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Chondrosarcoma of the temporomandibular joint. A case report and review of the literature

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Abstract

Chondrosarcoma is a malignant tumor characterized by the formation of cartilage, but not bone, by tumor cells. Only 5% to 10% of chondrosarcomas occur in the head and neck, representing 0.1% of all head and neck neoplasms, with the larynx and the maxillo-nasal region being the most common sites. This report describes an unusual case of chondrosarcoma in a 54-year-old man who presented with pain and swelling in the left preauricular area. Computed tomography demonstrated a soft tissue mass in the left temporomandibular joint without causing erosion of the adjacent bony structures. The tumor was treated by excision in a single block with perilesional tissues, preserving the facial nerve. Histopathologic examination revealed chondrocytes with irregular nuclei with S-100 immunocytochemical staining positive in 30% of the tumor. The diagnosis was a grade I chondrosarcoma. There was no evidence of disease at the 16-month follow-up. The occurrence of chondrosarcoma in the temporomandibular joint (TMJ) is an exceptional event, with only 16 cases described. We report a case of this unusual entity and review the literature.

Key words: *Chondrosarcoma, temporomandibular joint, bone tumors.*

Introduction

Chondrosarcoma is a malignant tumor characterized by the formation of cartilage, but not bone, by the tumor cells (1). After osteogenic sarcoma, chondrosarcoma is the most common bone tumor, representing 10-20% of primary bone tumors. Chondrosarcoma of the head and neck region is a rare disease, compared with the pelvis, ribs, femur and humerus, representing approximately 0.1% of all of head and neck neoplasms. Only 5% to 10% of chondrosarcomas occur in the head and neck, with the larynx and the maxillo-nasal region being the most common sites (2). Chondrosarcoma of the jaw occurs primarily in the anterior maxilla, where pre-existing nasal cartilage is present. The occurrence of chondrosarcoma in the temporomandibular joint (TMJ) is an exceptional event, with only 16 cases reported in literature (3-15) (Table 1). The following is a report of such a case.

Case Report

A 54 year-old man presented with a localized swelling in the left preauricular region that had been present for about three months. The mass, which was causing mild limitation and pain in mouth opening, was fixed to the deep structures and covered with normal skin and there were no cervical lymph nodes palpable. There was no facial palsy. The results of a routine hematologic examination were within normal range, and no signs of inflammation were detected. A fine-needle

