# Chondrosarcoma of the Mandibular Condyle: A Case Report

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#### KEY WORDS

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#### **ABSTRACT**

Chondrosarcomas are slow-growing, malignant mesenchymal neoplasms characterized by formation of cartilage by the tumoral cells. They display a wide range of morphological features from a well-differentiated growing mass resembling a benign cartilage tumour to a high-grade malignancy with aggressive local invasion. Only 5% to 10% of this neoplasm is confined to the head and neck region. Chondrosarcomas of the mandibular condyle may manifest the typical symptoms of the temporomandibular joint dysfunction syndrome. Tumours of the condyle can reach a large size without producing clinically obvious swellings. A rare case of chondrosarcoma of the mandibular condyle in a 34-years old woman is presented in this report. Patient's chief complaint was pain in the right temporomandibular joint when her mouth was in a maximum opening position. Mild malocclusion, figured as an occlusal discrepancy, was also detected. Radiographs illustrated erosion in the head of condyle. After condylectomy, the excised mass was histologically diagnosed as a grade II chondrosarcoma.

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### Introduction

Chondrosarcoma is a malignant neoplasm that is characterized by formation of cartilage, with progressive local invasion to the surrounding tissue. Currently, it is known as the second most common bone tumour, after osteosarcoma [1-2]. Chondrosarcomas occur in patients of all ages and the majority of them are over 50 years old. No significant gender or race predilection is noted yet [1-2].

Chondrosarcomas comprise 10-12% of all malignancies of bone tumours. They usually arise in the femur, humerus, pelvis, and sacrum [3]. Approximately 1% to 3% of chondrosarcomas occurs in the maxillofacial region, predominately in the anterior maxillary region, while mandibular body, ramus, nasal septum and paranasal sinuses are less affected. The favoured site of involvement in the mandible is the molar region, and they rarely occur in the ramus,

condyle, coronoid process, or symphysis [1, 4]. Chondrosarcomas are slow-growing hard tissue sarcomas which are characterized by formation of cartilage owing to tumoral cells. Primary chondrosarcomas develop de novo, whereas secondary chondrosarcomas arise from pre-existing chondroma or osteochondroma [1-2].

Clinical manifestations of mandibular chondrosarcomas are presence of an enduring mass, painless swelling or sometimes pain, paresthesia and trismus. Tooth mobility is observed in advanced stages of chondrosarcoma which is possibly attributable to the widening of the periodontal ligament space and to the bone loss as well, a condition which may resemble periodontal lesions [2, 4].

Radiographical features of this malignancy are presence of erosive radiolucent area with ill defined borders that may include calcification. In fact, bone

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destruction is often detected and occasional opacities caused by calcification are observed [2-3].

To the best of our knowledge, only 12 cases of chondrosarcomas, evolved in the symphysis of mandible, have been reported. In the mandible, remnants of Meckel's cartilage could be the sources of cartilaginous cells which are able to produce secondary cartilage in the mental region and the coronoid and condyloid processes. These structures could be a rational for development of a tumour in these regions [3-4].

Only 38 cases of primary mandibular chondrosarcoma in the mandibular condyle have been reported over the 40-year period of time up to 1974, in the English language literature. Three of these cases occurred within the mandibular condyle, however; in general, all kinds of temporomandibular joint tumours are rare [5]. Review of 3200 tumours of the head and neck by Nwoku and Koch, assessed in the clinic of maxillofacial and plastic surgery of the face from the Westdeutsche Kieferklinik, Dusseldorf, showed only 7 cases arising in the condylar head [6].

Most of neoplastic lesions of the mandibular condyle grow slowly, but eventually lead to some clinical signs such as deformity and deferred pain, clicking, shifting of the mandible to the unaffected side upon opening and also malocclusion. Some studies reported that chondrosarcoma of the mandibular condyle may provoke preauricular swelling, sharp preauricular pain and tenderness on pressure [7-9].

This report describes a new case of chondrosarcoma which has arisen from mandibular condyle. The clinical, radiographical, surgical and pathological aspects of this lesion are presented, and the relevant literature is reviewed.

## **Case Report**

A 34-years-old woman referred to the Dental School of Isfahan University of Medical Sciences, presenting with a small hard swelling in front of her right ear. When mouth was in its maximum opening situation, patient complaint of pain in the right temporomandibular joint area and mild malocclusion without any jaw deviation was also detectable.

