Chondrosarcoma of the Maxilla Extending to the Pterygomandibular Space: A Case Report and Review of the Literature

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Abstract Chondrosarcomas are rare tumors of which approximately 10 % are found in the maxillofacial region. They range from a well-differentiated growth resembling a benign cartilage tumor to a high-grade malignancy with aggressive local behaviour and the potential to metastasize. We report a case of high-grade chondrosarcoma of the maxilla extending to the pterygomandibular space which was treated with radical surgical excision without neck dissection and postoperative radiotherapy. The diagnosis was a conventional type of chondrosarcoma, Grade III. The postoperative course was uneventful, and the patient has had no recurrence in the 15-year follow-up period.

Keywords Chondrosarcoma · Maxilla ·
Pterygomandibular space

Introduction

Chondrosarcoma occurring in the head and neck region is relatively rare, the commonly affected sites being the maxilla, the mandible and the sino-nasal region [1–3]. Nearly 60 % of cases occur in the anterior alveolar region of the maxilla [2, 4, 5]. Its occurrence in the maxilla extending to the pterygomandibular space is extremely

rare. Painless swelling is the most common sign of chondrosarcoma of the maxilla and other symptoms are pain, paresthesia, trismus, and loosening of the teeth [2, 4, 5]. Chondrosarcomas are classified as conventional chondrosarcomas, clear cell chondrosarcomas, myxoid chondrosarcomas, dedifferentiated chondrosarcomas, and mesenchymal chondrosarcomas [6]. A lobuled growth pattern of hyaline cartilage with hypercellularity, nuclear pleomorphism, and binucleation is usually determined in histologic evaluation of this tumor [7]. In this paper, a case of chondrosarcoma arising from the maxilla extending to the pterygomandibular space is reported.

Case Report

The patient was a 21-year-old Japanese woman was referred by her general dentist to an oral and maxillofacial surgeon for evaluation of spontaneous pain and swelling of the gingiva after an extraction of the upper right third molar. There were no particular abnormalities in her general condition. The patient's past history and family history were also not contributory. The patient reported no medical problems or prior surgeries. She had a symmetric face, with mild trismus. Intraoral examination revealed right-sided oropharyngeal swelling with normal mucosa on the mass. On intraoral palpation, a relatively well-defined and elastic hard mass, approximately 3 cm in dimension, slight pressure pain was noted in the right maxillary tuberosity. Neck examination was normal. Computed tomography revealed a relatively well-defined mass of heterogeneous density measuring about 3 cm in diameter, situated in the maxillary tuberosity extending to anterior part of the right pterygomandibular space. The tumor extended from the lateral plate of the pterygoid process to the anterior border

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Fig. 1 **a** 3D-computed tomogram showing an expansive mass (*green*) ▶ of the maxilla extending to the pterygomandibular space. **b** Axial CT scans illustrating bone destruction of the posterior wall of the maxilla with focal calcification in a tumor (*red arrow*). **c** 15 years after surgery, axial CT scan showing no recurrence. (Color figure online)

of the mandibular ramus, and enclosed the right maxillary tuberosity. In the posterior wall of the maxillary sinus, there was partial bone destruction which had invaded the mucosa of the maxillary sinus (Fig. 1a, b). An incisional biopsy was performed under local anesthesia. The biopsy revealed chondrosarcoma with a high degree of cellular atypia. A surgical resection under general anesthesia was performed by labio-mandibular split approach. The tumor was resected en bloc after making suitable bony cuts through the hard palate, the right maxillary tuberosity, a part of the root of the lateral pterygoid plate, the posterior wall of maxillary sinus and the coronoid process without any complication (Fig. 2). Neck dissection was not performed since there was no lymphatic metastasis in the neck.

A temporary obturator was constructed to obturate the surgical defect.

Postoperative physical therapy was started 1 week after surgery after cessation of edema and pain, and continued over a period of 1 year using home care techniques, involving a mechanical jaw exerciser (Therabite®). Long-term complications involved trismus and difficulty with obturator removal. Six months postoperatively, the patient had scar release; a more suitable obturator was inserted.

Microscopic examination of sections stained with hematoxylin-eosin showed many atypical chondrocytes in hyaline cartilaginous matrix that were arranged in lobular patterns. There was a marked increase in cellularity. The pleomorphic tumor cells with irregular shaped nuclei and myxoid degeneration were seen (Fig. 3a, b). Mitosis was visible in a few cells. Highly positive reactions to S-100 protein (polyclonal, Dako, 1:2000 dilution) immunohistochemical staining was found in many tumor cells in areas with a cartilaginous matrix. The S-100 protein labeling index was 50 % in the tumor cells (Fig. 3c). The pathological diagnosis was a conventional type of chondrosarcoma, Grade III. The histopathological surgical margins were tumor-free; however, a partial site of them was close margin. Subsequently, the patient was irradiated with a total dose of 70 Gy on the surgical defect for 2 months postoperatively.

Due to the possibility of local recurrence and metastasis, resected patients should undergo routine lifelong surveillance. Ideally, surveillance consists of clinical examination and radiological examination of either MRI or CT scan every 3–6 months for the first 5 years and annually thereafter for a minimum of 15 years.

Fig. 2 Intraoperative view. The pterygomandibular space is reached via a labio-mandibular split approach

The postoperative course was uneventful and the patient had no recurrence for 15 years postoperatively (Fig. 1c), but now has been lost to follow-up.

