Chondroblastoma of the Temporal Base With High Mitotic Activity

—Case Report—

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Abstract

A 24-year-old man presented with a rare chondroblastoma of the temporal base manifesting as local pain accompanied by difficulty in opening the mouth. Gross total removal was achieved at initial surgery, but the tumor demonstrated rapid and destructive regrowth from a very small residual volume without definite histological malignant transformation. Growth activity estimated by MIB-1 staining increased spontaneously from 2.5% at the initial operation to 18.7% at recurrence. Further extensive radical tumor removal by surgeons from multiple disciplines was performed. The patient has been free of recurrence for 3 years without radiotherapy. Chondroblastoma of the temporal bone is widely accepted as a benign tumor and regrowth after gross total removal is very rare. However, some cases of chondroblastoma have potentially high mitotic activity.

Key words: chondroblastoma, temporal bone, regrowth, MIB-1

Introduction

Chondroblastoma is a rare tumor which accounts for less than 1% of all bone tumors and which most commonly occurs in the epiphyses of the long bones.^{4,27)} Although chondroblastoma may occur at various other locations, its appearance in the cranial bones is extremely unusual. We report here a case of chondroblastoma of the temporal base, which showed very rapid and destructive regrowth after initial surgery.

Case Report

A 24-year-old man presented with arthralgia of the right temporomandibular joint and painful swelling over the right temporal region associated with difficulty in opening the mouth. These symptoms progressed for 18 months with enlargement of the zygoma.

On admission, neurological examination showed no abnormalities, and routine blood tests were nor-

mal including serum alkaline phosphatase level. Physical examination disclosed that mouth opening was limited to 3 cm. Computed tomography revealed an extracerebral mass with a few surrounding calcified foci that had destroyed the temporal bone including the zygomatic bone. The tumor had expanded into the pterygoid fossa under the greater wing of the sphenoid bone. Magnetic resonance (MR) imaging demonstrated the mass as isointensity on the T₁-weighted images and mixed intensity on the T₂-weighted images (Fig. 1). Gadolinium administration caused homogeneous enhancement adjacent to the dura mater of the middle fossa, which showed focal thickening (Fig. 2 left). Right external carotid angiography demonstrated that the middle meningeal, accessory meningeal, and superficial temporal arteries were feeding the tumor, whereas other vessels did not show any additional arterial supplies to the tumor. Both gallium-67 citrate scintigraphy and technetium-99m-methylene diphosphonate scintigraphy showed very high accumulations in the tumor. The diagnosis was benign bone tumor originating in the temporal base.

Surgical plans were designed by specialists in

neurosurgery, otolaryngosurgery, and oral surgery. The feeding arteries were embolized with particles of polyvinyl alcohol before surgical removal. A right frontotemporal curved skin incision exposed the tumor in the right temporal base (Fig. 3). After craniotomy of the apparently normal surrounding temporal bone and removal of the zygomatic bone, the tumor was debulked. The capsule was bluntly

separated from the parotid gland and the pterygopalatine muscles. The tumor was impossible to separate from the dura of the middle fossa, so the affected dura mater was removed as thoroughly as possible. The edge of the thickened dura mater in the deep region of the middle fossa was coagulated with bipolar electrocautery. After dural plasty with autograft, dental and oral surgeons performed arthroplasty of the mandibular bone.

Histological examination of the resected tumor revealed round to ovoid tumor cells with varying

Fig. 1 *left:* Axial T₁-weighted (repetition time [TR] 437 msec, echo time [TE] 13 msec) magnetic resonance (MR) image showing a mass isointense to the gray matter in the right temporal bone. *center:* Axial T₂-weighted (TR 3000 msec, TE 80 msec) MR image showing varying mixed intensity of the mass. *right:* Computed tomography scan with bone window showing an extracerebral mass with spotty calcifications and peripheral “eggshell” ossification.

Fig. 3 Intraoperative photograph showing the tumor in the right temporal base (arrowheads). The tumor had invaded part of the zygomatic bone (arrow) and part of the temporal bone.

Fig. 2 *left:* Coronal T₁-weighted magnetic resonance (MR) image with gadolinium taken just before the initial operation showing the enhanced mass adjacent to the dura mater of the middle fossa and expanding into the pterygoid fossa. *center left:* MR image with gadolinium taken at 8 months after the initial operation showing no residual mass except for the thickened dura mater. *center right:* MR image with gadolinium taken just before the second operation showing regrowth of the tumor at the same position as the lesion in the initial image. *right:* MR image with gadolinium modified by fat suppression taken at 3 years after the second operation showing no regrowth of the tumor.