The pain and swelling had been present for 2 months. There were no complaints of malocclusion or pain in chewing activities. Intraoral examination showed no notable features. Further screening examinations showed that there was no cervical lymphadenopathy and all cranial nerves were intact.

Extraoral examination revealed indurated domeshaped swelling, sized about 3×2×1cm situated in front of right ear with unnoticeable facial asymmetry as the swelling was masked by the patient's hair. The overlying skin was intact and normal.

Panoramic radiograph showed erosion in the head of condyle (Figure 1). An expansile lytic lesion involving the condyle was seen in computed tomography (Figure 2). The radiographic diagnosis was a malignant intra-osseous neoplasm.



Figure 1 Panoramic radiography showed erosion in the head of condyle

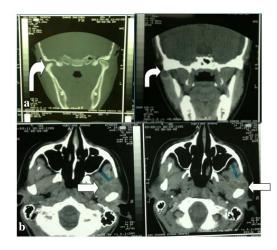


Figure 2a Coronal Computed tomography scans showing an expansile lytic lesion involving the mandibular condyle.

b Horizontal Computed tomography scans showing an expansile lytic lesion involving the mandibular condyle.

According to clinical and radiographical features, differential diagnoses were chondrosarcoma, osteosarcoma, and Ewing's sarcoma. Under general anaesthesia the tumour was exposed through a preauricular incision. The whole right condyle was resected. In macroscopic or gross examination, the excisional biopsy specimen was a gray-brown, cream coloured piece of head of condyle (40×30×20mm) with presence of cartilage and the cut-surface showed a lobulated pattern (Figure 3).

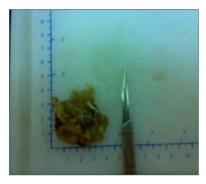


Figure 3 Tumor resected by condylectomy with surrounding capsules

Microscopic findings showed proliferation of chondroblasts in atypical lacunae that were arranged in lobular patterns with pleomorphism and mild-to-moderate nuclear hyperchromatism in dysplastic chondroid matrix. In some lacunae, there was binucleated or multinucleated chondroblasts. In addition there was ischemic necrosis and margins of tumour were not involved (Figure 4).

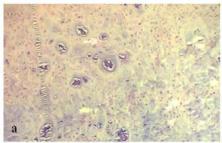
Based on these findings, our diagnosis was chondrosarcoma (Grade II). After condylectomy; the patient did not receive any other treatment. Therefore we decided to follow up the patient at regular intervals. The patient is in a good condition now.

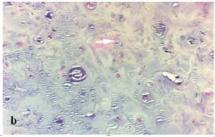
### Discussion

The importance of this case is ascribed to its rarity. In the present case, later diagnosed as condylar chondrosarcoma, the only clinical presentation was small swelling in front of right ear with pain and mild malocclusion in the right temporomandibular joint when her mouth was in maximum opening and not in the mastication. Therefore in all of the temporomandibular joint pains and malocclusion, even being mild, condylar chondrosarcoma should be considered in the clinical differential diagnosis. This tumour should be speculated after ruling out the prevalent causes of temporomandibular joint pain and malocclusion such as myofacial pain dysfunction syndrome, traumatic arthritis, osteoarthritis, rheumatoid arthritis and injuries of articular disc.

Radiographic features of this case were more representing a condylar malignancy than its clinical characteristics. Radiolucency with poorly defined margins and expansile lytic lesion with erosion of head of condyle was observed with occasionally mottled opacities induced by calcification. These radiographic features suggest chondrosarcoma, osteosarcoma, Ewing sarcoma and other malignant bone tumours to be included in the radiological differential diagnosis. Histopathological features of this case confirmed the diagnosis of chondrosarcoma and subsequently other diagnoses were ruled out. According to these findings, in the problematic cases of temporomandibular joint disturbances, excisional biopsy and histopathological consideration is highly recommended [8-9].

Chondromas of the jaws are exceedingly uncommon and most of them, proved to be low-grade





**Figure 4a** Chondrocytes in lacunae arranged in lobular patterns (hematoxylin and eosin; original magnification  $100 \times$  **b** atypical lacunae that were arranged in lobular patterns with pleomorphism and mild to moderate nuclear hyperchromatism in dysplastic chondroid matrix had been seen in magnification  $400 \times$ .

Chondrosarcomas after a concise investigation [1]. Therefore, even those chondrogenic tumours of the jaws which have benign appearance should be considered malignant unless proved to be otherwise. In one survey, 32% of patients with an early diagnosis of benign chondroma, chondroma or osteochondroma had an ultimate diagnosis of chondrosarcoma; and this correct diagnosis took about 12 months in average [2].

