

Temporal Bone Chondroblastomas

Allison Pontius, MD,* Paul Reder, MD,† and
Yadranko Ducic, MD, FRCS(C), FACS*†

Objectives: To review temporal bone chondroblastomas in regards to their presentation, radiographic findings, histopathology, and treatment.

Study Design: Case report and literature review.

Methods: A case report of a 38-year-old man is reviewed who presented with left-sided mixed hearing loss, otalgia, otorrhea, and a left external auditory canal mass. A computed tomography scan revealed a 6-cm mass involving the petrous and squamous portions of the left temporal bone.

Conclusions: Temporal bone chondroblastomas are extremely rare osseous tumors with only 35 cases previously reported in the literature. Presenting symptomatology and pathology may be confused with more common lesions seen in the temporal bone. Diagnostic radiology, including computed tomography and/or magnetic resonance imaging, as well as immunohistochemical staining with S-100 protein may assist in making the diagnosis. Treatment is complete surgical excision with preservation of vital neurovascular structures. (Am J Otolaryngol 2003;24:370-373. © 2003 Elsevier Inc. All rights reserved.)

Chondroblastoma is a rare osseous neoplasm, usually noted in the epiphyses of long bones in males under the age of 25 years. It accounts for only 1% of all tumors of bone and most commonly presents in the humerus. It was originally recognized as being distinct from giant cell tumors by Codman,¹ with the term *benign chondroblastoma* first introduced by Jaffe and Lichtenstein² in 1942. Chondroblastoma of the temporal bone is exceedingly rare with only 35 cases previously reported in the literature. In this brief article, we review a case of temporal bone chondroblastoma that presented with hearing loss, otorrhea, and otalgia and was initially presumed to be a cholesteatoma.

CASE REPORT

A 38-year-old man presented with a 2-month history of left-sided otalgia and otorrhea. Physical examination revealed a mass

in the external auditory canal. Audiogram showed a left-sided moderate-to-severe sensorineural hearing loss with a severe-to-profound conductive component. A biopsy of the mass visible within the external auditory canal was consistent with cholesteatoma. Computed tomography (CT) scan of the left temporal bone showed a 6-cm lesion with significant osseous destruction of the petrous and squamous portions (Fig 1). The patient was, however, lost to follow-up for the next 7 months. He then represented with progression of his pathologic process. He was now noted to have an appreciable mass within the left temporal region on physical examination. Other than the left-sided mixed hearing loss, cranial nerves 2-12 were intact, and the patient exhibited no peripheral or central neurologic deficits. There was no evidence of trismus. The patient was brought to the operating room where a combined intracranial/extracranial approach to expose the lesion safely and remove it completely was planned under the presumed diagnosis of a large, expansile cholesteatoma with intracranial and extratemporal extension.

The left temporal bone was exposed via a curvilinear incision over the mid-to-upper portion of the superficial temporalis muscle, arching over the squamous portion of the temporal bone, coming down posteriorly along the occiput and down into the upper portion of the neck. The overlying tissues were then

From the *Department of Otolaryngology-Head and Neck Surgery, University of Texas Southwestern Medical Center, Dallas, TX; and the †Division of Otolaryngology and Facial Plastic Surgery, John Peter Smith Hospital, Fort Worth, TX.

Address correspondence to: Yadranko Ducic, MD, FRCS(C), FACS, Director, Otolaryngology and Facial Plastic Surgery, John Peter Smith Hospital, 1500 S Main St, Ft Worth, TX 76104. E-mail: yducic@aol.com

© 2003 Elsevier Inc. All rights reserved.

