CASE REPORT OPEN ACCESS

# A case of chondrosarcoma of the temporal bone

Sumito Jitsukawa, Kenichi Takano, Ayumi Abe, Tetsuo Himi

## ABSTRACT

**Introduction: Chondrosarcoma is a malignant neoplasm of cartilage-forming-tissue and rarely encountered in the temporal bone. The clinical aspects and management of this rare lesion are discussed. Case Report: We report a chondrosarcoma of the temporal bone presenting with facial nerve palsy in an 80-year-old female. A computed tomography scan and magnetic resonance imaging showed a destructive mass located in the right mastoid air cells and extending into the venous sinus. We performed mastoidectomy and tympanotomy to determine a definitive diagnosis and to remove the temporal bone lesion. Conclusion: Chondrosarcoma should be considered in the differential diagnosis of patients with slowly progressive osseous tumors. It is important to obtain an early diagnosis and resection before the tumor becomes extensive.**

**Keywords: Chondrosarcoma, Temporal bone, Facial palsy**

Sumito Jitsukawa[1], Kenichi Takano[2], Ayumi Abe[1], Tetsuo Himi[3]

Affiliations: [1]MD, Graduate Student, Department of Otolaryngology, Sapporo, Hokkaido, Japan; [2]MD, PhD, Associate Professor, Department of Otolaryngology, Sapporo, Hokkaido, Japan; [3]MD, PhD, Professor and Chairman, Department of Otolaryngology, Sapporo, Hokkaido, Japan.

Corresponding Author: Kenichi Takano, Department of Otolaryngology, Sapporo Medical University School of Medicine, S1W16, Chuo-ku, Sapporo 060-8543, Japan; Tel: +81-11-688-9655, Fax: +81-11-615-5405; Email: kent@sapmed.ac.jp

Received: 27 May 2014
Accepted: 25 June 2014
Published: 24 August 2014

How to cite this article

Jitsukawa S, Takano K, Abe A, Himi T. A case of chondrosarcoma of the temporal bone. Edorium J Otolaryngol 2014;1:7–10.

Article ID: 100002SJO042014

*********

doi:10.5348/004-2014-2-CR-2

## INTRODUCTION

Chondrosarcomas are malignant neoplasms composed of cartilage-producing cells, rarely encountered in the temporal bone. It is difficult to differentiate the tumor from other conditions based on clinical signs alone, although once a tumor reaches a considerable size, a variety of cranial symptoms are observed. Herein, we report a case of chondrosarcoma of the temporal bone presenting with facial palsy.

## CASE REPORT

An 80-year-old female was admitted to our hospital three years ago with right-sided slowly progressive facial nerve palsy. No cervical lymph node swellings were noted on examination, and otoscopy revealed bilateral chronic otitis media. Her hearing test revealed bilateral and profound sensorineural hearing loss presented since childhood. Laboratory tests revealed normal white blood cell counts. A computed tomography (CT) scan showed a destructive mass which has approximately 14 mm major axis length located in the right mastoid air cells (Figure 1), and further investigation with magnetic resonance

imaging (MRI) revealed a homogeneous mass extending into the venous sinus (Figure 1). These findings suggested that this mass was compressed or it invaded the vertical segment of the facial nerve.

The patient subsequently underwent mastoidectomy and tympanotomy to determine a definitive diagnosis and to remove the temporal bone lesion. During surgery, a jelly-like whitish mass was observed at the peripheral mastoid air cells the mass was slightly more than 10 mm in size and was easily remobable with a curette. Although we performed intraoperative pathological examination twice, cartilaginous metaplasia with no evidence of malignancy was observed. The mass had anteriorly extended to the lower tympanic cavity and the area of the venous sinus and posteriorly to the siphon caroticum. Oozing bleeding occurred when removing the mass from the venous sinus, which was controlled by the application of fibrin glue. As the intraoperative pathological diagnosis indicated no evidence of malignancy, resection was performed by piecemeal removal. The vertical segment of the facial nerve was anteriorly compressed by the mass and the nerve was thread-like (Figure 2). The facial nerve was eventually conserved.

