A 62-year-old Man with Chondroblastic Osteosarcoma of Maxilla: A Rare Case

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Abstract

Osteosarcoma (OS) is a malignant tumor that induces formation of osteoid or immature bone and rarely occurs in maxilla. Maxillofacial OSs are reported to occur in the third to fifth decades that is 1-2 decades later than the mean age for OS of long bones. The present case is a 62-year-old man with facial asymmetry and pain in his left upper jaw. Intraoral examination showed a bony hard, tender swelling about 50 x 60 mm in diameter in the maxillary alveolar ridge extending unilaterally from the maxillary incisor to the molar region which had expanded the buccal and palatal aspects of alveolar ridge. Computed tomography showed a hyper dense mass in the left maxilla extending to maxillary sinus. The diagnosis of OS was confirmed by histopathologic evaluation. Following the initial surgery, a local recurrence developed in the facial region of the patient within 18 months. Radiographic evaluation of OS of maxilla is important for diagnosis. Early diagnosis and radical surgery are the keys to increasing survival rate.

Key words: Osteosarcoma, Chondroblastic Osteosarcoma, Jaw.

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Introduction

Osteosarcoma (OS) is a primary malignant tumor of bone which is characterized by formation of osteoid tissue. Although it is the most common malignancy of long bones after multiple myeloma (1, 2), it is a relatively rarer entity in the craniofacial region. About 6% of OSs arise in the jaws (3). The etiology of OS is unknown, but some risk factors such as a previous history of ionizing radiation, alkylating agent, retinoblastoma and benign bone lesions such as Paget disease and fibro osseous dysplasia have been associated with development of head and neck OS (2-4). Jaw Osteosarcoma (JOS) occurs with a peak in the third through fifth decades of life. The mean age is approximately 33 -39 years that are one to two decades later than the mean age for OS of long bones (3). Mandible is the most common site of involvement followed by maxilla and skull (4). Mandibular tumors arise more frequently in the posterior body and horizontal ramus whereas maxillary tumors occur in alveolar ridge, the sinus floor and palate (2, 5-7). Symptoms usually include painful swelling in the area and loosening of teeth, although paresthesia, nasal obstruction and ophthalmic complications such as proptosis may be noted. Radiographic findings vary from radiolucent, mixed radiopaque - radiolucent or entirely radiopaque lesion with irregular border. Widening of periodontal ligament space and enlargement of the mandibular canal are also widely described as important radiographic features. The radiographic feature of OS is not

pathognomic. (2-4, 6, 8, 9). Thus, incisional biopsy and histopathologic analysis are essential for final diagnosis and treatment planning. Depending on the relative amounts of osteoid, cartilage or collagen fibers produced by the tumor, pathologists subclassify OSs into three types: osteoblastic, chondroblastic and fibroblastic (3, 4, 6, 7, 9). This article reports a case of chondroblastic OS of maxilla. The aim of this case was to draw attention to the possibility of diagnosing this tumor based on its clinical and radiographic characteristics before its confirmation by histology.

Case Report

A 62-year-old man was referred to oral medicine department, with the chief complaint of facial asymmetry and swelling of his left upper jaw. He also had pain and paresthesia of the region innervated by infra orbital nerve .The pain was dull and continuous in nature and did not subside with medication. He had first noted the lesion about six months before and it had gradually increased in size (Fig. 1). The patient also explained difficulty in chewing on the affected side. His past dental history revealed previous extraction of the left maxillary lateral and canine six month before. Intraoral examination showed a bony hard tender swelling with a smooth surface about 50 x 60 mm in diameter in the maxillary alveolar ridge extending from the maxillary incisor to the molar region that expanded to the buccal and palatal aspects of alveolar ridge (Fig. 2). No cervical lymphadenopathy was detected. Systemic examination did not reveal abnormal clinical findings.

A first panoramic (six month prior to his visit) showed a mixed lesion with irregular borders in the left maxilla (Fig. 3) and in the next panoramic (at arrival), the lesion was completely radiopaque with irregular ossification in that area (Fig. 4). Computed tomography revealed an expansive lesion which involved the left side of maxilla and invaded into the lateral wall of nose and infero-lateral wall of the left maxillary sinus. Axial CT showed bicortical expansion and presence of radial spicules which spread outside the jaw bone on the palatal side, giving a “sun ray appearance” in relation to the molar teeth region (Fig. 5).

