schwann cell.

In addition, this case showed other points of difference from the reported cases. Patients in the reported cases showed symptoms of headache, tinnitus, and decreased facial sensation. Our case showed no such symptoms and only abducens nerve palsy despite the large size of the tumor. This is also related to the location of the causative Schwann cell, as mentioned earlier.

At our institution, at the time of surgical removal of schwannomas including vestibular schwannoma, subcapsular removal is preferred when gross total removal would significantly jeopardize cranial nerve function. Subcapsular removal of schwannoma is also recommended by other groups in terms of preservation of neural function.[14] We adopted subcapsular resection in the present case because tumor removal for the lesion had to be performed partially blind. This is because the surgical corridor of anterior transpetrosal approach is a horizontal direction from the lateral side, and the caudal part of the lesion was not fully visualized by the approach. Furthermore, in this case, CT showed the destroyed bone surrounding the internal carotid artery, so we could avoid injury using the subcapsular technique. CT performed 1 day postoperatively showed no sign of apparent residual tumor, and nearly total removal of the tumor was confirmed.

We must emphasize the need for follow-up (especially when a gross total resection is not performed), given the small risk of growth of residual tumor.

Conclusion

We emphasized the importance of considering the differential diagnosis of intraosseous schwannoma for osteolytic mass lesions within the petrous apex. From our experience, subcapsular removal of the tumor seems useful for preserving surrounding neural and vascular structures. The results of long-term follow-up are awaited.

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