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Surgical Management of Skull Base Chondroblastoma

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Objectives: **Chondroblastoma is a rare tumor accounting for 1% of primary bone tumors. Chondroblastoma involving the skull base is exceedingly rare with approximately 60 cases reported. We reviewed our experience with chondroblastoma of the skull base with an emphasis on current lateral skull base approaches and long-term tumor control.** ***Study Design and Setting:*** **A retrospective case review at a tertiary neurotology private practice group was performed over a 20-year period. Five patients were identified with skull base chondroblastoma. All patients underwent surgical intervention, and success of surgery was determined by disease-free status at last follow-up. Mean follow-up time was 5.8 years.** ***Results:*** **Two patients underwent gross tumor removal as primary therapy. One patient underwent partial tumor removal at an outside institution, and follow-up magnetic resonance imaging demonstrated rapid growth of residual tumor. This patient was successfully treated with gross total removal of residual tumor with an infratemporal craniotomy approach. Near total tumor removal was performed in two patients because of intimate involvement of vital structures. At last follow-up, no patient had radiographic evidence of tumor recurrence. There were no significant postoperative complications.** ***Conclusions:*** **Gross total or near total resection of skull base chondroblastomas through lateral skull base approaches results in long-term tumor control and low complication rates.** ***Key Words:*** **Skull base, chondroblastoma, temporal bone, neoplasm.**

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INTRODUCTION

Chondroblastoma is a rare neoplasm of bone origin that typically arises in the epiphysis of long bones. It comprises approximately 1% of all primary bone neoplasms. Chondroblastoma originating in the temporal bone was first described by Denko and Krauel[1] in 1955. Since this first description, approximately 60 cases of chondroblastoma of the temporal bone have been reported, including a few case series.

Surgical management of skull base neoplasms presents a unique challenge to the neurotologist. En bloc resections are usually not possible because of the critical structures intrinsic to the temporal bone. In addition, combinations of approaches are generally required for extensive lesions. The development and refinement of these approaches have led to excellent tumor control with preservation of vital structures. We report five cases of skull base chondroblastoma with an emphasis on clinical presentation, radiographic and pathologic findings, surgical management, and long-term outcomes.

PATIENTS AND MATERIALS

Over a period from 1985 to 2005, five patients were identified with histologically confirmed chondroblastoma of the skull base. One patient was initially evaluated and treated at an outside institution and presented for further management secondary to growth of residual disease. The remaining four patients underwent initial evaluation and treatment at the authors' institution. The mean age was 59 (range, 39–85) years. Four patients were female and one patient male. Clinical history, surgical approach, and follow-up data were obtained through retrospective chart review. Patients underwent high-resolution computed tomography (CT) or T1-weighted gadolinium enhanced magnetic resonance imaging (MRI) of the skull base per the treating surgeon's preference.

CASE REPORTS

Case 1

A 39-year-old female underwent partial resection of a left temporal bone lesion at an outside institution 2 months before presentation. The final pathology was thought to be a giant cell reparative granuloma. She was referred to our institution for further treatment because of significant growth of the residual mass. Her initial symptoms included severe headaches, left otalgia, and left facial paresthesias. Her past medical history was noncontributory. Physical examination revealed a well-healed craniotomy scar, no cranial nerve deficits, normal hearing, and no temporomandibular joint (TMJ) complaints. Audiometric testing was within normal limits. High-resolution CT and T1-gadolinium

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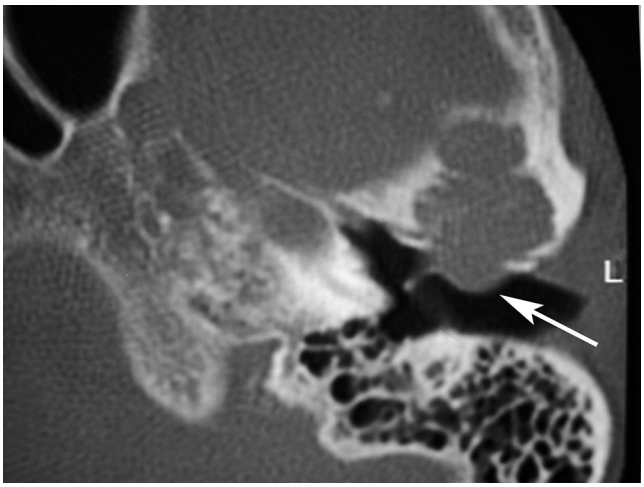

Fig. 1. Axial CT shows expansile lesion in the supra-glenoid region with scalloped borders eroding the anterior wall of the bony external auditory canal.