Fig. 4 Photomicrograph of the tumor specimen showing round to ovoid tumor cells with amounts of eosinophilic cytoplasm and focal chondroid intercellular matrix containing lace-like deposits of calcium granules. Hematoxylin and eosin stain, $\times 100$.

size and amounts of eosinophilic cytoplasm. The cells showed diffuse proliferation amid multinucleated giant cells. Focal chondroid intercellular matrix was present containing lace-like deposits of calcium granules (Fig. 4). Focal collection of many microvessels was observed. Immunohistochemical staining with MIB-1 showed a positive staining of 2.5% of cells. Immunohistochemical studies also revealed positive staining of the mononuclear cells for S-100 protein. The histological diagnosis was chondroblastoma. Invasive findings of the outer layer of the dura mater with focally increased vascularity were observed. Based on the intraoperative findings and postoperative MR imaging, the tumor was removed leaving possible residual tumor at the thickened dura mater (Fig. 2 center left).

Two years after gross total removal of the tumor, the patient presented with progressive headache in the right temple. Follow-up MR imaging showed regrowth of a mass of 7 cm diameter located in the skull base region inferior to the right zygomatic bone (Fig. 2 center right). In the second operation, the tumor was exposed through a wider surgical field than the initial operation. The tumor had adhered to the dura mater in the middle fossa and destroyed the temporal half of the skull base and the lateral wall of the orbit. Further radical dissection was performed including the petrosal bone, muscles, soft tissues, and dura mater of the temporal base. Dural plasty was performed after complete removal of the dura mater with tumor invasion. The tumor could be separated from the parotid gland and muscles. A large fat graft harvested from the abdominal wall

was used to obliterate the defect.

The histological diagnosis was chondroblastoma without definite malignancy, identical to the previous diagnosis. However, immunohistochemical staining with MIB-1 showed an increase from 2.5% to 18.7% of cells. Follow-up MR imaging 3 years after the second surgery showed no recurrence of the tumor (Fig. 2 right) and the patient has remained asymptomatic until now.

Discussion

Chondroblastoma of the skull is very rare, and is most often located in the temporal bone. A review of the 56 reported cases of chondroblastoma of the temporal bone including the present case shows that patients have a median age of 44 years and that the male to female ratio is 1.6:1.^{1-13,15-18,20,22-26,28} The most common symptoms are hearing loss, otalgia, and tinnitus due to involvement of the posterior half of the temporal bone and the inner ear.

Histological findings of typical chondroblastoma are mononuclear tumor cells of varying sizes, multinucleated osteoclast-like giant cells distributed throughout the tumor cells, and pericellular fine linear calcification, often showing the characteristic chicken wire pattern. Chondroblastoma can be differentiated from chondroma and chondrosarcoma by these findings. Additionally, immunostaining for S-100 protein is helpful in differentiating chondroblastoma from giant cell tumors. Specifically, the mononuclear tumor cells in chondroblastoma are focally positive for S-100 protein, whereas all giant cell tumor cells are negative.^{11,18}

A high rate of regrowth is reported after only curettage,¹² whereas regrowth after gross total removal is apparently rare. However, it should be kept in mind that the follow-up period was less than 2 years in most previously reported cases. These facts indicate that chondroblastoma is essentially benign, even though potentially malignant histological features such as mitotic activity, necrosis and vascular invasion have been observed in some cases of chondroblastoma.¹⁴ Nevertheless, in spite of limited local aggressiveness during growth, the biological behavior of these tumors is benign.

At the time of the initial operation, our patient was treated under a histological diagnosis of chondroblastoma without definite malignant features. However, the tumor showed rapid and aggressive regrowth, and the MIB-1-positive index at the second operation was very high, suggesting that residual chondroblastoma can grow spontaneously without histological malignant transformation after surgery, and that the tumor can enlarge rapidly even

if the residual volume is very small. In the present case, clinical testing showed hypervascularity and dural invasion, which seem to be associated with the unusually rapid progression of the tumor. However, hypervascularity of the tumor was found in three of the seven cases in which angiographic findings were reported, but without significant regrowth of the tumor.^{4,8,18,20,23,24,26} Furthermore, the present case, together with 15 previously reported cases which included detailed operative descriptions, showed dural adhesion in 13 cases (81%) and dural invasion in three cases (19%).^{1,3,4,8,12,15,17,18,20,23-26,28} Thus, these features are not necessarily rare in this tumor. In several interesting cases, non-cranial "malignant chondroblastoma" showed increased mitotic activity and distant metastasis with typical features of chondroblastoma that were histologically similar to those found in our patient.^{19,21} Complete tumor removal may be essential to prevent recurrence in those cases of chondroblastoma that show potentially high mitotic activity. Radiotherapy was reported to be effective in some cases, but the need for this treatment is not widely accepted.^{9,23} In the case of our patient, we chose not to perform postoperative irradiation because of the possibility of malignant transformation, and therefore cannot provide further evidence about its efficacy.

In conclusion, the present case of chondroblastoma of the temporal base showed high mitotic activity. The potential for rapid regrowth after gross total removal of such a tumor indicates that complete surgical removal with the cooperation of a neurosurgeon, an otolaryngologic surgeon, an oral surgeon, and other specialists as deemed necessary is essential to achieve a good outcome for patients with chondroblastoma.

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