Based on the clinical and radiological findings, a provisional diagnosis of malignancy was given. Incisional biopsy verified the diagnosis of chondrosarcoma (Fig. 6). The patient was evaluated for possible metastasis by Chest X-ray and abdominal ultrasonography both of which proved to be normal. A whole-body bone scan with IV injection of 99m TC-MDP showed an increased activity in the left maxilla (Fig. 7). After the evaluation, a surgical operation was performed and the patient underwent a wide excision of the tumor with hemimaxillectomy of the left side. The histopathologic examination of the specimen was chondroblastic OS (Fig. 8).

The margins of the surgical resection were negative for the tumor. The patient refused to receive chemotherapy and also did not complete treatment; therefore, six months later, the lesion recurred. Unfortunately he died 18 months after recurrence of his lesion.

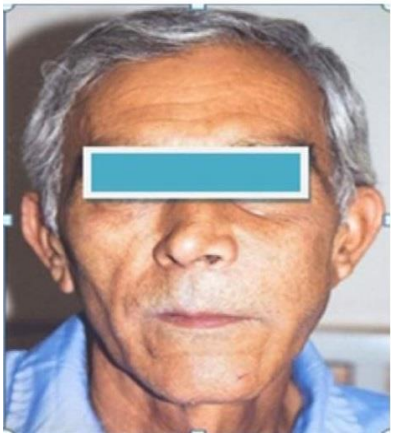

Figure 1. Extra oral view showing asymmetry of face

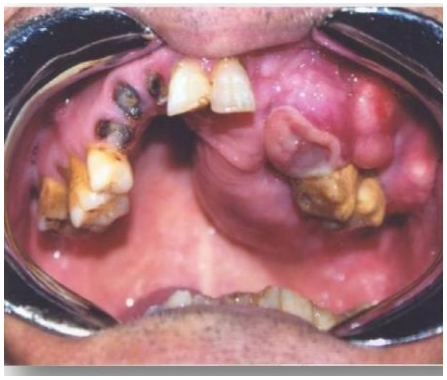

Figure 2. Intra oral view showing buccal and palatal expansion

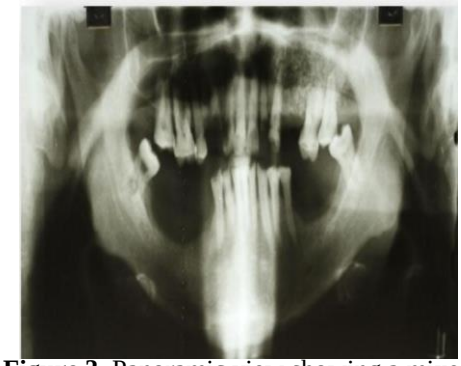

Figure 3. Panoramic view showing a mixed RO-Rl lesion in the left upper jaw in 6 months ago

