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Mesenchymal Chondrosarcoma Simulating a Periodontal Disease: A Case Report

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Abstract

Mesenchymal chondrosarcoma (MCS) is a rare high-grade variant of chondrosarcoma. MSC's represent about 1% to 9% of chondrosarcomas. Less than 500 cases of MCS have been reported in literature, so far. As opposed to other variants, MSC develops outside of the bone in almost third of the cases. Pain and swelling are the most common symptoms at presentation. The overall prognosis of MSC is poor, with frequent distant metastases. Histopathologically, MCS typically exhibits a biphasic pattern consisting of both small cells, and islands of atypical cartilage. The immunohistochemical profile of this lesion overlaps with several other entities. Resection with wide margins is usually the treatment of choice. MCS is believed to be more responsive to chemotherapy and radiotherapy, than the other variants of chondrosarcoma. We are reporting a case of mesenchymal chondrosarcoma on the buccal alveolar mucosa of the first and second left mandibular molars, in a 25 year old female.

1. Introduction:

We are reporting a case of mesenchymal chondrosarcoma on the buccal alveolar mucosa of the first and second left mandibular molars, in a 25 year old female.

Mesenchymal chondrosarcoma (MCS) is a rare, high-grade variant of chondrosarcoma. As opposed to other variants, MSC develops outside of the bone in almost third of the cases. It typically affects women in their second and third decades of life. Patients usually present complaining of pain and swelling of a short duration. MSC runs an aggressive clinical course with frequent distant metastases to the lungs, lymph nodes, and other bones (1, 2). The overall prognosis is poor, with a 5- year survival rate of 55%. Improved survival rates are noted in MSCs of the head and neck, this may in part be due to the easier detection at these locations (3).

Histopathologically, MCS typically exhibits a biphasic pattern comprising small dark cells, and islands of atypical cartilage. Microscopically, the differential diagnosis of MCS can be intricate, and encompasses, fibrosarcoma, dedifferentiated chondrosarcoma, hemangiopericytoma, osteosarcoma, lymphoma, and Ewing's sarcoma/primitive neuroectodermal tumor group of tumors. Studies using Immunohistochemical markers frequently reveal a positive reaction with S100 protein, vimentin and CD99; whereas, cytokeratin, EMA, and actin are usually negative (4,5). However, and due to the variable immunohistochemical profile, MSC is diagnosed primarily based on its histomorphological pattern.

Less than 500 cases of MCS have been reported in literature, so far, thus and Owing to its' rarity, MCS is still poorly understood. Resection with wide margins is typically the treatment of choice, and several groups opted to treat MCS in accordance to Ewing's sarcoma treatment protocol (1,2). MCS is believed to be more responsive to chemotherapy and radiotherapy, than the other variants of chondrosarcoma (6).

1.1. Case report:

Our patient is a 25 year old female patient who was referred to our institution with a chief complaint of "I have a bump on my gum". The patient stated that a month prior to her visit to our facility, she was diagnosed at another clinic to have a periodontal disease and was managed accordingly. She also reported that the lesion did not respond to periodontal therapy and that the lesion has doubled in size over the last two month and that it was not painful.

Physical examination of the case revealed a firm, soft tissue swelling, on the buccal alveolar mucosa, of teeth numbers 20 and 21. The overlying mucosa appeared to be intact and of normal color and texture (fig. 1). Radiographically, a panorex radiograph revealed no notable changes (fig. 2). An incisional biopsy was performed at our maxillofacial surgery clinic, and the specimen was submitted for histopathological examination (fig's 3-9).

Histopathologic examination revealed three overlapping patterns, with the first pattern consisting of islands of low-grade malignant cartilage and, the second pattern consisted of sheets of undifferentiated small spindle to round basophilic cells. The third pattern consisted of focal areas resembling a hemangiopercytoma. We rendered a diagnosis of mesenchymal chondrosarcoma. The patient has since been referred to a head and neck surgery center, where she underwent a mandibular segmental resection, based on imaging results (MRI and CT). The patient has also been scheduled for chemotherapy.

