

SHORT REPORT

Mesenchymal Chondrosarcoma of the Mandible

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Mesenchymal chondrosarcoma (MCS) is one of the most unusual neoplasms. It was first described in 1959 by Bernstein and Lichtenstein as a distinct variant of chondrosarcoma (CS) (1). It usually appears in the second and third decades of life (2). This neoplasm affects females more frequently than it does males (F/M = 1.4/1) (3). One of the most affected regions is the facial skeleton, especially the jaw. The lesion was originally grouped with osteosarcoma. These neoplasms are characterized by sheets or clusters of highly undifferentiated, small, ovoid cells that alternate with small zones of neoplastic cartilage. The prognosis for patients with MCS is unpredictable. This type of neoplasm shows aggressive local behavior as well as a high metastatic potential. Due to these features and the high risk of recurrence, the prognosis is poor (3).

In this report we present a case of MCS primarily involving the mandible and review its clinical presentation, histopathology and treatment.

Case Report

A 30-year-old male patient applied to our clinic in March 2000 with complaints of pain and a mass on his left jaw. The patient stated that he had first been examined by a dentist. Antibiotic therapy was given by the dentist, who assumed that the mass was due to a dental abscess. Since the complaints continued and the mass was growing continuously, the patient was referred to our

clinic. The patient stated that 1 tooth had been extracted from the side of his jaw 3 years before and that he had lost 7 kg in body weight within the last 2-3 months. His medical and family histories were non-contributory. On physical examination we determined that there was a fixed, rough and painful mass (6 x 5 x 5 cm) on the left angulus mandible. Oral examination revealed that the third molar was eroded, that there was a macerated area (0.5 x 0.5 cm) on the left retromolar region and that the gingiva was hyperplastic. The first and second molar teeth were mobile and perioral paresthesia was present on left side. There were no other pathological findings upon systemic examination.

The biochemical parameters of the patient were within the normal ranges. (Hb: 13.1 gr/dl, Hct: 35.4%, sedimentation rate: 13 mm/h). Ultrasonography (USG) of the neck revealed that there was an anechoic mass (6 x 5 x 5 cm) along with cortical thickness and irregularities on the left side of the mandible. Abdominal USG did not show any pathology. Abdominal and lung computerized tomography (CT) were also negative. CT of the neck demonstrated that there was a massive lesion along with changes in the bone structure invading the soft tissue of the left mandible (Figure 1).

Intraoral incisional biopsy was performed and undifferentiated tumor cells with a round-to-oval nucleus and clear cytoplasm were observed microscopically. Immunohistochemically, these cells were positive for vimentin, but negative for S-100 protein, neuron specific

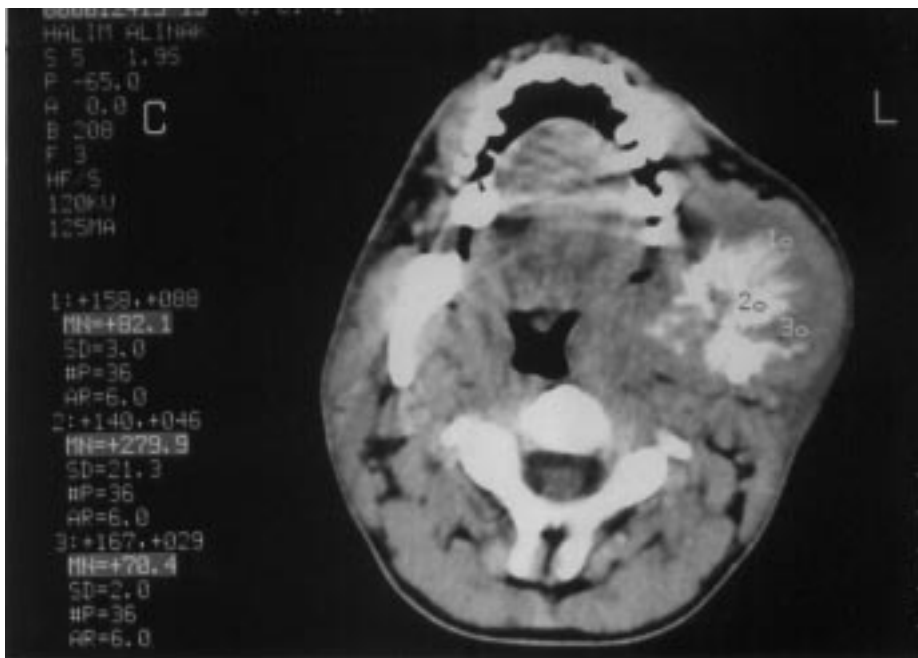


Figure 1. Computerized coronal tomography of the neck: a massive lesion with changes in bone structure invading the soft tissue of the left mandible.

