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Case report

# A case of chondrosarcoma in the temporomandibular joint

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ABSTRACT

We report a case of a 78-year-old woman with chondrosarcoma in the temporomandibular joint. On examination, swelling of the right preauricular region and pain when opening the mouth was observed. Imaging revealed an irregular-shaped radiopaque mass in the right mandibular condyle with bone resorption. After performing tumorectomy under general anesthesia, histopathological examination revealed that tumor cells had slightly hyperchromatic nuclei that lacked significant pleomorphism, displayed polygonal or stellate profiles of the soma, and formed nodular nests in the homogeneous fibrous cartilage matrix. The pathological diagnosis was chondrosarcoma, Grade II of the right temporomandibular joint.


## 1. Introduction

Chondrosarcoma is a malignant cartilaginous neoplasm that frequently develops in the bones of the trunk, such as the pelvis, femur, and humerus [1], but rarely in the temporomandibular joint [2]. Here, we report a case of chondrosarcoma in the right temporomandibular joint.

## 2. Case report

A 78-year-old woman presented with swelling of the right preauricular region and pain when opening the mouth. Swelling had developed approximately 2 months earlier and slowly enlarged since its first appearance. The patient had a history of thyroid adenoma, osteoarthritis of the knee, hypertension, and cataracts. Her face was asymmetric, and diffuse swelling with tenderness was noted over an area of approximately 40 × 36 mm in the right preauricular region, with pain when opening the mouth (Fig. 1). No abnormalities were detected in blood chemistry tests. The patient provided written informed consent for this case report.

X-ray radiography showed an irregular-shaped radiopaque mass in the right mandibular condyle. On CT, a multilocular mass was observed surrounding the right mandibular condyle. Imaging showed the density of the mass to be slightly less than that of bone, and bone resorption was noted in the right mandibular condyle. Weak enhancement was also observed in the surrounding soft tissue.

The cervical lymph nodes were not swollen, but magnetic resonance imaging (MRI) showed an irregular-shaped mass around the head of the mandible that gave isointense signals on T1-weighted imaging (WI) and heterogeneous low- and high-intensity signals on T2-WI. Although the mass-like shadow could not be enhanced, the unclear boundary of mass in the surrounding soft tissue could be enhanced by dynamic MRI (Fig. 2).

The clinical diagnosis was a right temporomandibular joint tumor. Tumorectomy was performed under general anesthesia via an Al-Kayat-Bramley incision. Rapid intraoperative pathological examination indicated that the lesion was a chondrosarcoma. The peri-tumor tissue was resected while confirming the integrity of the facial nerve. The upper region included the articular disk of the temporomandibular joint. In the lower region, the root of the condylar process was exposed and cut to excise the tumor.

The excised specimen comprised of a mass containing a calcified substance that was elastic, soft and surrounded the head of the mandible. Grossly, the surgically resected specimen, which surrounded the mandibular condyle, was elastic and soft. Histopathological examination of formalin-fixed, paraffin-embedded sections stained with hematoxylin and eosin revealed characteristic features of Grade II chondrosarcoma. At low magnification, the tumor consisted of eosinophilic, pale, and basophilic areas, which were intermingled with each other (Fig. 3a). In the eosinophilic areas (indicated by arrows), tumor cells had slightly

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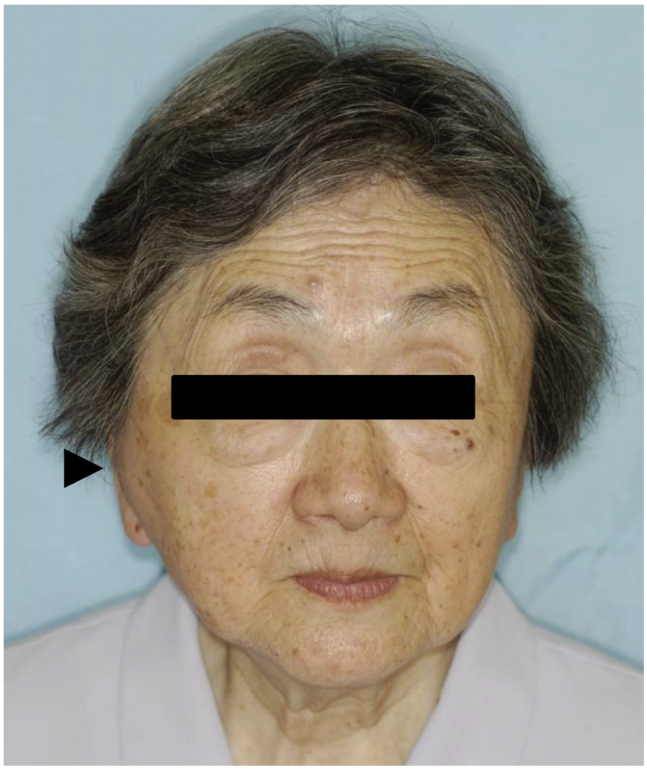

