



Case Report

Promising Outcome of Radiation Therapy for Chondroblastoma of Temporal Bone in Childhood: A Case Report

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Abstract

Chondroblastoma is a rare cartilaginous benign bone tumor. Chondroblastoma in the temporal bone is also quite rare. Total excision is the main treatment. Data regarding tumor response to radiation therapy (RT) is insufficient. We describe a case of chondroblastoma that was treated with RT following subtotal tumor resection. In this case, the patient was a 14-year-old male who presented with a three-month history of ear fullness and hearing loss in his right ear. Magnetic resonance imaging revealed a mass partly filling the right external auditory canal and the inferior part of the middle ear. Histopathological findings indicated chondroblastoma. Subtotal tumor resection was performed due to risk of complications. RT was planned upon the growth of the tumor during follow-up. Treatment with subtotal resection and postoperative RT has been successful and the patient had no recurrence in the course of the 12-year follow-up. In chondroblastoma, complete surgical resection is still the gold standard. But the success of subtotal resection followed by adjuvant RT should also be kept in mind for cases where the total excision would pose high morbidity.

Keywords: Chondroblastoma, hearing loss, temporal bone, radiation therapy, pediatric otorhinolaryngology, case report

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Introduction

Chondroblastoma is a tumor rarely seen in temporal bone in childhood. It accounts for less than 1% of primary bone tumors (1). It is a benign cartilaginous tumor that often affects the epiphysis of long bones

and is seen in the second to third decades of life (2). Chondroblastoma usually originates from the squamous part of the temporal bone (3). Clinical picture shows a painful mass with hearing loss and tinnitus. While total excision is the mainstay of the treatment, there is insufficient data

regarding tumor response to radiation therapy (RT). In this case report, we present our experience with the long-term follow-up of a teenager with chondroblastoma who had undergone RT following subtotal tumor resection.

Case Presentation

A fourteen-year-old male was admitted with ear fullness and hearing loss for three months in his right ear in 2008. A fragile and hemorrhagic mass filling the right external auditory canal (EAC) was observed in examination. The tympanic membrane could not be evaluated. No pathology was observed in the left ear. Pure tone audiometry (PTA) revealed slight conductive hearing loss with 3-frequency average of air and bone conduction thresholds of 30 and 5 dBHL, respectively, in the right ear. Air and bone conduction thresholds were 5 for each dBHL on the left. Non-contrast computerized tomography (CT) of the temporal bone showed an expansile and destructive lesion in right temporal bone (Figure 1a). Magnetic resonance imaging (MRI) of the temporal bone showed a mass partly filling the right EAC and the inferior part of the middle ear. Longest axis in the superior to inferior direction was 18 mm. The mass was irregularly shaped. It was hypointense in T1W, isointense in fat-saturated T2W images and showed contrast enhancement after post-contrast fat-saturated T1W images (Figure 1b). Following a biopsy, intact canal wall mastoidectomy was performed with posterior tympanotomy through the facial recess. It was noted that the reddish, fragile and bloody mass in the inferior part of the middle ear had remodeled the inferior part of the posterior

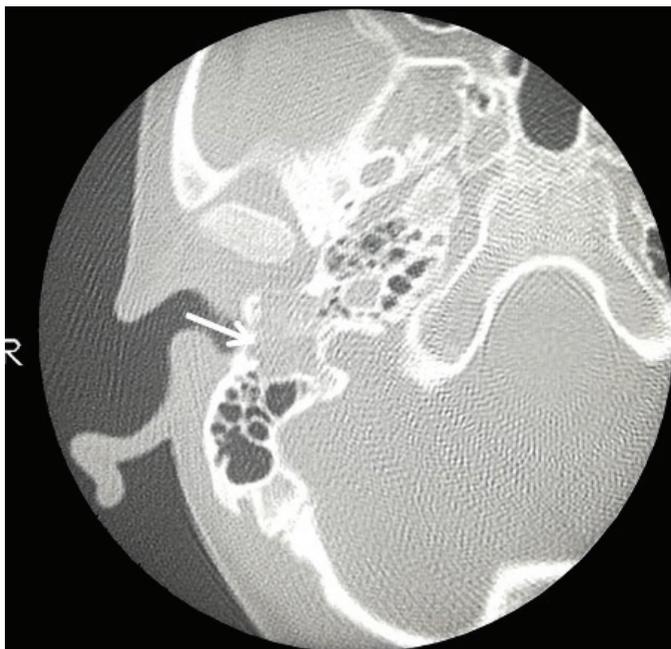


Figure 1a. Axial thin-section non-contrast CT image of temporal bone showing destructive and expansile lesion in right temporal bone (arrow)