Histopathological features of chondrosarcomas range from well-differentiated appearing like benign cartilage tumours to high-grade malignancies with aggressive local invasion [1-2]. These tumours may be divided into three histopathological grades of malignancy. The grading system correlates well with the rate of tumour growth and prognosis for chondrosarcomas of the extragnathic skeleton [1].

Grade I chondrosarcomas, closely mimic the appearance of a chondroma, composed of chondroid matrix and chondroblasts that show only subtle variation from the appearance of normal cartilage and have no metastasis.

Grade II chondrosarcomas, which recur more often than grade I lesions, show a greater proportion of moderately sized nuclei and increased cellularity particularly about the periphery of the lobules in myxoid and less common hyaline matrix and occasionally exhibit mitotic figures. The rate of metastasis is approximately 10%.

Grade III chondrosarcomas are highly cellular and may show a prominent spindle cell proliferation, and mitotic figures are prominently increased. The rate of metastasis in grade III lesions is more than 70%.

The 5-year survival for chondrosarcoma is approximately 90% for grade I, 81% for grade II and 43% for grade III [1-2].

Chondrosarcoma may be misdiagnosed with chondroblastic osteosarcoma or even Ewing's sarcoma, because of their similarity in histological features [1-2].

Differential diagnosis of chondrosarcoma from chondroma and chondroblastic osteosarcoma is difficult but important. Chondroma usually arises in small bones, and is extremely rare in the jaws and facial bones. It is wise to consider any cartilaginous tumours of the jaws, malignant or potentially malignant rather than benign. It is also important to differentiate chondrosarcoma from chondroblastic osteosarcoma in the jaw; since prognosis of chondrosarcoma is more favourable than chondroblastic osteosarcoma [4]. Chondrosarcoma grows more slowly than chondroblastic osteosarcoma and metastasizes late in the course of tumour growth [1].

Osteosarcomas of the jaw that have chondroblastic differentiation make confusion with chondrosarcomas. The lack of neoplastic osteoid, neoplastic bone or alkaline phosphatase expressing tumour cells rule out a chondroblastic osteosarcoma [3]. Chondroblastic osteosarcoma, in contrast to the present case, often contains chondroblastic areas with highly pleomorphic chondroblasts in addition to dysplastic osteoid tissue and more pleomorphic sarcomatous background.

The prognosis for chondrosarcoma is related to the size, location, and grade of the lesion. The most important factor is the location of the tumour because this has the greatest influence on the ability to achieve complete resection [1]. Richter et al. stated a male predominance for cases of mandibular chondrosarcoma reported in the literature [10].

The prognosis of patients with chondrosarcoma is varied from series to series. Shafer et al. reported that chondrosarcomas of the jaws were exceedingly dangerous and often resulted in death [11]. In the National Cancer Database series of 400 head and neck chondrosarcomas, only approximately 12% of patients had regional or distant metastasis at the time of diagnosis, with a tendency for metastasis in higher grade tumours and also for sinonasal lesions. The overall 5 and 10 year disease- specific survival rates in this series were 87.2% and 70.6% respectively [1].

Positive margins in histological appearance and high-grade tumour (Grades II and III) indicate poorer prognosis. Treatment of these lesions is wide surgical excision with negative margins [1-2]. Chondrosarcomas usually develop slowly but may be invasive. Lymph node metastasis of jaw chondrosarcomas is rare; hence, elective neck dissection is not routinely

required. Distant metastasis is also rare and usually occurs in more advanced or recurrent tumours. Distant metastasis to the lungs, sternum and vertebrae has been reported [1-3].

For high-grade chondrosarcomas, radical surgery may be needed. Chondrosarcoma is simply implanted in soft tissue where it can grow rapidly and invade to adjacent tissues. There is some controversy about the radiosensitivity of these tumours. Conventional chondrosarcoma are regarded as a radio-resistant tumour, so radiotherapy is generally introduced for high-grade lesions as an assisting therapy and also for surgically unresectable lesions [1-3]. Conversely, Harwood and others reported that chondrosarcoma was radiosensitive and potentially radio-curable [12].

The present case was a relatively small sized grade II tumour arising in the mandibular condyle region and was treated with condylectomy. The resected specimen showed tumour-free margins, suggesting a favourable prognosis.

However, long-term follow-up is essential, because chondrosarcomas show a wide disparity in the interval between recurrences and also in their possibility of metastasis.

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