**Fig. 1.** Photograph of patient. Diffuse swelling was noted over an area of the right preauricular region (arrow).

hyperchromatic nuclei without prominent pleomorphism, displayed polygonal or stellate profiles of the soma, and formed nodular nests in the homogeneous fibrous cartilage matrix. In contrast, in pale areas (indicated by asterisks), tumor cells were sparsely distributed in the myxoma-like cartilage matrix (Fig. 3b). In basophilic areas, tumor cells had normochromatic nuclei, displayed round-shaped figures, and were irregularly distributed in the hyaline cartilage-like matrix with focal calcification (Fig. 3c).

Microscopic evaluation after the surgery did not detect any definitive evidence of residual tumor. In addition, no malignant lesions were found by PET-CT. Panoramic radiography of the immediate post-surgical site is shown in Fig. 4. The patient was pain free with satisfactory mouth opening (Fig. 4). There was no evidence of recurrence after a follow-up of 7 years.

## 3. Discussion

Chondrosarcoma in the head-and-neck region is relatively rare, accounting for 1%–12% of all cases [3]. Regarding the region of development, chondrosarcoma most frequently develops in the maxilla, followed by cervical vertebrae, mandible, skull, sphenoid and ethmoid sinuses, frontal sinus, nasal septum and orbit [4].

From 1966–2015, 21 cases of chondrosarcoma in the temporomandibular joint were reported in the English literature (Table 1) [5–22]. The ages of these patients, including the present case, ranged from 23 to 78 years (mean, 46.5 years), and the ratio of male to female patients was 10–12 (Table 1). These values differ from those cited in reports on facial skeletal chondrosarcoma, of which the number of male patients was slightly higher than that of female patients [3,4,26].

The most common symptom of chondrosarcoma in the temporomandibular joint is preauricular swelling (16/22 cases), frequent pain (11/22 cases), and mildly or severely limited mouth opening (8/22 cases). Hearing loss was reported in three cases, with case 3 [7] being due to pressure from tumor on the auditory tube, resulting in otitis media, and cases 6 and 10 [10,14] being due to the occlusive effect of the tumor on the ear canal.

Although the onset of symptoms is generally 3–36 months before the establishment of a final diagnosis, in cases 7, 9, 11, and 13 (Table 1), onset occurred from 72 to 120 months prior to diagnosis. Chondrosarcoma can be primary or secondary, depending on whether the tumor develops *ex novo* or from a preexisting benign lesion, such as a chondroma or exostosis [12]. Cases with delayed diagnosis of chondrosarcoma were most likely secondary chondrosarcoma.

Pathognomonic imaging findings associated with chondrosarcoma consist of local bone destruction and involve the condyle and an expansive mass with calcification. In most cases, an increase in the articular space and length of the condylar neck is observed. Computed tomography (CT) and MRI have demonstrated local bone destruction and spreading of the tumor, which is important for preoperative planning as well as for differential diagnosis [9]. Regarding histological study, hyaline cartilage consists of chondrocytes and a ground matrix, which has a high aqueous content, causing high intensity on T2-WI [27].

Cohen et al. [27] stated that low-grade chondrosarcomas, which are often difficult to diagnose on plain radiography and to differentiate from enchondromas, are characterized by the MR tandem of low signal intensity septa on T2-WI together with septal or ring-and-arc enhancement. In the present study, resorption of the mandibular condyle and an irregular-shaped mass with calcification were apparent on imaging. MRI showed an irregular-shaped mass that gave isointense signals on T1-WI and heterogeneous low-high intensity signals on T2-WI. The mass-like shadow was not enhanced, but an unclear boundary was enhanced in the surrounding soft tissue. Histological examination of the chondrosarcoma revealed proliferation of hyaline cartilage with sarcomatous stroma containing stellate, spindle-shaped, or rounded cells. The presence of mitotic figures was rare.

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