CT: Computed tomography

wall of the EAC and reached the mastoid cavity. The fallopian canal was eroded in the middle part of the mastoid segment of the facial nerve. The ossicles were tumor-free. Upon obtaining the frozen section result as a benign lesion with no further detail, we tended to act based on the permanent histopathology report. Hearing levels could be preserved after the biopsy. The fact that permanent histopathologic examination confirmed the diagnosis of chondroblastoma led us to aim at intended-total excision of the lesion. During the main operation, the EAC was lowered in order to improve tumor exposure. The tumor was removed via canal wall down (CWD) mastoidectomy to the maximum extent possible and some residue was left behind the temporomandibular joint (TMJ) and on the jugular bulb in order to prevent inevitable injury. Meatoplasty was performed. The patient was discharged without any complications. The hearing level remained almost at the same level. Histopathologic examination of the main operation specimen revealed the same diagnosis (Figure 2). A wait-and-see policy was followed in order to observe the fate of the residual tumor. In a follow-up examination two years after the operation, it was noted that the entrance

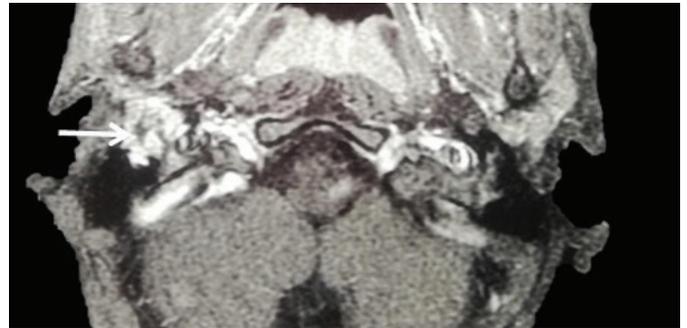


Figure 1b. Axial post contrast fat-saturated T1W image shows tumor in the right temporal bone (arrow)

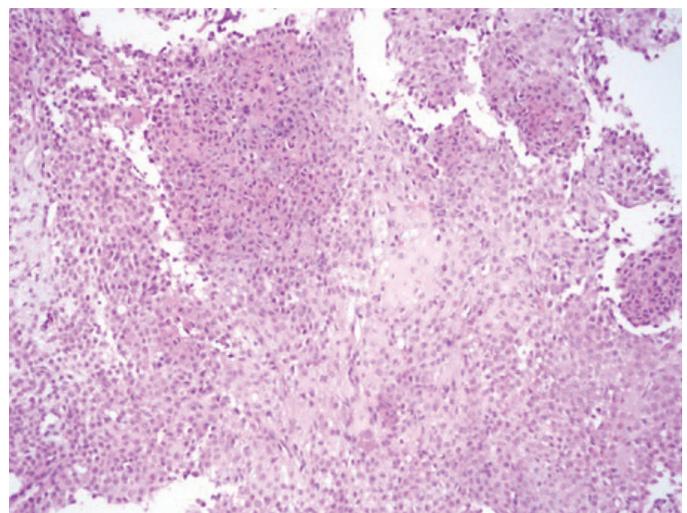


Figure 2. Sheet-like proliferation of chondroblasts with extracellular matrix. Scattered osteoclast-like giant cells are present and focal mineralization of matrix surrounding single cells can be seen (H&E 200X)

of the CWD mastoidectomy cavity was narrowed inferiorly by the tumor. PTA revealed increased conductive hearing loss in the right ear (Right: 45/10 dBHL, Left: 5/5 dBHL). Follow-up MRI showed increased dimensions of the residual tumor. It extended medially towards the vertical segment of the internal carotid artery and anteriorly towards the TMJ (Figure 3). Considering the morbidity of the required surgery led us direct the child to RT. Sixty Gy intensity modulated image guided RT was delivered in 30 fractions with 2 Gy fraction dose for six weeks (Figure 4). Grade 2 peripheral facial paresis and synkinesis developed on the right side a few months later which healed spontaneously in the next months. He developed total hearing loss in the right ear due to RT (PTA: Right air/bone: 110+/67+dBHL, Left air/bone: 10/5 dBHL).

The patient has been on follow-up for 12 years and does not have any complaints. A very recent MRI showed no evident contrast enhancement in the temporal bone (Figure 5). He is still on our yearly follow-up program.

Discussion

Chondroblastoma is a histologically benign but clinically aggressive tumor (4). Its etiology is unknown. It is caused by immature chondroblasts in the epiphysis of long bones (5). While the disease usually involves long bones and especially the femur and humerus, the involvement of flat bones such as the skull is extremely rare (<2%) (6). The temporal bone is the most frequently involved cranial bone. The squamous part is usually involved due to its cartilaginous origin (3).

In a systematic review of 100 cases, the mean age at diagnosis of skull and facial bone chondroblastoma was 42.3 years (2). In the same study, 4 out of 100 cases were diagnosed under the age of 18. There also was a slight male predominance (ratio, 1.3:1). Likewise, our patient is a 14-year-old male.