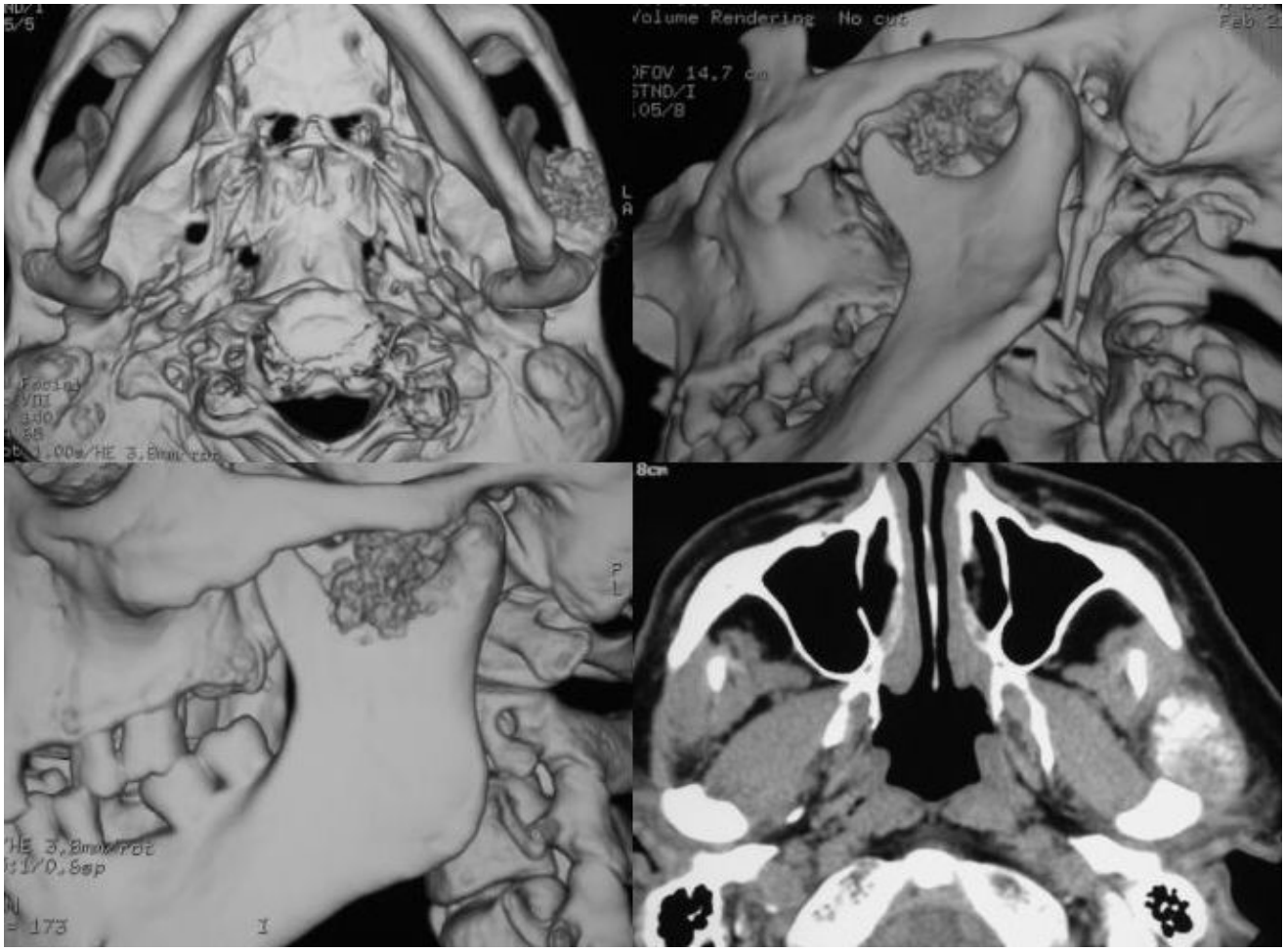

Fig. 1. CT scan and 3D reconstruction showing the mass arising adjacent to the temporal condyle, with extension into the infratemporal fossa.

aspiration biopsy was not diagnostic. The computed tomography (CT) and 3D reconstruction showed a mass arising adjacent to the temporal condyle, with a slight extension into the infratemporal fossa, but without causing erosion of the adjacent bony structures (Figure 1).

The tumor was surgically resected through a Finochetto-Joel incision. The mass appeared under the branches of the facial nerve and showed no visible invasion into the surrounding soft tissues (Figure 2). There was no adhesion between the lesions and the disc. An intraoperative frozen section showed that the specimen was made up of cartilaginous tissue, but a definite diagnosis was impossible. The mass was dissected from the adjacent structures (temporal condyle, sigmoid notch, and zygomatic arch) and removed in a single block with perilesional tissues. The facial nerve was preserved. The size of the excised mass was 22 x 13 x 5 mm and appeared to be glistening cartilaginous tissue, grayish-white, with a marked lobular pattern on the cut surface.

Microscopic examination of sections stained with hematoxylin-eosin revealed chondrocytes with irregular nuclei

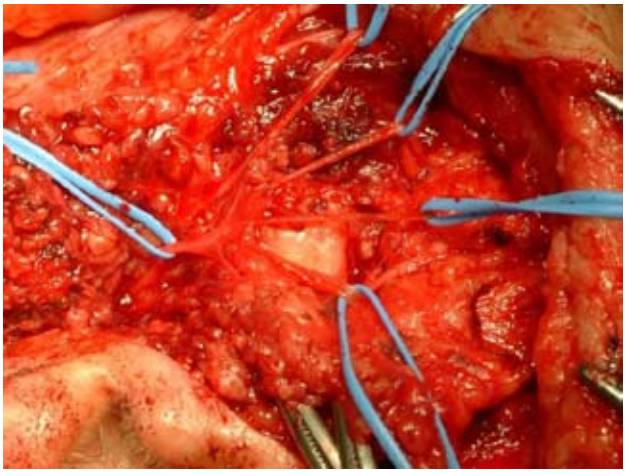

Fig. 2. Intraoperative view of the greyish-white mass under the branches of the facial nerve.