Although the pathological diagnosis during surgery was of cartilaginous metaplasia, further pathological evaluation of the resected specimens resulted in a diagnosis of grade I chondrosarcoma. This was based on the observation composed of sheets and chords of uniform cells overlying a myxoid matrix and enlargement and hyperchromasia of the nuclei (Figure 3). We considered postoperative radiotherapy because the tumor was not removed en bloc and some previously reported cases underwent radiotherapy. However, postoperative therapy was not performed because of the low malignant potential and her disagreement. The patient showed no evidence of recurrence or metastasis after a one-year follow-up.

This report has been approved by the Institutional Review Board for human subjects at Sapporo Medical University.

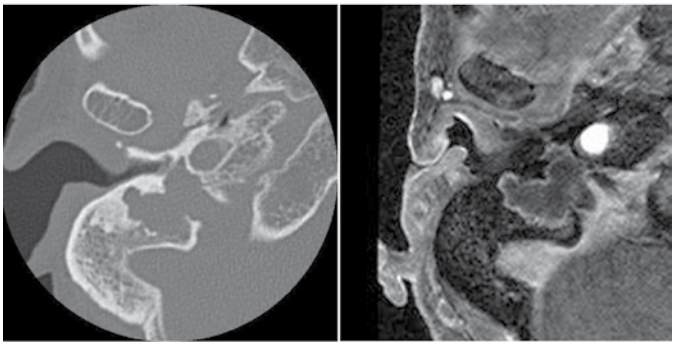

Figure 1: Axial computed tomography scan (left) showing the right mastoid destruction mass. An axial T1-weighted magnetic resonance imaging with gadolinium (right) showing a homogenous mass with rim enhancement and encroaching the venous sinus.

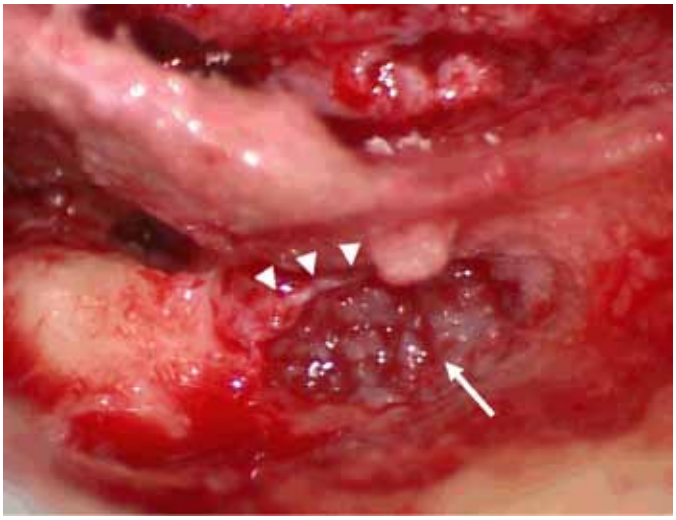

Figure 2: Intraoperative findings showing a jelly-like whitish mass with bone destruction (arrow) in the mastoid cavity, and anterior compression of the vertical segment of the facial nerve (arrow heads).

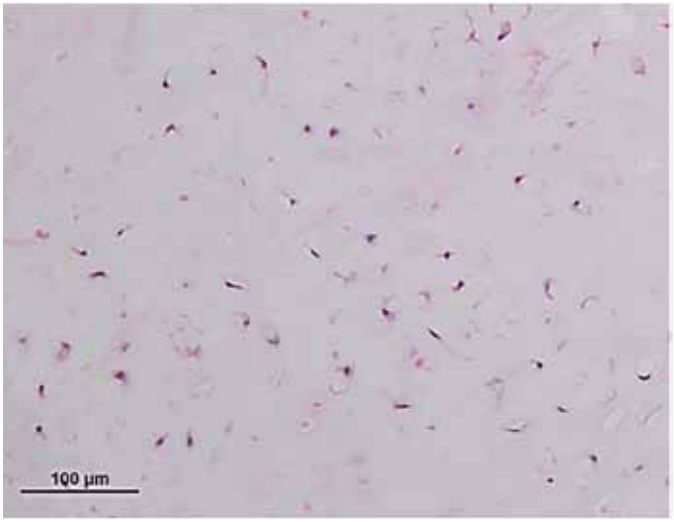


Figure 3: Histopathology of a well-differentiated (grade I) chondrosarcoma, characterized by the observation composed of sheets and chords of uniform cells overlying a myxoid matrix and enlargement and hyperchromasia of the nuclei. There is minimal cellular atypia, with only occasional mitotic figures.