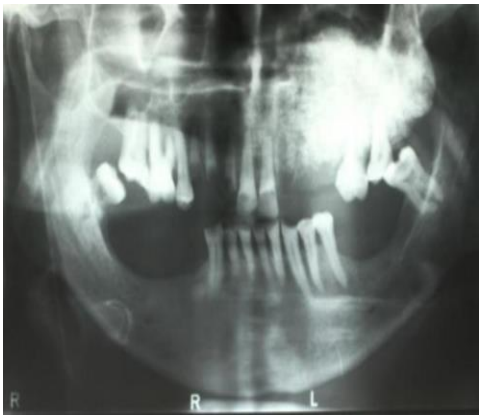

Figure 4. Panoramic view showing a radiopaque lesion in the left upper jaw

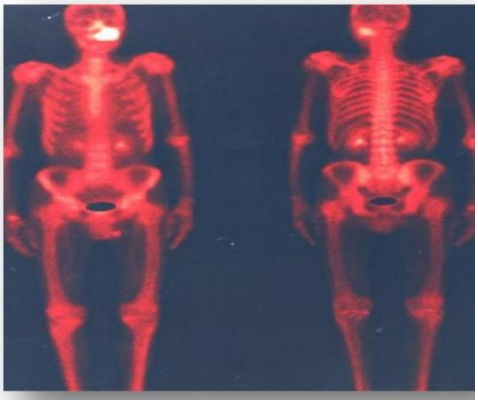

Figure 7. whole body scans with active site in left maxilla

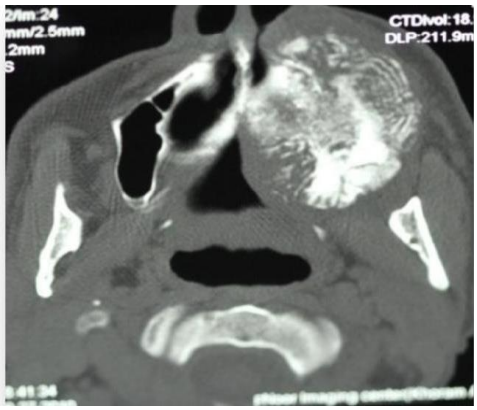

Figure 5. axial CT showing the lesion invade to nose and sinus and bicortical expansin

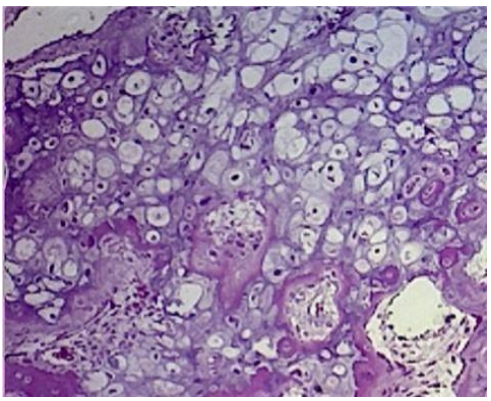

Figure 8. photomicrographs of excisional biopsy: osteoid, chondroid and atypical cells with hemorrhage are seen(X100)

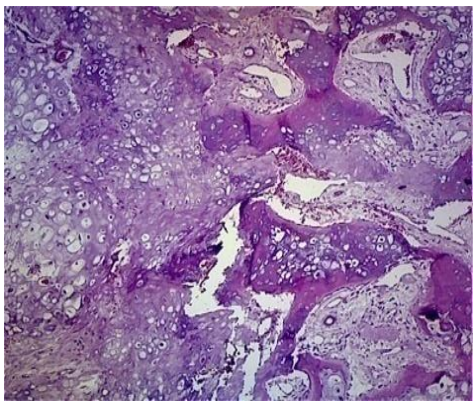

Figure 6. photomicrograph of incisional biopsy show chondroblastic osteosarcoma(X40)

Discussion

OS is the most common malignant bone tumor after multiple myeloma; however, JOSs are rare and their biological characteristics are different of other bones (3, 4). At the time of diagnosis, the patients are approximately 2 decades older than those with OS of other bones, which have a peak incidence in the third through fifth decades of life, better 5-year survival rate, rare metastases and local recurrences(3). OS is classified into two types, primary and secondary. The etiology of the primary type is unknown presumably due to genetic influence or other environmental factors. Secondary OSs arises in various clinical settings such as Paget disease, giant cell tumor, multiple osteochondroma, chronic osteomyelitis and history of radiation exposure (2, 3, 9, 10) In this case, the etiology was unknown. The clinical features of this case were similar to previous studies, regarding sex, site of lesion and chief complaints but the

age was higher than those. August et al. (11), in a study of 30 patients (20 men and 10 women, mean age of 34 years), found that seventeen lesions occurred in mandible and 13 in maxilla. Swelling without pain was the most common presenting symptom. Thirteen lesions were initially misdiagnosed as odontogenic infections. Numbness as a presenting symptom was statistically associated with poor prognosis. The average duration of symptoms before diagnosis was 3–4 months. The most frequent locations in the craniofacial region were alveolar ridge and antrum of maxilla and the body, symphysis and ascending ramus of mandible. However, our patient had a painful swelling, paraesthesia and nasal obstruction. Yamamoto et al. (12), described a case of maxillary OS with unusual image findings in an 11-year-old boy. The lesion was in the center of palate. No abnormality was observed in the maxillary bone on panoramic or occlusal radiograph.