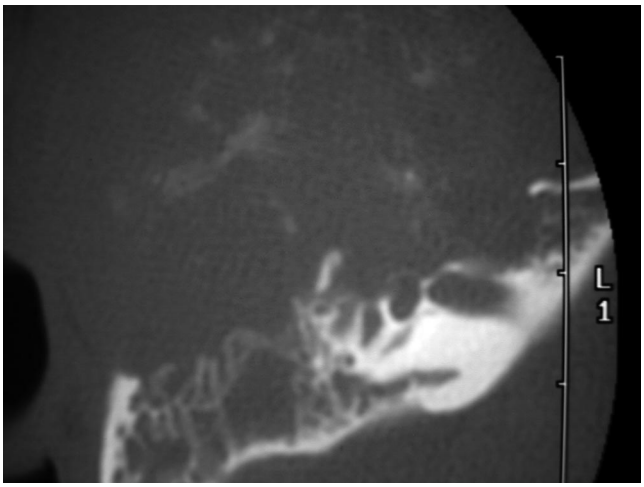

Fig. 2. Axial CT shows destruction of the external auditory canal, middle ear, cochlea, geniculate fossa and floor of the middle cranial fossa. Calcifications can be seen dispersed throughout the mass.

enhanced MRI demonstrated a residual lesion that was located anterior to the left condyle and extended medially to involve the foramen spinosum and foramen ovale. A preauricular, infratemporal approach was used for tumor removal with the use of intraoperative facial nerve monitoring. The tumor was transdural and involved the epitympanum, lateral petrous apex, and the geniculate ganglion. All involved bone was removed, resulting in gross total resection. Final pathology was consistent with a chondroblastoma of the left temporal bone. She had no cranial nerve deficits postoperatively and showed no evidence of recurrence by the most recent T1 gadolinium-enhanced MRI 4 years after resection.

Case 2

An 85-year-old female presented with a long history of bilateral hearing loss (HL), left greater than right. A few months before presentation, she noticed increased difficulty using the telephone with her left ear. Physical examination revealed a subcutaneous smooth mass in the left anterior external auditory canal that moved with TMJ movement. She had no cranial nerve deficits. Audiometric evaluation revealed bilateral, symmetric sensorineural HL with decreased left speech discrimination scores. High-resolution CT revealed a 1.3×0.8 cm mass superior to the TMJ and extending to the middle fossa dura and toward the middle ear (Fig. 1). This mass was thought to be a cholesterol granuloma, and she subsequently underwent a mastoidectomy. Pathology was consistent with a chondroblastoma. She subsequently underwent a transmastoid-subtemporal approach with removal of the root of the zygoma and supra-auricular temporal bone. The tumor required dissection from the TMJ and temporal lobe dura. There were no postoperative complications or cranial nerve deficits. Her most recent high-resolution CT 3 years later showed no evidence of recurrence.

Case 3

A 39-year-old female presented with right-sided HL for approximately 1 year and acute onset of right facial paresis for 5 weeks. Physical examination revealed collapse of her right external auditory canal. Her right facial nerve function was House-Brackmann grade 3/6. Audiogram revealed a right profound HL. She had no other cranial nerve deficits and did not complain of TMJ pain. A high-resolution CT demonstrated a large lesion centered above the glenoid fossa (Fig. 2). An open biopsy of the external auditory canal mass was performed and was consistent with a chondroblastoma. An infratemporal craniotomy and condylectomy was performed. The anterior and superior external auditory canal was involved with tumor. The condyle was removed to obtain adequate tumor margins. A large region of supra-auricular temporal bone measuring 6 cm $\times$ 5 cm $\times$ 7 cm was involved with tumor and removed. The superior semicircular canal and cochlea were involved with tumor, and the labyrinth and cochlea were removed. Tumor was dissected from the facial nerve at the internal auditory canal fundus, the fifth cranial nerve, and from a dehiscent petrous carotid artery. Small tumor remnants were left on the facial nerve and fifth cranial nerve. In addition, the tumor involved the middle fossa dura. At the end of the procedure, the external auditory canal was oversewn. Her facial nerve function returned to a House-Brackmann grade 1/6. She complained of transient facial paresthesias that eventually resolved. Her most recent T1 gadolinium-enhanced MRI 3 years later demonstrated no evidence of recurrence.