Hearing loss (especially the conductive type), tinnitus, cranial nerve involvement, facial swelling, and otalgia are the

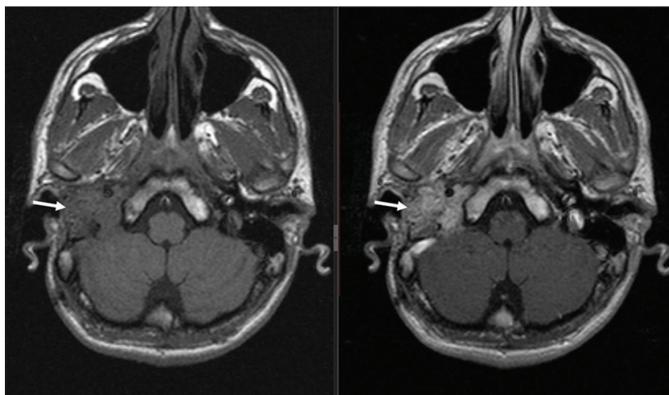


Figure 3. Axial pre- and post-contrast T1W images show increased dimension of the residual tumor (arrow)

most common symptoms (4, 6, 7). TMJ dysfunction can also be seen. Diagnosing can be difficult. Chondroblastoma can be mistaken for the granulation tissue, giant cell reparative granulomas, giant cell tumors and osteosarcoma (3).

Radiological imaging is important for diagnosis. CT imaging typically shows internal calcification associated with intra-tumor calcium and hemosiderin deposition and rarely expansile intraosseous soft tissue mass with increased intravenous contrast enhancement (6).

The amount and morphologic pattern of calcification are variable on CT (8).

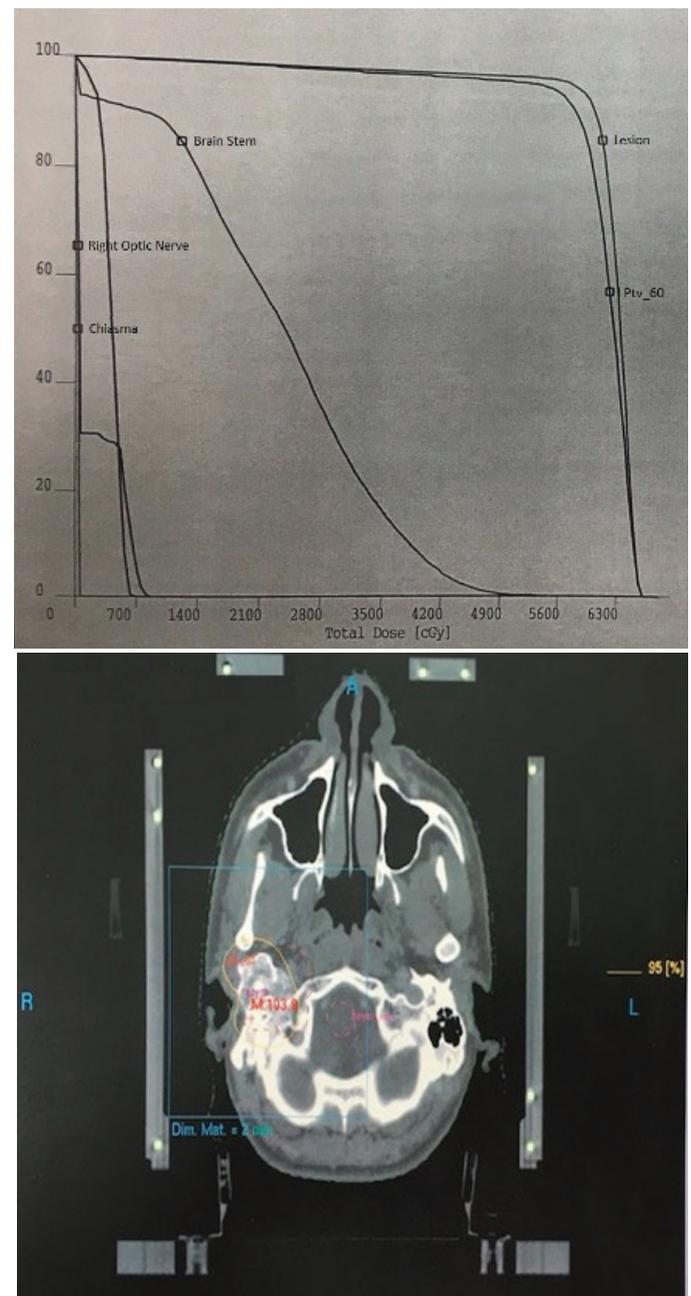


Figure 4. Dose volume histogram and digitally reconstructed radiography image of the planning target volume of the lesion