Discussion

Chondrosarcomas are uncommon malignant neoplasms of cartilage that can occur anywhere in the body but are most commonly found in the long bones, and to reported extents of 1–12 % in the head and neck [3, 8]. Chondrosarcoma of the head and neck are rare, accounting for less than 5 % of all tumors of the head and neck region [9, 10]. The median age of patients with this tumor is around 30 years [2], and the patients are reportedly divided equally between males and females, or with a slight preponderance of males. The most frequent site of involvement in the maxillofacial region is the maxilla, followed by the mandible, and the nasal septum [1–4, 8]. To our knowledge, chondrosarcoma of the maxilla extending to the pterygomandibular space, as found in our case, is extremely rare, with no case reported previously.

The diagnosis of chondrosarcoma is made histologically and they are categorized into three grades on the basis of the degree of cellularity, nuclear size and atypia, and mitotic activity. Grade I chondrosarcomas have abundant

chondroid matrix, clusters of chondrocytes with normal or slightly enlarged nuclei, rare nucleoli, absent mitoses, and occasional binucleation. Grade II chondrosarcomas appear to have less chondroid matrix and more chondrocytes than grade I tumors, presence of rare mitoses, slightly enlarged vesicular hyperchromatic nuclei, and multinucleation. Grades III chondrosarcomas display a myxoid matrix with irregularly shaped chondrocytes and increased cellularity, nuclear pleomorphism, and mitoses compared with the low-grade chondrosarcomas [9, 11].

In histopathologic evaluation of chondrosarcoma of the jaw and facial bones, three grades, based on cellularity, pleomorphism, size of nuclei, binucleate cells are usually considered [12]. Myxoid degeneration is an indication for malignancy in cartilage tumors and is widespread in the large chondrosarcoma [7]. Myxoid degeneration was seen in our case.

The treatment of choice for this tumor is wide surgical resection of all the involved structures with negative margins and preservation of function if possible [3]. Incomplete or inadequate surgical margins have been associated with recurrences. The overall recurrence rate within a 10-year duration was 7.25 % (29/400 patients) in a large series studied by Koch and colleagues [8]. Metastatic neck disease on presentation has been reported to be extremely rare in several studies [8]. In a review of 400 cases of head and neck chondrosarcomas, only 5.6 % presented with regional metastatic disease and 6.7 % had distant metastasis [8]. Furthermore, minimal microscopic disease was found in patients who have undergone elective neck dissections [8]. Elective neck dissection in the clinically and radiographically negative neck is therefore not recommended.

The efficacy of radiotherapy in the treatment of chondrosarcoma is controversial. Chondrosarcoma was traditionally regarded as a radioresistant tumour, and radiotherapy was therefore generally reserved for high-grade lesions (as a postoperative adjuvant therapy [13]) and for surgically unresectable lesions [13, 14]. The total dose of radiation ranged from 40 to 70 Gy delivered in 30–35 fractions, which is comparable to treatment regimens used for other sarcomas. Harwood et al. [15] reported that chondrosarcoma was radiosensitive and potentially radio-curable. Krochak et al. [16] reported survival at 5 years for 38 patients who underwent radical radiotherapy. Thirteen of 25 patients with favorable features were progression-free at the 4-year follow-up, which led the authors to conclude that chondrosarcoma might not be radioresistant.

Tumor grade and resectability are the most important prognostic factors for head and neck chondrosarcomas [1–10, 14, 17, 18]. Tumor site is another important prognostic determinant [18]. Factors indicating poorer prognosis include histologically positive margins and high-grade tumor differentiation (Grades II and III) [3]. Some sites in

Fig. 3 **a** Grade III chondrosarcoma with increased cellularity, many clustering the cells, modest hyperchromasia and pleomorphism in lacunae that were arranged in lobular patterns (hematoxylin–eosin stain; original magnification $\times 4$). **b** Higher-magnification photomicrograph showing swollen, pleomorphic, multinucleated cells (hematoxylin–eosin stain; original magnification $\times 200$). **c** Immunoreactivity of the tumor cells for S-100 protein. The S-100 protein labeling index is 50 % in tumor cells (original magnification $\times 200$)

the head and neck region, because of the presence of vital structures, are associated with a poorer prognosis, even with relatively small tumors [4]. Since death in patients with chondrosarcoma in the head and neck often results from intracranial invasion of the tumor due to local recurrence rather than distant metastasis, treatment requires extended surgery. The median size of these tumors is around 4 cm [4]. However, Mark et al. [1] found a predominance of tumors of 5 cm in diameter. Prado et al. [13] reported that tumors smaller than 5 cm treated surgically and having adequate margins had a better prognosis. Thus, prognosis depends on histological grading, anatomical location, tumor size and adequacy of the surgical resection.

Overall, the prognosis is very good with adequate primary treatment. A review of the American College of Surgeons' national cancer database of 179 head and neck chondrosarcomas cases showed a disease-specific survival of 87.2 % at 5 years and 70.6 % at 10 years [8]. Hong et al. [14] reported that a 5-year disease-specific survival rate of 73 %. A worse prognosis has been associated with high-grade tumors and both regional and distant metastatic disease [8].

In conclusion, the mainstay of treatment for chondrosarcoma of the maxilla is radical surgery. Radiotherapy may be used for prevention of local recurrence. The crucial factors determining the prognosis of chondrosarcoma in the head and neck region are the anatomic localization, size, extension, the histologic grade, and surgical resectability of the tumors.

Chondrosarcoma of our case was a conventional type, Grade III, myxoid degeneration, less than 5 cm occurring in the maxilla extending to the pterygomandibular space, the histological surgical margins were tumor-free, however, a partial site of the specimen was close margin, therefore postoperative radiotherapy was performed with 70 Gy was administered to the surgical defect. There was no evidence of local recurrence or distant metastasis 15 years after surgery and radiotherapy. So, it is absolutely essential that carefully further follow-up with radiographic examinations be required because of the possibility that radiation-induced sarcoma may occur.

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Conflict of interest None.

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