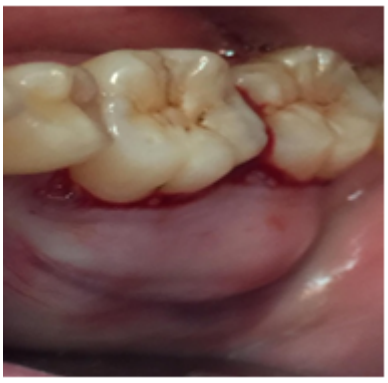

Figure 1: Intra-oral clinical pictures of the swelling on the alveolar buccal mucosa of teeth numbers 20 and 21

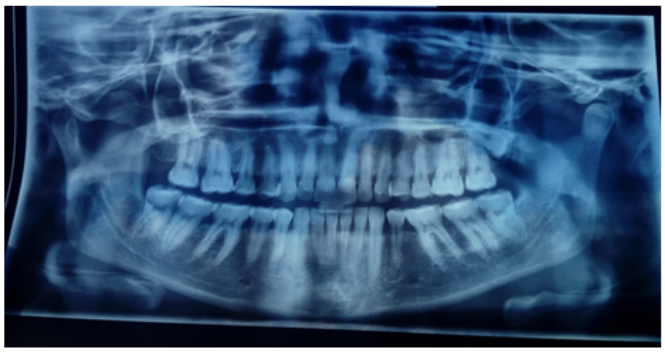

Figure 2: A Panoramic radiograph of the patient, showing no notable changes.

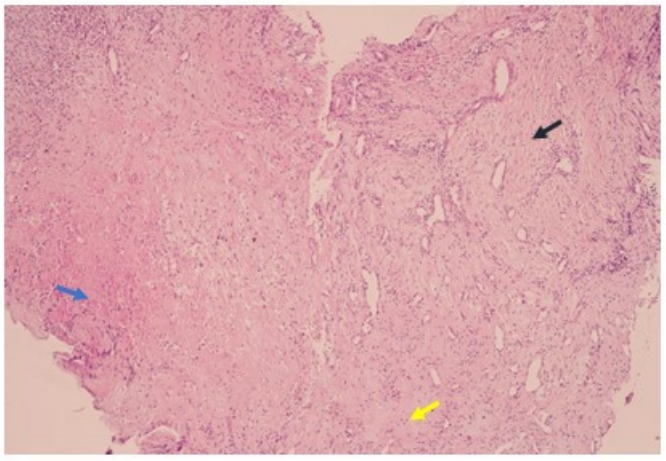

Figure 3: Photomicrograph of the biopsy specimen exhibiting overlapping zones of mature appearing cartilage (yellow arrow) with focal areas resembling a hemangiopericytoma (black arrow). Note: areas of necrosis were also evident (blue arrow) (H&E, orig. mag. X10

2. Discussion:

Mesenchymal chondrosarcoma is an uncommon yet aggressive variant of chondrosarcoma. This variant is different from other variants of chondrosarcoma in that about third of the cases are located extra skeletally with the orbits, meninges and lower extremities being the most commonly affected parts of the body. MCS runs a rapid aggressive clinical course. It is debated that MCSs of the head and neck exhibit a less aggressive clinical behavior, than those located elsewhere. How-

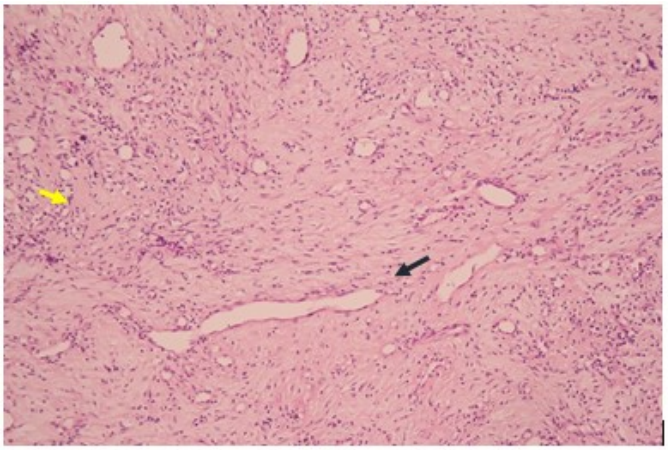

Figure 4: Photomicrograph of the biopsy specimen exhibiting areas of mature appearing cartilage (yellow arrow) and areas resembling a hemangiopericytoma. (H&E, orig. mag. X20)

