Maxillary Ameloblastoma with Intracranial Invasion

—Case Report—

Kazuei SATO, Satoshi SUDO*, Yasuhiko FUKUYA*, and Hideo SAKUMA**

Departments of Neurosurgery, *Plastic Surgery, and **Pathology, Ota Atami Hospital, Koriyama, Fukushima

Abstract

A 79-year-old male presented with recurrent maxillary ameloblastoma with intracranial invasion into the left orbit, previously histologically diagnosed as benign ameloblastoma. Skull x-ray films and computed tomography showed the multicystic mass had destroyed the skull base. The tumor was nearly completely removed. However, microscopic examination revealed residual tumor cells around the left optic nerve. Histological examination found no malignant transformation in the tumor specimen. Aggressive complete removal of maxillary ameloblastoma should be attempted even in cases of intracranial invasion.

Key words: ameloblastoma, intracranial invasion, computed tomography, successful removal

Introduction

Ameloblastoma is a relatively rare and histologically benign tumor derived from the odontogenic apparatus. The recurrence rate is high because the absence of tumor capsule allows infiltration into the surrounding tissues.3,12 Eighty percent of ameloblastomas originate in the mandible, and 20% in the maxilla.3,15,17 Maxillary ameloblastomas are inherently more difficult to treat and are considered more aggressive than the mandibular type.3 Maxillary ameloblastoma may become locally aggressive and potentially lethal when the tumor has invaded the skull base or given rise to distant metastases.5,11,18

We describe a rare case of maxillary ameloblastoma involving the left anterior and middle cranial fossa and the dura, and discuss the treatment.

Case Report

A 79-year-old male was referred to our clinic on March 20, 1992, because of a growing orbital mass. He was first admitted to another hospital in 1981, aged 68 years, because of swelling of the left maxillary region, when a radical maxillectomy was performed (the first operation). Histological examination revealed benign ameloblastoma. Over the next 9 years, he was asymptomatic until exophthalmos and paresthesia in the distribution of the maxillary branch of the left trigeminal nerve developed. Computed tomography (CT) showed a mass in the inferior and posterior region of the orbit. He was referred to the Department of Plastic Surgery of our hospital in June, 1990. In July, a tumor in the inferior region of the orbit was removed in a second operation performed with an otologist. A third operation in collaboration with an ophthalmologist involved almost total removal of the recurrent tumor with orbital exenteration in June, 1991. The defective skin area was repaired by pedicle skin graft. Histological examination showed ameloblastoma without malignant transformation. He later noted painful progressive swelling of a mass in the left orbit, and was admitted to our hospital on April, 1992.

The diffuse swelling involving the left maxillary region and the orbit was elastic hard. The overlying skin was thin and reddish. He complained of mild frontal headache. Neurological examination revealed decreased sensation to touch and pin-prick in the distribution of the ophthalmic and maxillary branches of the left trigeminal nerve, and facial nerve paresis on the left side. A plain skull x-ray film showed indistinctly outlined radiolucent lesions in the left frontal basal and anterior temporal basal areas. A compact bone was seen in the upper

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lateral orbital margin (Fig. 1). CT revealed a multicystic round mass in the left maxillar, orbital, and frontotemporal region destroying the skull. Postcontrast CT showed the tumor was partially enhanced (Fig. 2 left). Left carotid angiography showed faint staining of the tumor fed by the lingual artery, facial artery, palatine branches of the maxillar artery, middle meningeal artery, and ophthalmic artery, and superomedial displacement of the middle cerebral arteries. The above findings suggested that recurrent maxillary ameloblastoma had extended into the skull with dural involvement.

The feeding vessels of the tumor, the lingual artery, facial artery, and palatine branches of the maxillar artery, were embolized with particles of Gelfoam® 3 days before the operation. A skin incision was made along the old scar of the skin graft at the left maxillar orbital region and the tumor mass was dissected from the nasal wall (Fig. 3). Bleeding from the tumor was easily controlled. A hair line skin incision was made for frontotemporal craniotomy. The tumor was a multicystic mass containing old bloody fluid surrounded by a thick wall, and invaded the intracranial space through the bone defect around the superior orbital fissure. The extradural tumor was excised immediately at the optic canal. Gross inspection revealed no tumor invasion around the optic nerve. The intradural tumor, covered with the inner layer of the dura mater and adhering to the temporal lobe, was excised at the margin of the tumor after dissection from the temporal lobe. The tumor specimen was $6 \times 5 \times 7.5$ cm. The part of the skull attached to the tumor was drilled off or rongeured out. The dural defect was replaced with lyophilized dura and fatty tissue, and then sealed with fibrin glue. Skin graft was used to repair the defective nasal mucous membrane and seal the frontal sinus. Pedicle flaps of the superficial temporal muscle and bisected frontal bone flaps were used in the reconstruction of the skull base.

Histological examination showed a moderately lateral orbital margin (Fig. 1). CT revealed a multicystic round mass in the left maxillar, orbital, and frontotemporal region destroying the skull. Postcontrast CT showed the tumor was partially enhanced (Fig. 2 left). Left carotid angiography showed faint staining of the tumor fed by the lingual artery, facial artery, palatine branches of the maxillar artery, middle meningeal artery, and ophthalmic artery, and superomedial displacement of the middle cerebral arteries. The above findings suggested that recurrent maxillary ameloblastoma had extended into the skull with dural involvement.