0196-0709/03/2406-0000\$30.00/0

doi:10.1053/S0196-0709(03)00084-X



Fig 1. Coronal computed tomography scan showing destruction of the squamous portion of the temporal bone and temporomandibular joint by an expansile mass.

elevated, and the temporal bone was completely exposed. The sternocleidomastoid muscle was released from the mastoid tip, and the incision was carried through the periosteum with reflection of the auricle anteriorly. The external auditory canal was transected within its osseous portion immediately lateral to the lesion present within the canal. Next, a mastoidectomy was performed. At this point, it was noted that there was a yellowish-brown mass filling the mastoid air cells with extension superiorly causing erosion through the tegmen mastoideum into the squamous portion of the temporal bone. The temporalis muscle was then elevated and released superiorly to expose the underlying tumor that was protruding through the temporal fossa directly from the middle cranial fossa. The tumor had eroded through the anterior external auditory canal and the tegmen, as well as the temporomandibular joint. The middle ear was not grossly involved with tumor; however, the incus was noted to be partially eroded. The malleus and the remaining incus were removed to allow for obliteration of the eustachian tube orifice. The stapes and the stapedius tendon were visualized and left intact. The facial nerve was fully intact and within its canal despite erosion of the temporal bone down to and including the scutum and roof of the epitympanum. The tympanic membrane and middle ear mucosa were fully removed, with subsequent obliteration of the middle ear, external auditory canal, and eustachian tube. Next, a middle fossa craniotomy was

performed to fully expose the intracranial portion of the tumor. During dissection of the tumor, it was noted to extend into the temporal lobe of the brain through the dura. The dural defect that was created was then repaired with a dural patch graft after complete tumor extirpation. Reconstruction of the skull base defect was performed with a pedicled temporalis muscle flap. A titanium mesh/hydroxyapatite cement cranioplasty was then performed to rehabilitate the calvarial defect. A 2.0-mm dynamic titanium mesh was fashioned to cover the defect with 0.5 to 1.0 cm of overlap at the periphery. The titanium mesh scaffold was rigidly fixed in place with a series of titanium screws, and then the hydroxyapatite cement was placed on the scaffold, completely covering it. This was allowed to set for 20 minutes; then the scalp was closed over bulb drains.

The patient's postoperative course was uncomplicated without development of a cerebrospinal leak. He has maintained an excellent contour in the temporal region with a well-healed incision. His cranial nerve and neurologic examination were normal, other than for the expected loss of hearing on the operated side. The patient remains clinically and radiographically disease free at 12-month follow-up.

Histologic evaluation of the specimen revealed the presence of sheets of polyhedral, mononuclear tumor cells with extensive areas of golden brown pigment deposition. Interspersed with the mononuclear cells were numerous, multinucleated giant cells (Fig 2). The decalcified sections contained multiple areas of chondroid differentiation associated with microscopic calcification that was in a "chicken wire" configuration. An occasional mitosis was seen. The tumor was highly vascularized throughout with no obvious areas of necrosis.

DISCUSSION

Chondroblastoma of the temporal bone was first reported by Denko and Krauel³ in 1955. Since that time, 36 cases of temporal bone chondroblastoma, including this one, have been presented in the literature.

The age range of patients presenting with temporal bone chondroblastoma has been re-

ported between 2 and 70 years of age, with a mean age of 33 years old. There is a male preponderance with a male-to-female ratio of 2 to 1.⁴ The most common presenting symptoms include hearing loss, otalgia, tinnitus, aural fullness, and vertigo or dysequilibrium. The most common presenting sign is that of a mass within the external auditory canal or temporal area. The differential diagnosis of a temporal bone chondroblastoma includes cholesteatoma, giant-cell tumor, aneurysmal bone cyst, solitary bone cyst, enchondroma, chondrosarcoma, and other osteogenic and cartilaginous tumors.