Case 4

A 70-year-old male presented with a 3-month history of progressive right HL and tinnitus. On physical examination, he was noted to have a bluish mass located in the external auditory canal. Audiometric evaluation revealed a left moderate sensorineural HL and a right mixed HL. CT demonstrated a 3 cm ovoid, expansile mass in the anterior temporal bone, just superior to the TMJ. The lesion extended as far medial to the eustachian tube and foramen spinosum. The tumor also involved the middle ear and ossicles. The otic capsule appeared intact. Biopsy in the office was inconclusive. He later underwent a middle fossa craniotomy with debulking and curettage of the lesion. The tumor was found to be adherent to temporal lobe dura. It was separated from the malleus and incus without disturbing the ossicles. A near total resection was performed and the final pathology was consistent with a chondroblastoma. He had no postoperative cranial nerve deficits, and his hearing remained stable. His most recent CT scan 18 years later showed no recurrence.

Case 5

A 62-year-old female presented with left HL, otalgia, and aural fullness for 15 months. Physical examination revealed a left external auditory canal mass occluding most of the canal. Audiometric testing revealed normal hearing on the right and a

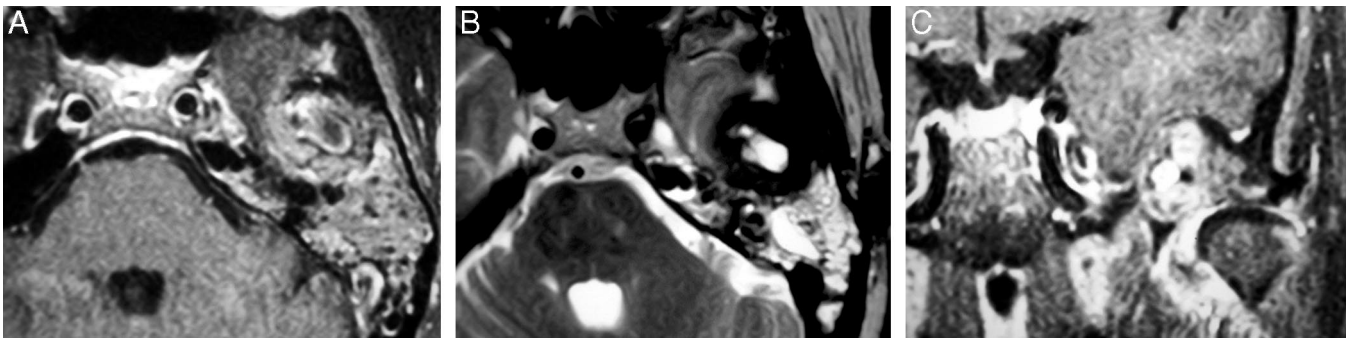

Fig. 3. (A) Axial T1-weighted, gadolinium enhanced MRI shows oviod heterogeneously enhancing mass medial to the glenoid fossa on the floor of the middle cranial fossa. (B) Axial T2-weighted MRI shows hypointense ovoid mass on the middle fossa floor with a markedly hyperintense center, lying anterior to the hyperintense liquid-containing mastoid antrum. (C) Coronal T1-weighted, gadolinium enhanced MRI shows a heterogeneously enhancing rounded mass expanding the floor of the middle fossa in the supra-glenoid region. The mass bulges superiorly, elevating the temporal lobe and inferiorly towards the infratemporal fossa.

conductive HL on the left. MRI and CT revealed an expansile osteolytic process eroding the articular margin of the mandibular condyle with extension into the middle ear and middle cranial fossa (Fig. 3). She subsequently underwent an infratemporal fossa craniotomy with facial nerve monitoring. Her external auditory canal was oversewn. Tumor was found filling the eustachian tube and involved a dehiscent carotid artery. The temporal lobe dura was involved with tumor and subsequently removed. The temporal bone lateral to the foramen ovale was removed and the glenoid fossa was opened. Tumor involved the infratemporal fossa and the mandibular ramus. Gross total resection was achieved with the pterygoid muscles as a medial margin. She experienced no cranial nerve deficits and had an uneventful recovery. Six months after resection, a T1 gadolinium-enhanced MRI showed no evidence of recurrence.