## DISCUSSION

Chondrosarcoma is a malignant neoplasm of cartilage-forming-tissue and represents less than 1% of all intracranial tumors [1]. The petro-occipital and spheno-occipital areas, the sphenopetrosal synchondrosis, and a large part of the petrous portion of the temporal bone are sites in the mature skull that undergo endochondral development. It is considered that islands of residual endochondral cartilage may be present in these areas and

chondrosarcomas may develop from these chondrocytes. In this case, because the tumor was mainly located around the vertical segment of the facial nerve, it may have been derived from the petro-occipital area.

Chondrosarcomas of the temporal bone have several clinical presentations with symptoms that are closely related to the anatomical sites of destruction or compression. It has been reported that initial complaints include hearing loss, diplopia, dysphagia, vertigo, and facial palsy. Computed tomography and MRI scans have roles in the diagnosis and treatment of chondrosarcoma of the temporal bone. The typical CT appearance is of a destructive mass with scalloped erosive borders, occasionally with calcification. The MRI scan shows low to intermediate signal intensity on T1-weighted and high signal intensity on T2-weighted images, often with marked heterogeneous enhancement.

Chondrosarcomas are graded based on the degree of cellularity and nuclear atypia into grade I (well differentiated), grade II (moderately differentiated), and grade III (poorly differentiated). This grading system is useful because it reflects prognosis based on tumor biology distinct from the tumor location or stage of presentation. For example, a five-year survival rates of grades I, II, and III chondrosarcomas from all body sites have been reported as 90%, 81%, and 43%, respectively [2]. Although en bloc resection is the optimal treatment with curative potential, partial tumor excision can be an acceptable surgical treatment providing a superior prognosis [3, 4] with well-differentiated grade I chondrosarcomas, as in the present case. In addition, radiation therapy and/or chemotherapy have been considered to be futile, although proton-beam radiation has reportedly achieved good control of residual postoperative tumor growth [5].

## CONCLUSION

Chondrosarcoma of the temporal bone is extremely rare. However, it should be considered in the differential diagnosis of patients with slowly progressive osseous tumors. It is important to obtain an early diagnosis and resection before the tumor becomes extensive.

*********

### Acknowledgements
We would like to thank our patient and her family for allowing us to publish her medical condition.

### Author Contributions
Jitsukawa Sumito – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Kenichi Takano – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Ayumi Abe – nalysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Tetsuo Himi – nalysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

### Guarantor
The corresponding author is the guarantor of submission.

### Conflict of Interest
Authors declare no conflict of interest.



## REFERENCES

1. Cianfriglia F, Pompili A, Occhipinti E. Intracranial malignant cartilaginous tumours. Report of two cases and review of literature. Acta Neurochir (Wien) 1978;45(1-2):163–75.
2. Evans HL, Ayala AG, Romsdahl MM. Prognostic factors in chondrosarcoma of bone: A clinicopathologic analysis with emphasis on histologic grading. Cancer 1977;40(2):818–31.
3. Burkey BB, Hoffman HT, Baker SR, Thornton AF, McClatchey KD. Chondrosarcoma of the head and neck. Laryngoscope 1990;100(12):1301–5.
4. Rapidis AD, Archondakis G, Anteriotis D, Skouteris CA. Chondrosarcomas of the skull base: Review of the literature and report of two cases. J Craniomaxillofac Surg 1997;25(6):322–7.
5. Munzenrider JE, Liebsch NJ. Proton therapy for tumors of the skull base. Strahlenther Onkol 1999;175 Suppl 2:57–63.