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Histological examination showed a moderately
differentiated follicular ameloblastoma. The outer layer of the dura mater from around the optic nerve was invaded by tumor cells (Fig. 4).

His postoperative course was uneventful, and no radiotherapy nor chemotherapy were given. He was discharged on the 23rd postoperative day. Postoperative CT revealed no residual tumor mass (Fig. 2 right).

**Discussion**

This case illustrates two distinct features: maxillary ameloblastoma can be locally aggressive and incurable, and should be extensively excised even if the tumor invades the intracranial space. Robinson\(^{15}\) and Small and Waldron\(^{16}\) considered ameloblastoma to be benign, but despite the lack of cellular malignant characteristics, these tumors tend to grow continually and invade the surrounding tissue, because of the composition of proliferating odontogenic epithelium in a connective tissue stroma. Intracranial ameloblastoma has only been sporadically reported.\(^{2,5,6,10,12,18}\) The tumor can spread through two routes, direct invasion or distant metastasis. A few ameloblastoma (malignant ameloblastoma) cases have metastasized to the skull bone.\(^{12,14}\) Approximately 2% of ameloblastoma is malignant, with the diagnosis based on histological atypia and distant metastases.

Symptoms attributable to maxillary ameloblastoma first appear at a mean age of 32.7 years (range

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**Table 1** Cases of intracranial invasion by maxillary ameloblastoma

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age*/ Sex</th>
<th>Site of invasion**</th>
<th>Treatment***</th>
<th>Outcome, disease duration (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kyriazis <em>et al.</em> (1971)(^{13})</td>
<td>73/F</td>
<td>orbit (5), MCF (8)</td>
<td>partial maxillectomy (0), wide local excision (3, 5)</td>
<td>died, 8</td>
</tr>
<tr>
<td>Weiss <em>et al.</em> (1985)(^{10})</td>
<td>72/M</td>
<td>orbit (5), MCF (6)</td>
<td>wide local excision (1), partial maxillectomy (2), radiotherapy (5), craniotomy and partial removal (6)</td>
<td>died, 7</td>
</tr>
<tr>
<td>Bredenkamp <em>et al.</em> (1989)(^{2})</td>
<td>53/M</td>
<td>orbit (0), MCF (0), maxilla (0), orbit (2), MCF (14)</td>
<td>64-Gy radiotherapy (0)</td>
<td>alive, 1</td>
</tr>
<tr>
<td>15/M</td>
<td></td>
<td></td>
<td>enucleation (0), radical maxillectomy (2), orbital exenteration (2), 65-Gy radiotherapy (4), chemotherapy (8), partial removal (8–14)</td>
<td>died, 15</td>
</tr>
<tr>
<td>Present case</td>
<td>68/M</td>
<td>orbit (9), MCF and ACF (11)</td>
<td>radical maxillectomy (2), debulking (9), orbital exenteration (10), craniotomy and debulking (11)</td>
<td>alive, 12</td>
</tr>
</tbody>
</table>

*Age at discovery. **Number in parentheses indicates years until each anatomic site became involved. ***Number in parentheses indicates years until treatment was undertaken. ACF: anterior cranial fossa, MCF: middle cranial fossa.
Maxillary ameloblastoma has a 2.4:1 male to female ratio. Five cases of maxillary ameloblastoma involving the middle cranial fossa have been reported (including this case) (Table 1). The mean period from onset to intracranial invasion was 7.8 years. The orbit was the first vital structure involved, followed by the skull base, parasellar region, and middle cranial fossa. Intracranial invasion is frequently followed by gradual painful swelling of the affected site, visual disturbance, disturbed ocular movement, and epistaxis. Symptoms attributable to increased intracranial pressure are not common.

Patients with intracranial involvement have a poor prognosis, mainly because complete surgical excision is impossible when the tumor involves the central nervous system. Even radical surgery to remove ample margins of healthy bone, which will include areas of microscopic spread, cannot completely remove the tumor. Histological examination of removed tumors has shown microinvasion into the intertrabecular spaces of the cancellous skull bone with pseudopods of tumors. In our patient, bones around the osteolysis were rongeured out or drilled away, and the dural margins around the removed tumor were microscopically tumor-free. However, microscopic tumor cell invasion was seen around the extradurally resected optic nerve.

Chemotherapy is not effective for reducing tumor size or palliation of the disease. Radiotherapy and cryotherapy achieve some palliation with decreased tumor size and pain, but probably offer no chance of cure. Therefore, radiotherapy should only be used as a primary treatment when surgery is impossible because of the patient's age or condition. Surgical removal should be repeated if possible. Our 79-year-old patient again tolerated the surgical procedure very well, so no additional treatment was given.

We consider that removal of maxillary ameloblastoma, even if not complete, achieves a better prognosis when the patient is treated in collaboration with plastic surgeons, otologists, and ophthalmologists.

References


Address reprint requests to: K. Sato, M.D., Department of Neurosurgery, Ota Atami Hospital, 5-240 Atami, Atami-machi, Koriyama, Fukushima 963-13, Japan.