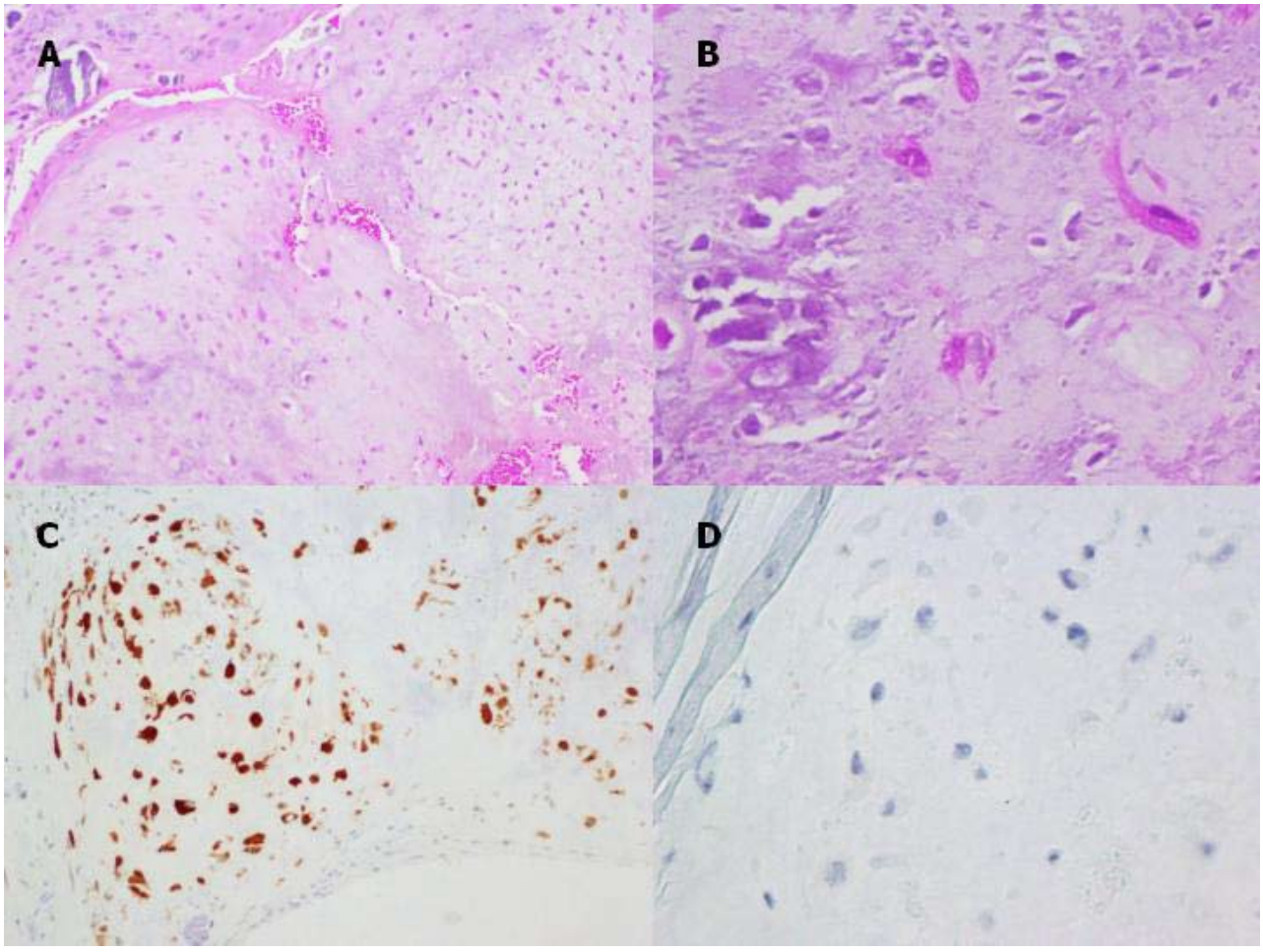

Fig. 3. A.- Hematoxylin-eosin stain showing chondrocytes arranged in lobular pattern (x 40). B.- Hematoxylin-eosin stain showing variable cartilage cell shape. Mitoses were not seen (x100). C.- S-100 immunocytochemical staining was positive in 30% of the tumor cells. D.- Inmunohistochemical study with Ki67 showing less than 5% of cellular activity.