enolase, cytokeratin, desmin, FVIII, and CD68. In the light of these findings, the case was diagnosed as poorly differentiated malign mesenchymal tumor.

The patient was then operated on. A left hemimandibulectomy was performed with wide surgical margins under general anesthesia. The postoperative course was uneventful. No chemotherapy or radiation treatment were given preoperatively. The lesion was pink and fleshy, mimicking other sarcomas. There were foci of calcification.

The microscopic appearance of the tumor was islands of well differentiated cartilage that were juxtaposed to small round-to-oval cells (Figure 2). These cells simulated Ewing's sarcoma. In some regions of the tumor, the small malignant cells were arranged around vascular spaces (hemangiopericytomatous pattern) (Figure 3). The chondroid islands were calcified, but there was no ossification. Benign giant cells were also present in a few areas (Figure 4).

Examination of surgical specimens showed that the surgical margins were free of tumor. In view of the disease the patient was given a course of radiotherapy and a combination of chemotherapies. Currently

(postoperative 35th month) the patient is alive and there has been no local recurrence or distance metastasis.

MCS is an uncommon but characteristic malignant tumor first described by Lichtenstein and Bernstein (4). CSs are also rare malignant neoplasms in the head and neck (5). MCSs constitute 3% to 9% of all CSs. In a comprehensive evaluation of 400 skeletal extraosseous CSs of all types, Huvos et al. (6) found 35 cases of MCS (8.7%). Forty-six cases have been published in the literature of which 42 were reviewed by Takahashi et al. in 1993 (7). Nakashima et al. (8) reported that MCSs can occur in both osseous and extra-osseous sites in a ratio of 2: 1. Furthermore, 3% and 25% of all skeletal MCSs occur in the maxillofacial region (9). The maxilla and mandible are common sites of skeletal origin. The maxilla was the more common intraosseous site in Christensen's report, but in our case the location of the MCS was the mandible (10). Extraskelatal tumors commonly arise from the orbit, meninges, nasal and paranasal mucosa, and the parapharyngeal space, which are among the extraskelatal sites reported in the literature (11). MCS of the jaw is always seen in the second and third decades of life. The sex incidence is approximately equal. The most common location of CS in the maxilla is in the premolar