Radiographically, these tumors present as an osteolytic lesion of the temporal bone. They are sharply separated from the surrounding normal bone by a thin margin of increased osseous density.⁵ On CT, chondroblastoma typically appears as a high-density mass with mild homogenous enhancement and central, small unenhanced areas. Calcifications may also be seen within the mass. The appearance of chondroblastoma on MRI is variable but is usually low-to-intermediate intensity on T1 and T2 images. The cystic or fluid-filled areas may show areas of hyperintensity on T2 images.¹³ There is partial enhancement seen with the use of gadolinium.⁶

On gross pathological examination, the tumor has a reddish-gray or brown appearance and can be very vascular or even hemorrhagic. It contains gray, gritty regions with flecks of yellowish calcific material.⁵ Histologically, chondroblastomas contain cellular areas with benign multinucleated cells in variable

numbers. Mitotic figures, although usually present, are sparse, and the predominant cells are rounded or polyhedral in shape. The designation of a tumor as chondroblastoma depends on foci, typically rounded, of chondroid matrix. Dystrophic calcification in these cartilaginous islands is common.⁷ It is not uncommon to have large zones within a chondroblastoma that do not contain cartilage, and this can lead to the mistaken diagnosis of giant cell tumor because they both contain other morphologically identical cells.⁷ To differentiate chondroblastoma from other pathological processes with histological similarities, immunoreactivity of S-100 protein has been shown to be related to chondroid tissue formation. Chondroblastoma has been shown to have a strong immunoreactivity to S-100, whereas giant-cell tumor does not.^{4,8-12}

In the past, treatment for chondroblastoma has included curettage, irradiation, total en bloc excision, and surgery combined with radiation. The most effective modality based on the literature review appears to be total excision with or without radiation.⁴ However, because of the potential late development of chondrosarcoma after radiation treatment of chondroblastoma at sites other than the temporal bone, radiation is not currently recommended in the treatment of temporal bone chondroblastoma unless unresectable disease is encountered or left behind or the patient is not a good surgical candidate secondary to comorbid conditions and advanced age. There are no reports of metastatic temporal bone chondroblastoma, although there has been

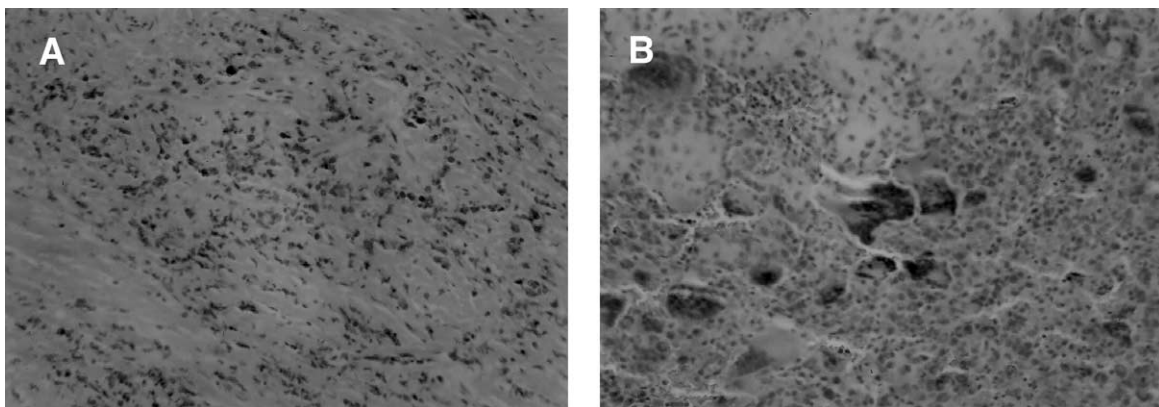


Fig 2. Hematoxylin and eosin stain (50× magnification) showing polyhedral, mononuclear tumor cells with numerous multinucleated giant cells, and multiple areas of chondroid differentiation.

one case of pulmonary metastasis from a non-temporal bone chondroblastoma.⁷ Currently, chemotherapy does not have a role in the treatment of temporal bone chondroblastoma.