RESULTS

Demographics and Clinical Presentation

Table I summarizes the demographics, clinical presentation, surgical approach, and follow-up for this series of patients. Three tumors involved the left temporal bone, whereas two involved the right. Four of five patients presented with an external auditory canal mass and HL. Two patients complained of otalgia. One patient presented with House-Brackmann grade 3/6 facial paresis that resolved after surgery. Another patient complained of facial paresthesias. No other cranial nerve neuropathies were noted, and no patient complained of trismus or TMJ pain.

Tumor Characteristics

Surgical approach was dependent on extent and location of tumor, patient's age, and surgeon's preferences. Table II summarizes the structures involved with tumor. Three patients underwent an infratemporal fossa craniotomy, one patient underwent an extended middle fossa craniotomy, and one patient underwent a transmastoid-subtemporal approach. In all five cases, tumor involved temporal lobe dura and required resection of the involved dura. The TMJ was directly involved in two cases, with one patient requiring a condylectomy to obtain adequate margins. The tumor had exposed the petrous carotid

TABLE I.
Demographic Data, Presenting Symptoms, Surgical Approach, and Follow-Up in Five Patients With Chondroblastoma.

Patient	Age (yr)	Sex	Side	Symptoms	Surgical Approach	Extent of Resection	Follow-Up (yr)
1	39	Female	Left	1. Otalgia 2. Headaches 3. Facial paresthesias	ITFC	Gross total	4
2	85	Female	Left	1. Hearing loss 2. EAC mass	Transmastoid-subtemporal	Gross total	3
3	39	Female	Right	1. Hearing loss 2. EAC mass 3. Facial nerve paresis	ITFC	Near-total	3
4	70	Male	Right	1. Hearing loss 2. EAC mass 3. Tinnitus	Extended MFC	Near-total	18
5	62	Female	Left	1. Hearing loss 2. EAC mass 3. Otalgia	ITFC	Gross total	1

ITFC = infratemporal craniotomy; EAC = external auditory canal; MFC = middle fossa craniotomy.

TABLE II.
Significant Structures Involved With Tumor.

Involved Structures	No. of Patients
Temporal lobe dura	5
External auditory canal	4
Temporomandibular joint	2
Carotid artery	2
Ossicles	2
Trigeminal nerve	1
Facial nerve	1
Cochlea	1
Vestibular labyrinth	1

artery in two cases, and tumor was successfully removed from the exposed carotid artery in both cases.

The middle ear space was involved with tumor in two patients and involved the ossicular chain in both cases. Tumor was successfully dissected free from the ossicular chain in one patient. The other patient had direct involvement of the cochlea and superior semicircular canal, requiring removal of the labyrinth and cochlea. The trigeminal nerve was directly involved in one patient, and a small remnant of tumor was left. The patient that presented with facial nerve paresis had tumor involving the nerve. The integrity of the nerve was maintained, although a small amount of tumor was left on the facial nerve sheath.

Complications

One patient developed an infected bone flap associated with an epidural abscess 6 months after surgery. All hardware was removed, and she had no subsequent complications. There were no incidences of meningitis, cerebrospinal fluid leak, postoperative cranial nerve deficits, or recurrence. One patient had transient trigeminal paresthesias that resolved, and another patient recovered from her preoperative facial nerve paresis.

DISCUSSION

Chondroblastoma, first reported by Codman[2] in 1931 as a chondromatous giant cell tumor, is an unusual benign neoplasm of bone that represents less than 1% of all bony neoplasms. In 1942, Jaffe and Lichtenstein[3] renamed the tumor a chondroblastoma, emphasizing the origin of the tumor to be the chondroblast. The vast majority of chondroblastomas arise in the epiphysis of long bones in children and young adults in their second or third decade of life with a predilection for males.[4] In contrast, temporal bone chondroblastomas arise in older patients, as demonstrated in a series of temporal bone chondroblastomas by Bertoni et al.[5] with a mean age of 47.8 (range, 29–70) years. Our series is also characterized by an older patient population (mean, 59 yr).