that were arranged in lobular patterns (Figure 3A). The cartilage cell shape was variable and bizarre. Mitoses were not seen (Figure 3B). S-100 immunocytochemical staining was positive in 30% of the tumor cells (Figure 3C) and inmunohistochemical study with Ki67 was made, showing less than 5% of cellular activity (Figure 3D). The diagnosis was a grade I chondrosarcoma.

The patient was discharged one week after the operation in good condition and was not referred for radiation therapy because all margins were tumor-free. There was no evidence of recurrence of the tumor 16 months after surgery, and the patient was continuing to receive routine follow-up at the time of writing.

Discussion

Chondrosarcomas are malignant tumors of cartilage. Chondrosarcoma represents 10-20% of all malignant bone tumors, and of these, 1 to 12% originate in the head and neck region, with the most common sites of occurrence in the head and neck being the larynx, followed by the mandible, nasal cavity and maxilla (16,17). In our review, only 16 cases of chondrosarcoma of the TMJ were identified in the literature (3-15). When all head and neck sites are considered, the tumor has a slight male predilection and occurs more frequently in the fourth to seventh decades of life (16). However, our review of chondrosarcomas of the TMJ suggest a female predominance (Table 1).

The onset of symptoms is generally 3 to 24 months before the establishment of a final diagnosis, an exception being a case of Nitzan et al. (12) and Mostafapour et al. (15) (case 2) (Table l), in which they had begun about 6 and 8 years before respectively. The most common symptom is a swelling in the preauricular region, frequently with pain and mild limitation in mouth opening. In three cases, a reduction in hearing was also reported.

In conventional radiographic investigation and on the CT scan, no pathognomonic findings are associated with chondrosarcoma (18), which appears as a mass with single or multiple radiolucent areas that may contain calcifications and cause condylar deformity, bone destruction and sometimes erosion of the surrounding bony structures (the glenoid space, the auditory canal, base of the cranium and infratemporal fossa). In most cases, an increase in the articular space, length

Table 1. Reported Cases of Chondrosarcoma of the Temporomandibular Joint.

Reference	Sex	Age (years)	Symptons duration	Main complaint	Imaging presentation	Treatment	Follow-up
Gringrass (3) 1954	F	46	12 m	Pain in TMJ Swelling	Slight TMJ space Subcortical sclerosis	Surgery	No report
Lanier and Wilkinson (4) 1971	F	48	24 m	Pain Swelling	Condyle resorption	Surgery	Few months
Richter et al (5) 1974	M	75	10 m	Hearing loss Pain Swelling	Erosion glenoid fossa Increased length condylar neck TMJ space widening	Surgery	12 months
Tullio and D'Errico (6) 1974	F	17	8 m	Swelling	Condyle resorption	Surgery	No report
Nortje et al (7) 1976	M	40	6 m	Dull pain Swelling	TMJ space widening Condyl elongated and distorted	Surgery	24 months
Sato et al (8) (Sudo) 1977	-	-	3 y	Pain	-	Surgery and Irradiation	-
Sato et al (8) (Kato) 1977	-	-	18 m	Swelling Pain, trismus	Radiopacity Radiolucency	Surgery	-
Sato et al (8) (Kato) 1977	-	-	4 m	Swelling Pain, trismus	Radiopacity Radiolucency	Surgery	-
Cadenat et al (9) 1979	F	60	-	Swelling	-	Surgery	6 months
Morris et al (10) 1987	F	29	24 m	Headache Swelling	Mass from condyle to infratemporal fossa	Surgery and Irradiation	6 months
Wasenko and Rosenbloom (11) 1990	F	49	-	Swelling Pain Hearing loss	Mass from condyle to infratemporal fossa with calcification	Surgery	No report
Nitzan et al (12) 1993	F	36	6 y	Spontaneous pain; swelling	TMJ space radiolucent lesion resorption of condyle	Surgery	7 years
Sesenna et al (13) 1997	F	60	12 m	Swelling	Mass from condyle to infratemporal fossa Calcification	Surgery	5 years
Batra et al (14) 1999	M	65	18 m	Swelling Hearing loss	Mass anterior to ear canal encasing the mandibular condyle	Surgery	7 months
Mostafapour (15) et al 2000 (case 2)	F	23	8 y	Left facial swelling	Left pterigoid space mass with involvement of TMJ	Surgery	-
Mostafapour (15) et al 2000 (case 3)	F	52	1,5 y	Right cheek mass	Mass on right TMJ Involvement of petrous temporal bone and middle fossa	Surgery and Irradiation	6 months
Our case 2007	M	54	3 m	Swelling Pain trismus	Mass adjacent to the temporal condyle, extension into the infratemporal fossa	Surgery	16 months