Review of the literature shows that patients with temporal bone chondroblastoma treated with curettage developed unacceptably high rates of recurrent disease requiring secondary procedures.⁴ Patients who underwent an en bloc resection or total excision of tumor had no evidence of recurrence at an average follow-up of 27 months.⁴ Current recommendations for treatment include a combined intracranial/extracranial procedure to ensure complete removal of the tumor while preserving important neurovascular structures. After closure of the dural defect, we chose to reconstruct the defect with a pedicled temporalis muscle flap followed by a titanium mesh/hydroxyapatite cement cranioplasty of the temporal bone calvarial defect.¹⁴ This provides a watertight closure to prevent postoperative cerebrospinal fluid leak while also providing a natural contour to the temporal bone defect.

CONCLUSIONS

Temporal bone chondroblastoma is a rare entity with only 35 cases previously reported in the literature. It usually presents with hearing loss, otalgia, tinnitus, and an external auditory canal mass. It should be considered in the differential diagnosis of cholesteatoma, giant-cell tumor, aneurysmal bone cyst, solitary bone cyst, enchondroma, chondrosarcoma, and other osteogenic and cartilaginous tumors. Initial biopsy results may be misleading and not provide the diagnosis; however, the use of S-100 protein may help in identification of this rare lesion. Additional tools, such as computed tomography and magnetic

resonance imaging should be used for diagnosis and surgical planning. Treatment remains complete surgical excision with preservation of vital neurovascular structures.

REFERENCES

1. Codman EA: Epiphyseal chondromatous giant cell tumors of the upper end of the humerus. *Surg Gynecol Obstet* 52:543-548, 1931
2. Jaffe HL, Lichtenstein L: Benign chondroblastomas of bone: A reinterpretation of the so-called calcifying or chondromatous giant cell tumor. *Am J Pathol* 18:969-991, 1942
3. Denko JV, Krauel LH: Benign chondroblastoma of bone: An unusual location in the temporal bone. *Arch Pathol* 59:710-711, 1955
4. Varvares MA, Cheney ML, Goodman ML, et al: Chondroblastoma of the temporal bone: Case report and literature review. *Ann Otol Rhinol Laryngol* 101:763-769, 1992
5. Blaauw G, Prick JJ, Versteeg C: Chondroblastoma of the temporal bone. *Neurosurgery* 22:1102-1107, 1988
6. Watanabe N, Yoshida K, Shigemi H, et al: Temporal bone chondroblastoma. *Otolaryngol Head Neck Surg* 121:327-330, 1999
7. Dahlin DC, Ivins JC: Benign chondroblastoma: A study of 125 cases. *Cancer* 30:401-413, 1972
8. Nakajima T, Kameya T, Watanabe S, et al: An immunoperoxidase study of S-100 protein distribution in normal and neoplastic tissues. *Am J Surg Pathol* 6:715-727, 1982
9. Stefansson K, Wollmann RL, Moore BW, et al: S-100 protein in human chondrocytes. *Nature* 295:63-64, 1982
10. Ushigome S, Takakuwa T, Shinagawa T, et al: Ultrastructure of cartilaginous tumors and S-100 protein in the tumors with reference to the histogenesis of chondroblastoma, chondromyxoid fibroma and mesenchymal chondrosarcoma. *Acta Pathol Jpn* 34:1285-1300, 1984
11. Monda L, Wick MR: S-100 protein immunostaining in the differential diagnosis of chondroblastoma. *Hum Pathol* 16:287-293, 1985
12. Karabela-Bouropoulou V, Markaki S, Prevedorou D, et al: A combined immunohistochemical and histochemical approach on the differential diagnosis of giant cell epiphyseal neoplasms. *Pathol Res Pract* 184:184-187, 1989
13. Kobayashi Y, Murakami R, Toba M, et al: Chondroblastoma of the temporal bone. *Skeletal Radiol* 30:714-718, 2001
14. Ducic Y: Titanium mesh and hydroxyapatite cement cranioplasty: A report of 20 cases. *J Oral Maxillofac Surg* 60:272-276, 2002