Since initial findings of OS can be less suggestive, it is necessary to be more conscious when such patients are being diagnosed, as they often remain undiagnosed for considerable durations and this delay can negatively affect prognosis. A number of various infections, benign neoplastic diseases or malignancies can produce some of the OS's signs and symptoms. Various differential diagnosis of OS due to its nonspecific symptoms and radiographic features can be considered (2). Amini Shakib et al. (13) described an OS of maxilla of an edentulous patient that was misdiagnosed as epulis fissuratum. Lindquist et al. (14), reported that widening of periodontal ligament space and inferior dental canal, together with sunburst effect are almost pathognomic of JOS. Yesilova et al. (15), reported a 50-year-old woman complaining of her remaining root and pain in the right maxillary posterior region which had lasted for two months. On panoramic radiograph, the borders of the lesion were ill-defined. Also the "sun-burst appearance" was seen. They believed the lesion was malignant because of the radiological findings, clinical appearance and the history of the patient.

Our case, in terms of clinical and radiographic presentations, is very similar to the cases mentioned above, and it imitates many specifications of jaw osteosarcomas that led us to a possible diagnosis of osteosarcoma.

Since not all the lesions show such peculiar characteristics, diagnosis of this type of malignancy only through plain radiographic feature is difficult (2-4). The extent of OS in bone and soft tissue was best determined by cross sectional imaging techniques such as computed tomography and magnetic resonance imaging (2, 3, 8, 9). Atypical sunray speculations were seen in our case that was strongly suggestive of OS.

According to the predominant type of extracellular matrix present, OSs are classified histopathologically into osteoblastic, chondroblastic, fibroblastic subtypes. The osteoblastic type consists of tumor osteoid surrounded by bizarrely arranged osteoblastic cells. (16, 3) In chondroblastic OS, tumor cells lie in the lacunae and form lobules. Fibroblastic OS is the last variant where the tumor cells are spindle-shaped and characteristically arranged in a herring bone pattern typically resembling fibrosarcoma. The formation of tumor osteoid differentiates this variant of OS from fibrosarcoma (2, 3, 6, 9). In our case, due to the small size of the sample and possibility of a limited investigation of the biopsy specimen, the interim diagnosis of chondrosarcoma was established. After removing the whole lesion and performing histopathologic assay, the definitive post-operative diagnosis of chondroblastic osteosarcoma was verified. This diagnosis is based on observing direct production of osteoid from malignant mesenchymal cells and malignant cartilage with a lobular pattern.

Recently IHC is used to detect a variety of OSs like chondroblastic OS from chondrosarcoma or low-grade OS from benign histological mimics. For example in chondosarcoma IHC is positive for S100 and vimentin and negative for cytokeratin and epithelial membrane antigen (EMA). In chondroblastic OS, IHC is positive for Vimentin, EMA, S100 and rarely cytokeratin (3, 9).

The treatment of choice for JOS is wide surgical resection. Chemotherapy is useful for OS of long bones, leading to significant changes in the disease-free survival rate. But use of chemotherapy in JOS is controversial, because of its rare occurrence and lack of standardized chemotherapy protocols making it difficult to assess the effectiveness of the adjuvant therapy (10). In many cases the treatment of choice is radical surgical excision and it provides a 5-year survival rate of over 80%. It appears that it does not have much effect on survival rates. It can be described on the basis of its rare and late metastases. In brief, OSs of the jaw are less aggressive than those of long bones and have a different behavior from them (3, 10). Treatment includes all combinations of surgery, chemotherapy and radiotherapy. In August et al. (11), study Patients receiving chemotherapy with four or more agents showed a trend toward better survival with 71% alive and disease-free at the time of review. Higher age was statistically associated with decreased survival. The average age of survival was 27 years. Older patients suffered more local recurrences which, in all but one case, resulted in mortality. Our patient avoided receiving chemotherapy and finally died after 18 months.

Conclusion

In summary, OS of maxilla has an aggressive biological behavior. Therefore, early diagnosis and radical surgery with wide surgical margins are the keys to a good outcome. This article would bring attention of dental professionals to approaching such cases with greater concern so as to diagnose them at an early stage leading to better prognosis.

Competing Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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