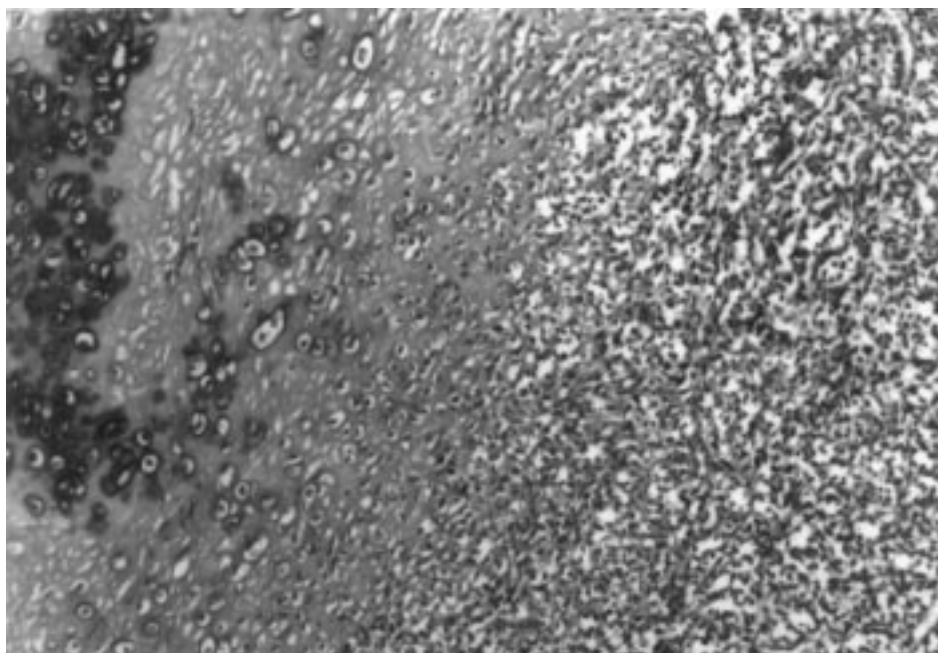


Figure 2. On the left, well differentiated chondroid areas, and on the right, undifferentiated neoplastic cells with small, round nucleus and clear cytoplasm (H&E, x100).

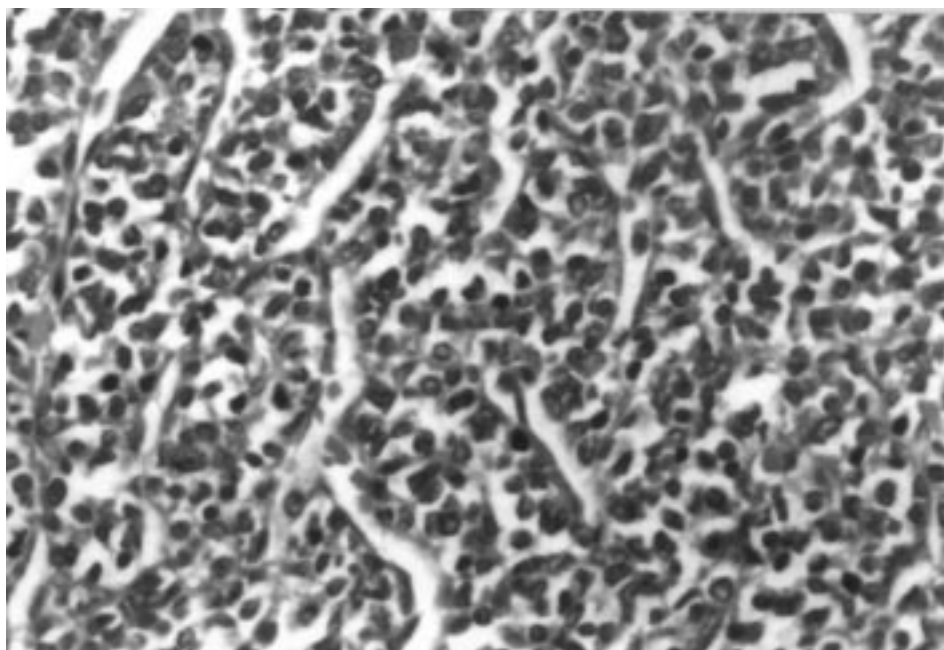


Figure 3. Vascular clefts among neoplastic cell groups (hemangiopericytomatous pattern). (H&E, x400).

and molar area. In the mandible the most common location is the premolar-molar area but the symphysis or coronoid condylar processes may be involved (12). In our

case, the location of the mass (6 x 5 x 5 cm) was between the subcondylar and the mentum of the left mandible.