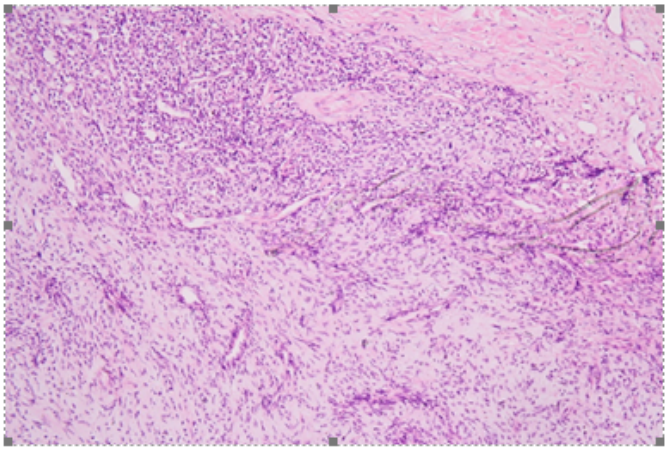

Figure 5: Photomicrograph of biopsy specimen showing sheets of basophilic cells (H&E, orig. mag. X20)

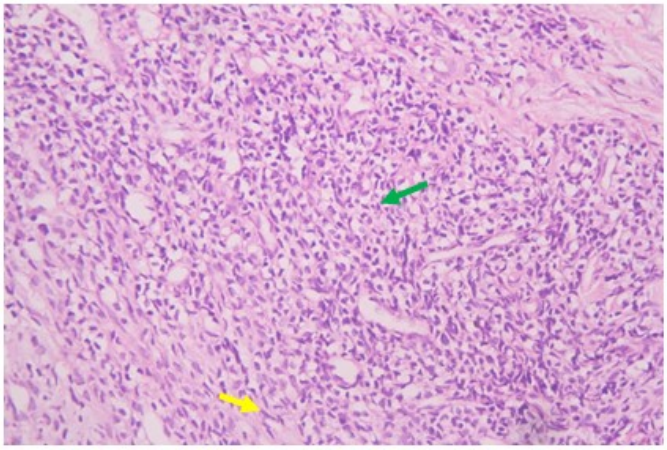

Figure 6: Photomicrograph of the biopsy specimen showing sheets of basophilic cells exhibiting a round (green arrow) to spindle morphology (yellow arrow) and scattered blood vessels (H&E, orig. mag. X40)

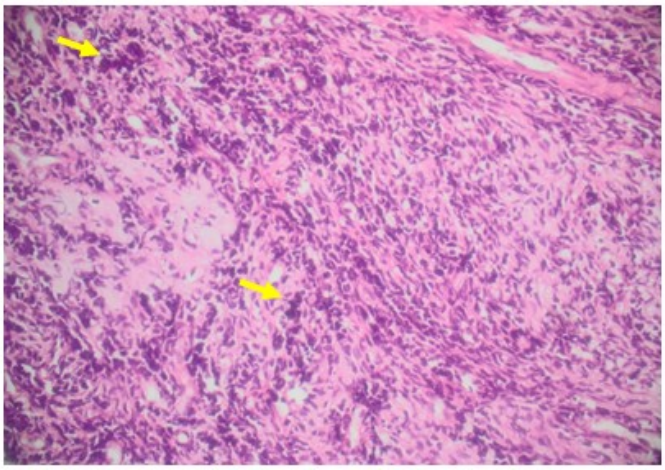

Figure 7: Photomicrograph of the biopsy specimen exhibiting basophilic cells with 'smudging' (crush artifact) reminiscent of a lymphoma (yellow arrow) (H&E, orig. mag. X20)

ever, this is attributed by many authors to the fact that lesions affecting the head and neck are readily noticed, early on in the course of the disease.