In this series, the most common presenting symptom was HL, consistent with previous reviews.[6,7] HL may be conductive because of occlusion of the external auditory canal or ossicular involvement or sensorineural if the labyrinth is involved. Two of the five patients had otalgia but none presented with specific TMJ pain or trismus. Surprisingly, both patients with direct involvement of the TMJ identified during surgery did not present with specific TMJ complaints. Four of the patients presented with an anterior external auditory canal mass. Movement of the mass with jaw opening confirmed involvement of the TMJ. One patient had a facial nerve palsy at presentation that resolved after surgery. Facial palsy is a rare presentation but has been reported previously.[8]

Chondroblastomas of the skull base generally involve the lateral temporal bone.[5] By imaging, all of the tumors in our series appeared centered above the supraglenoid region of the temporal bone (Fig. 1). On CT, the margins are sharply defined to moderately well-defined. Scalloped borders with a sclerotic rim are generally present. Although commonly reported in the literature,[9,10] calcifications were observed in only the largest tumor in our series (Fig. 2). On MRI, chondroblastomas are intermediate in signal intensity on T1-weighted images and heterogeneous after gadolinium-enhancement (Fig. 3A and B). The enhancement in our series is much less homogeneous than in some of the previously reported cases.[10,11] However, on T2-weighted images, their heterogeneity with components of marked hyperintensity (Fig. 3C) is consistent with the limited number of previously reported cases with MRI.[10–12]

Tumors involving the skull base and characterized by giant cells include cholesteatoma, reparative giant cell granuloma, giant cell tumor, aneurysmal bone cyst, brown tumor of hyperparathyroidism, and chondroblastoma. On gross examination, chondroblastomas are typically red to brown and granular. Microscopically, the lesion is cellular with sheets of mononuclear polyhedral cells admixed with multinucleated giant cells (Fig. 4). The mononuclear cell membranes appear thick and sharply defined. The nuclei are round to slightly indented with inconspicuous nucleoli and moderate to large amounts of eosinophilic cytoplasm.[13] Intracytoplasmic glycogen granules are sometimes present. Reticulum fibers appear to surround each individual cell. Mitotic figures are rarely encountered. Areas of hemorrhage are frequently seen and are

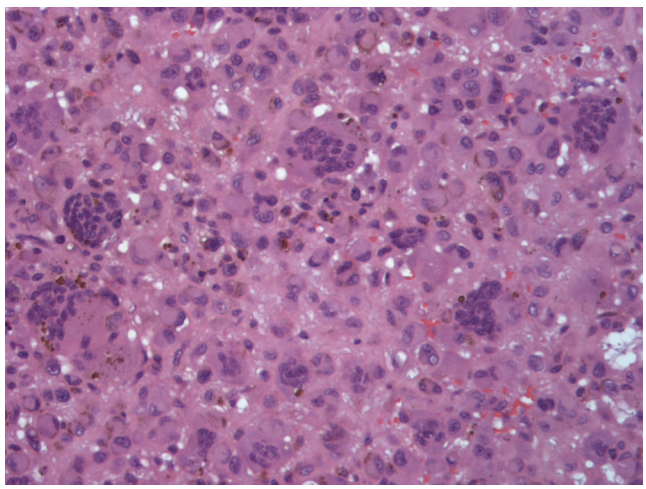

Fig. 4. Light microscopy demonstrates sheets of polyhedral cells admixed with scattered multinucleated giant cells. A few brown hemosiderin granules are present.

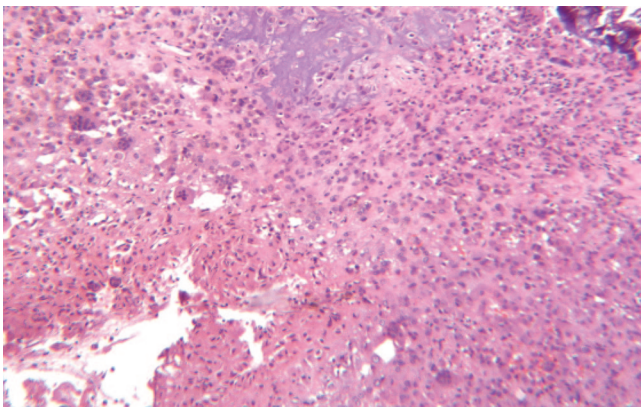

Fig. 5. Light microscopy demonstrates an area of chondroid differentiation and sheets of polyhedral tumor cells.