of the condylar neck, and radiopacity of the condyle can be seen. Specifically, for TMJ chondrosarcoma, radiographic evidence of widening of the joint space was reported in most of the cases, including our case.

Histopathologically, chondrosarcomas of the TMJ appear similar to chondrosarcomas of the head and neck or other regions of the body. These lesions appear as lobulated, cellular neoplasms. Microscopically, this tumor showed a hyaline cartilaginous proliferation, with a sarcomatous stroma containing stellate, spindle-shaped, or rounded cells (16). The presence of mitotic figures is rare, and their absence does not rule out the diagnosis of chondrosarcoma. Criteria for diagnosis of malignancy in these tumors include an increased number of cells, increased size of the nuclei, cells with binucleate forms, or giant cell tumor formation.

This tumor has been classified into grades I, II, and III based on the frequency of mitoses, cellularity, and nuclear size (19), although these data are from chondrosarcomas arising from all joints, not just the TMJ. There can be difficulty in distinguishing between a well-differentiated chondrosarcoma (grade I) and a chondroma. Fine-needle aspiration biopsy, although proposed by several authors, does not always give exact results in this region because of the need to distinguish the tumor from osteogenic sarcoma, parotid pleomorphic adenoma, and chondroma (13).

The clinical behaviour of this tumor is variable and linked to histologic grading. Local recurrence, however, is more common than distant metastasis, and there is a relatively good relation between tumor grade and prognosis, although the most important prognosis factor is resecability (6), which makes complete escision of the tumor the most single significant factor in the prognosis. The current grading system and subsequent measures of survival are based on studies of all joints, not just the TMJ. The paucity of patients with this tumor involving the TMJ has precluded a useful analysis of survival in these patients. Thus there are no large series reporting effectiveness of adjuvant therapy in these patients.

The survival rate of these tumors is reported as 44-87.5% (16), and the most common cause of death in chondrosarcoma is recurrence, not metastasis. Histologic differentiation also influences the metastasis rate, which varies from 10% for grade II cases to 71% for those in grade III. Metastasis from grade I chondrosarcoma has not been reported. The most frequent metastatic sites are the limbs and the lungs; involvement of the regional lymph nodes is unusual (13).

For the subset of patients with chondrosarcoma of the TMJ, wide local resection is also the treatment of choice. The overall low incidence of regional metastasis may suggest that neck dissection is not indicated (20). Chondrosarcoma, although traditionally regarded as a radioresistant tumor, was reported by Harwood et al. (21) to be radiosensitive and potentially radiocurable. In the reviewed literature rarely includes irradiation as single modality treatment, and is used most commonly as adjunct. The primary role of radiation therapy has been for the treatment of unresectable disease and, after surgery, resection of incompletely resected lesions. Chemotherapy has a limited role in chondrosarcoma, but can be applied as an adjuvant therapy in cases with aggressive behaviour, rapid local recurrence and high grade chondrosarcomas (20). Our case received a correct surgical therapy, involving removal of the neoplastic mass with perilesional tissue, so irradiation and chemoteraphy were not needed. No recurrence was reported after 16 months follow-up, but 10 to 15 years follow-up has been reported as necessary to fully characterize survival after treatment of chondrosarcoma (20).

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