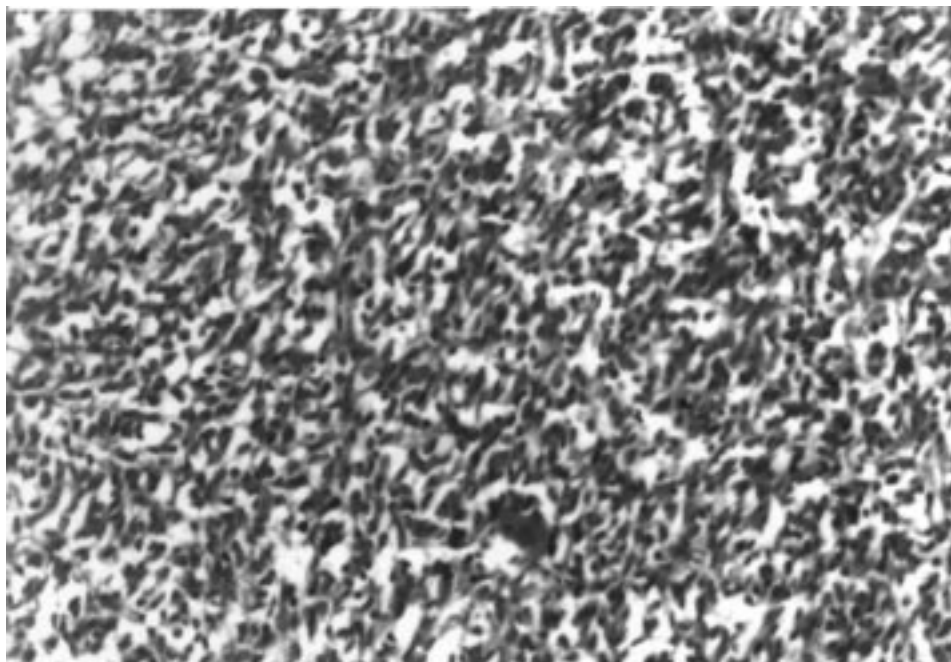


Figure 4. Giant cell formation, between poorly differentiated neoplastic cells (H&E, x200).

There were no specific clinical signs or symptoms. The predominant symptom was usually a painless mass or swelling (53%) although a painful mass is also frequently (16%) reported (13). Facial paralysis, bleeding from the lesion, nasal obstruction, epistaxis, paresthesia, difficult hearing and incidental findings are also observed. Dental complaints may be the initial symptoms. Injudicious local surgery, dental extractions or even biopsy may provoke more rapid growth (3). Our patient stated that he had first been seen by a dentist and that antibiotic therapy had been given. Three years before, 1 tooth had been extracted from the side of the patient's jaw. Clinically, MCSs have some major problems including high malignant potentiality, tumor recurrence and a low survival rate.

Three dimensional CT is very useful for MCS. On CT, the tumor presented as a well defined mass with multiple areas of fine and coarse calcification. The most common radiographic appearance of MCS is radiolucency. Takahashi et al. (7) reported that MCS of the mandible has a radiolucent osteolytic shadow. CT in our case showed a massive lesion with changes in bone structure invading the soft tissue of the left mandible.

The most effective therapeutic modality is wide surgical excision (14). Mandible-wide local excision with a

tumor free margin of 2 to 3 cm is recommended (12,15). According to Nakashima et al. (8), extensive resection has less recurrence and a better survival rate than limited surgical resection (1). Although there have been reports of resolution of this tumor with chemotherapy and radiation alone (2), the benefit from chemotherapy and radiation is as yet unclear (15). However, experience of these methods is limited and there is no evidence that these therapies improve the prognosis (16). However, postoperative radiotherapy and chemotherapy offer a good prognosis and eradicate micrometastases that have not been previously detected.

The prognosis for MCS is poor because tumors have a tendency to late recurrence either locally or as metastasis (14). Metastasis of MCS is hematogenous and the most common site is the lung. Five-year survival rates for craniofacial CSs are 40%-60%, and at least 60% of patients have recurrences within 5 years of initial treatment (12). Nakashima et al. (8) found that 43 out of 71 patients (61%) developed metastasis after an average of 4.3 years. Takahashi et al. (7) reported that 6 out of 14 patients with mandible tumors died after an average survival time of 29 months and that survival time is longer with maxillary than with mandibular lesions. Huvos and Marcove (17) identified 35 patients, 8 of

whom died of the disease within 5 years, and 6 between 5 and 10 years after diagnosis. Overall there was a 10-year survival rate of 28% (2).

We present a case of MCS, because MCS is an uncommon tumor and is rarely found in the jaw. An adequate biopsy is enough for diagnosis. It may be radiolucent, radio-opaque or mixed. MCS requires surgical excision with wide margins. Pre- and postoperative chemotherapy or radiation may be a choice of treatment, although their effectiveness is unclear. MCS

may be considered in the differential diagnosis of mandibular masses.

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