characterized by hemosiderin granules. Area of cartilage differentiation are often present (Fig. 5). A distinctive microscopic finding is the presence of zones of lacy calcifications, so-called "chicken wire" calcification (Fig. 6). By electron microscopy, the chondroblastoma cells closely resemble normal cartilage cells grown in tissue culture.[14,15]

Chondroblastomas express S-100 protein and vimentin.[16] They may also be immunoreactive for neuron specific enolase, muscle specific actin, and low molecular weight keratin.[17,18] The tumor is occasionally misdiagnosed as a giant cell tumor because of the presence of giant cells. However, the expression of S-100 in the tumor cells differentiates it from a giant cell tumor.[16]

Previously reported treatment for chondroblastoma of the skull base have included curettage, surgical resection, radiation therapy, and combined surgery and radiation therapy. Recurrence rates after curettage have been reported to be as high as 43%.[19] Radiation therapy should be reserved for poor surgical candidates or unresectable disease because there is a potential late development of malignant degeneration.[20] Although en bloc resection is the treatment of choice for chondroblastoma of long bones, en bloc resection of skull base lesions is often not achievable. However, gross total removal of the tumor is usually possible with current skull base techniques.

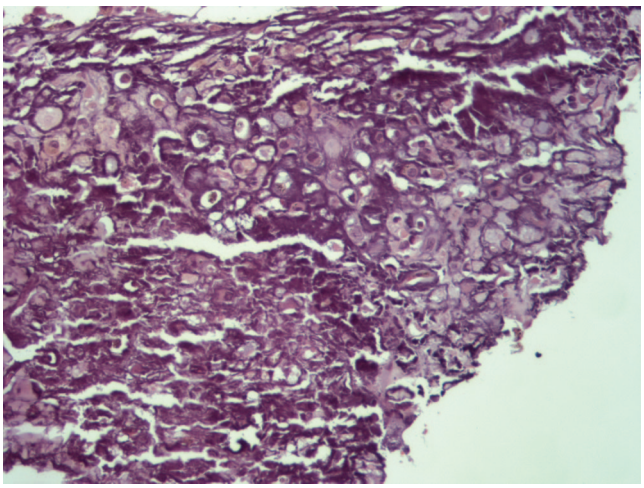

Fig. 6. Light microscopy demonstrates lacy calcifications in a "chicken wire" pattern.

Surgical removal using current skull base surgical techniques is the mainstay of treatment. The goal should be total gross removal of the tumor while maintaining the function of critical structures such as the cranial nerves and carotid artery. Because these tumors arise from the anterior temporal bone near the TMJ, adequate exposure to the zygoma and TMJ are essential. In addition, medial spread along the middle fossa floor toward the geniculate ganglion and foramen ovale are also important considerations in surgical planning. With tumor limited to the middle fossa floor, an extended middle fossa craniotomy may be sufficient. However, with larger tumors involving the TMJ, a preauricular, infratemporal fossa approach provides more adequate exposure. In limited cases, an extended transmastoid-subtemporal approach with removal of the root of the zygoma may be adequate. With large tumors, a combination of approaches may be required for adequate exposure. Vital structures such as the carotid artery, facial nerve, and labyrinth should not be sacrificed for tumor removal unless significant preoperative deficits are present. Both patients in this series with near total removal did not have radiographic evidence of recurrence with follow-up.

Metastasis has been reported in chondroblastoma of the ribs.[21] However, there have been no reported cases of metastasis from skull base chondroblastoma, so metastatic work-up is not indicated unless there are other suggestive symptoms. Local recurrences have been reported but are less when total gross resection can be achieved.[22] Currently, there is no role for chemotherapy in the treatment of chondroblastoma.

CONCLUSION

Chondroblastoma of the temporal bone is a rare neoplasm that arises near the glenoid fossa and can involve the anterior middle fossa floor. Patients typically present with HL and an anterior external auditory canal mass. Chondroblastomas appear as well-defined osteolytic lesions with calcifications being a common finding. Histologic diagnosis can be challenging, and immunohistochemical staining techniques can be helpful. Dural involvement was found in all patients in this series and should be considered in preoperative planning. Treatment should be aimed at gross total removal and may require a combination of skull base approaches; however, vital structures should be conserved when possible.

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