Histopathologically, this lesion is made up mainly of two populations of cells, undifferentiated round or spindled cells with intervening areas of well-differentiated hyaline cartilage. Both cell types are believed to originate from primitive cartilage forming mesenchyme.

MSC can prove to be a diagnostic challenge, especially if the cartilaginous areas are inconspicuous, this is particularly true for small biopsies. The most frequent items on the differential diagnosis list for MSC are hemangiopericytoma, dedifferentiated chondrosarcoma, lymphoma and Ewings sarcoma. Immunohistochemical analysis is not always decisive in this regard, as there are some overlapping immune positivity between MSCs and other tumors on the differential diagnosis list, like CD99 and S100 protein.

Wide resection is the recommended surgical approach. Adjuvant chemotherapy is usually scheduled after the surgical procedure and was documented to improve survival rates. The role of Radiotherapy however is still debatable, to date.

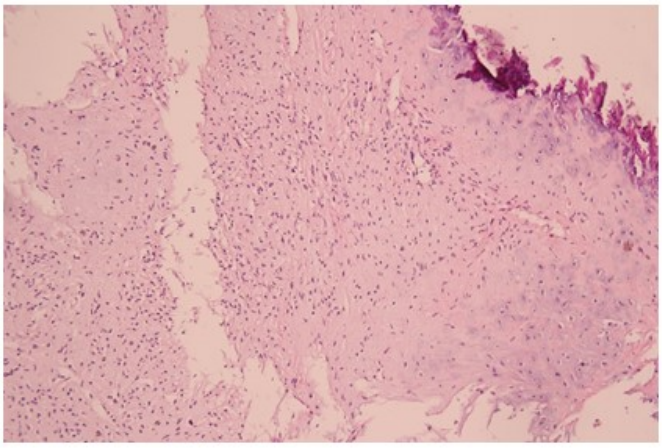

Figure 8: Photomicrograph of the biopsy specimen exhibiting areas of low-grade chondrosarcoma. Note: calcification at the periphery (H&E, orig. mag. X20)

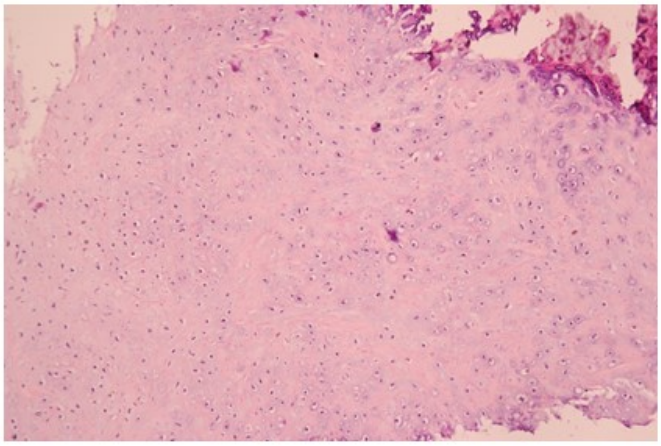

Figure 9: photomicrograph of the biopsy specimen exhibiting areas of low-grade chondrosarcoma. (H&E, orig. mag. X20)

Retrospective studies have correlated the size of tumor and the presence of metastatic deposits to the prognosis of the disease.

The case reported here in emphasis the fact that, some really evil tumors can mimic some really benign/inflammatory conditions and vice versa. Also, this report highlights the fact that, MCSs of the head and neck are readily discovered and hence may be managed at an earlier stage than would MSCs in other locations.

2.1. Conflict of interest:

None declared.

2.2. Ethical statement:

The work has been approved by the ethical committee of Future University in Egypt and the patient gave an informed consent to the work.

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