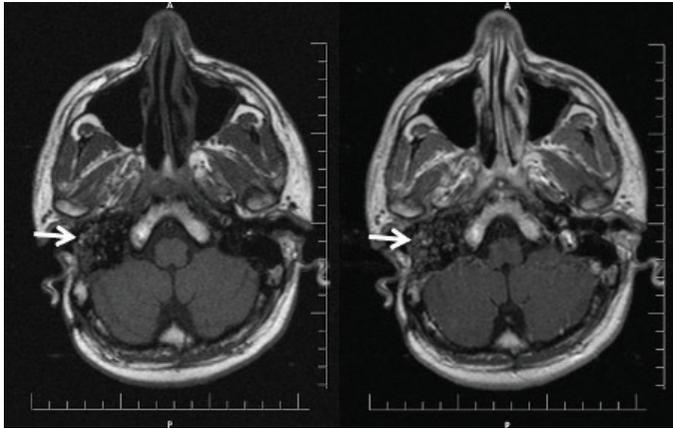


Figure 5. Axial pre- and post-contrast T1W images show no significant tumor enhancement (arrow)

MRI typically shows hypo-intermediate signal in T1 imaging and high signal in T2 due to bleeding into the mass. The cause of heterogeneity in T2 images is intense cellularity and dense vascular fibrous tissue. Post-gadolinium increase, which is a prominent hyperintensity component, shows heterogeneity in T1W imaging. In addition, MRI better describes intracranial/other soft tissue involvement such as dura and brain (6). The exact diagnosis is made pathologically. It is very difficult to diagnose with frozen section due to its similarity to other benign and malignant giant-cell neoplasms (2, 3). Diagnostic histopathological findings are the presence of chondroblast, osteoclastic-like giant cells, and chondromyxoid stroma around neoplastic cells. Chicken wire calcification is typical. These tumors secrete s-100 and vimentin (6).

The standard procedure for chondroblastoma is complete or wide/en bloc surgical resection. Complete removal of the tumor is recommended because of high local recurrence rate after incomplete surgery which is around 25% (4). However, complete tumor removal may not be possible in some circumstances, i.e., tumor proximity to vital structures or involvement of these structures. Also, less aggressive surgeries can be considered to preserve the quality of life in benign tumors like chondroblastoma. In these cases, adjuvant RT can be given. There is no clear information in the literature, possibly due to small sample size, about the use of adjuvant RT in case of residual disease. In fact, it is not recommended to be given due to malignant transformation risk (4). In a review conducted in 2020, only 11 of the 100 cases had a history of RT. Proton therapy combination with photon therapy were given in one patient and targeted biologic therapy using denosumab was given in one other patient (2). In a series of three cases in 2020, chondroblastoma were treated with a functional surgical approach. One case had recurrence after 125 months of follow-up and was treated with RT successfully (gamma knife radiosurgery; maximum

dose of 23 Gy). No recurrence occurred in the following 100-month period (total follow-up period was 225 months) (7). However, in our case, total removal was not possible and control MRI showed residual tumor growth thus warranting RT as the best alternative treatment modality with many reports showing its effectiveness in tumor growth control (9).

A wait-and-see policy was adopted because of the possible side effects of RT, but because the patient came from countryside, close follow-up was recommended, but could not be done. This case is also important in terms of showing the importance of close follow-up in patients with residual tumors.

In this case report, we presented the long-term outcome of a young male with temporal bone chondroblastoma who received RT after the regrowth of the subtotally resected tumor. Clinical results favor RT, although this does not come to mean that RT can replace the surgical mode of treatment. Surgical treatment is still favorable wherever total excision is feasible.

Conclusion

Chondroblastoma is a rare tumoral formation seen in the mastoid bone in the first few decades. Although complete surgical resection is still the gold standard today, the success of subtotal resection followed by adjuvant RT should also be kept in mind for cases where total excision would pose high morbidity. It should be considered that hearing was sacrificed to RT and RT carries a potential risk of malignancy although tumor control was achieved for a long time in our case.

Informed Consent: Written informed consent for the publication of this report was obtained from the patient.

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Authorship Contributions

Surgical and Medical Practices: B.S., E.Ç., Concept: B.S., Design: B.S., Data Collection and/or Processing: F.C.A.Ö., E.Ç., U.B., M.B., Analysis and/or Interpretation: F.C.A.Ö., U.B., M.B., Literature Search: F.C.A.Ö., Writing: F.C.A.Ö., B.S.

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Main Points

- Chondroblastoma is a tumor rarely seen in temporal bone in childhood.
- Total surgical resection is the main treatment.
- Although complete surgical resection is still the gold standard today, the success of subtotal resection followed by adjuvant radiation therapy should also be kept in mind for cases where the total excision would pose high morbidity.

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