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A rare case of maxillary osteoclastoma with pulmonary and calvarial metastasis

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Abstract

An uncommon, benign, locally aggressive osteolytic neoplasm called a giant cell tumor (also known as an osteoclastoma or myeloid sarcoma) of the bone typically affects matured long bones and accounts for 5 to 9% of all primary bone tumours. It mainly targets the epiphysis of long bones. Most often benign, they rarely spread to the lungs. The treatment plan usually ranges from simple curettage to wide resection, with typically a good prognosis. The recurrence rate has been reported to be between 40% and 60% and 5% to 7% of recurrent cases are malignant. However, such tumor growth is rarely encountered in craniofacial bones like the temporal, ethmoid, sphenoid, maxilla, and mandible which accounts only for 2% of cases. Herein this article we present and discuss the clinical presentation, radiological features, histopathological investigations, treatment procedures and follow-ups done in such a rare case of giant cell tumor involving maxilla in a young female patient.

Keywords: Giant cell tumor, Osteoclastoma, Maxilla, Metastasis

Introduction

Cooper in 1818 first described the giant cell tumor of bone and later Nelaton presented its local aggressiveness followed by Virchow who revealed its malignant potential ^[1]. A giant cell tumor of bone (otherwise named osteoclastoma or myeloid sarcoma) is an uncommon, benign, locally aggressive osteolytic tumor primarily affecting the skeletally matured long bones of the body ^[2]. It usually arises from the epiphysis of matured long bones and in case it affects immature bone it arises from the metaphysis and extends into the epiphysis of the involved bone. It accounts for 5-9% of all primary bone tumours. Although, its occurrence in the craniofacial region is very rare and accounts for only 2% of cases. In the craniofacial region, it mostly affects the temporal bone followed by the sphenoid and mandible. Its occurrence in the maxilla is very rare and it is seldomly reported. Most of these tumors are benign they rarely metastasize to the lungs ^[3]. The aetiology of this tumour is not clear but some authors suggest that it arises from mononuclear cells with RANKL expression and osteoblastic progenitor characteristics ^[4]. A histopathological examination is needed for a confirmatory diagnosis. Imaging modalities like CECT, CT, and CBCT are also recommended as diagnostic approaches. A Chest X-ray or CT Chest helps to diagnose the metastasis status ^[5]. The treatment plan ranges from simple curettage to wide resection with a good prognosis. The recurrent rates of this tumour account for about 40-60%. Hence, we report and discuss the clinical diagnostic challenge, radiographic features, histopathological investigations, treatment procedures and follow-ups done in such a rare case of giant cell tumour involving maxilla in a young female patient reported to our department.

Case Report

A 30-year-old young female patient reported to the Department of oral medicine and Radiology with a chief complaint of swelling on the left side of the face for the past 6-7 months. The patient was normal 6-7 months back and she noticed swelling on the left side of the face which was small initially and gradually increased to the present size. The swelling was not associated with pain.

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After being asked questions regarding her systemic health, the patient gave a history of joint pain in the lower extremities for the past 3 years. She also gave the history of gradual bending of the phalanges of her right thumb, index and ring finger, and left thumb and index finger for one year. On general examination, all her vital signs were within normal limits. Her past dental history, family history, and personal history were not contributory.

On extraoral examination, facial asymmetry was present due to diffuse swelling present on the left side of the face which extends anterior-posteriorly from the ala of the nose to the outer canthus of the eye and superior-inferiorly from the infraorbital rim to the occlusal plane. The skin overlying the swelling appeared normal in colour as that of the adjacent area (Figure 1a). On palpation, the swelling was firm in consistency, slightly compressible, non-tender, afebrile, and non-pulsatile. In comparison with the right side, the nasolabial fold on the left side of the face was obliterated with the lifting of the ALA of the nose. Left submandibular lymph nodes are palpable, single, round in shape, soft inconsistency, and tender on palpation.

On intraoral examination, grade II mobility was present with 11, 12, 21, 22, 23, 24, 25, 26, 27 teeth with poor oral hygiene.

A well-defined swelling of size 5x5 cm approx. seen extending anterior-posteriorly from the left maxillary central incisor to the maxillary tuberosity region, superior-inferiorly from the buccal vestibule to the alveolar crest of the involved tooth, and mediolaterally from the buccal vestibule to palate crossing the midline. The mucosa over the swelling was slightly brownish in appearance as compared to that of the adjacent mucosa. Expansion of both buccal and palatal cortical plates with obliteration of the buccal vestibule was present (Figure 1b).

On palpation, the swelling was firm in consistency, slightly compressible, non-tender, afebrile, and non-pulsatile. With this overall clinical presentation, a provisional diagnosis of a Benign Tumor involving the maxilla on the left side was made. Considering other systemic symptoms, size, site, extent, and severity of the lesion we gave hyperparathyroidism as the first differential diagnosis followed by giant cell lesion/tumour, fibrous dysplasia, aneurysmal bone cyst, and ameloblastoma. On laboratory investigations, serum alkaline phosphatase level was elevated (1888 IU/L) with serum calcium and phosphorus level being within the normal limits.

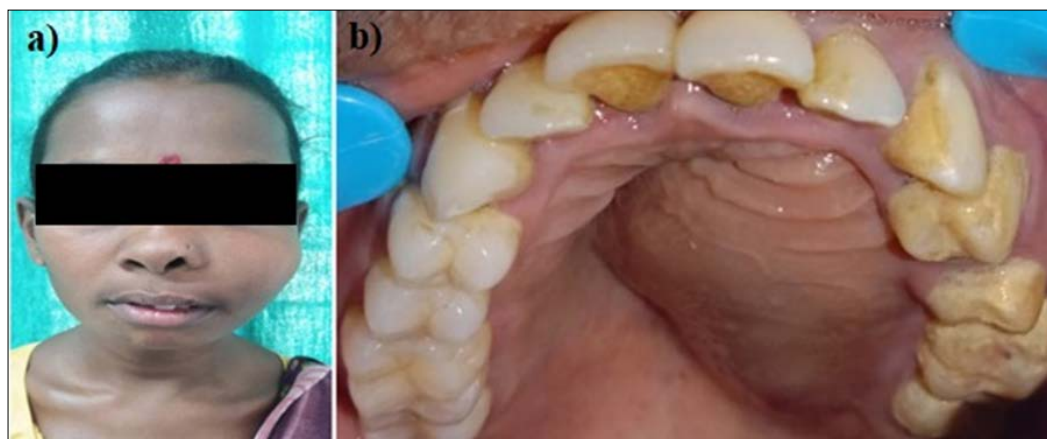


Fig 1: a) Extraoral photograph showing extent of the lesion; b) Intraoral photograph showing lesion in the left maxilla with a slightly brownish appearance

On radiographic examination, the hand-wrist radiograph shows the bending of terminal phalanges of the thumb, index, and ring finger. The lateral cephalogram of this patient shows

multiple tiny osteolytic changes over the entire skull calvaria (Figures 2a and 2b).

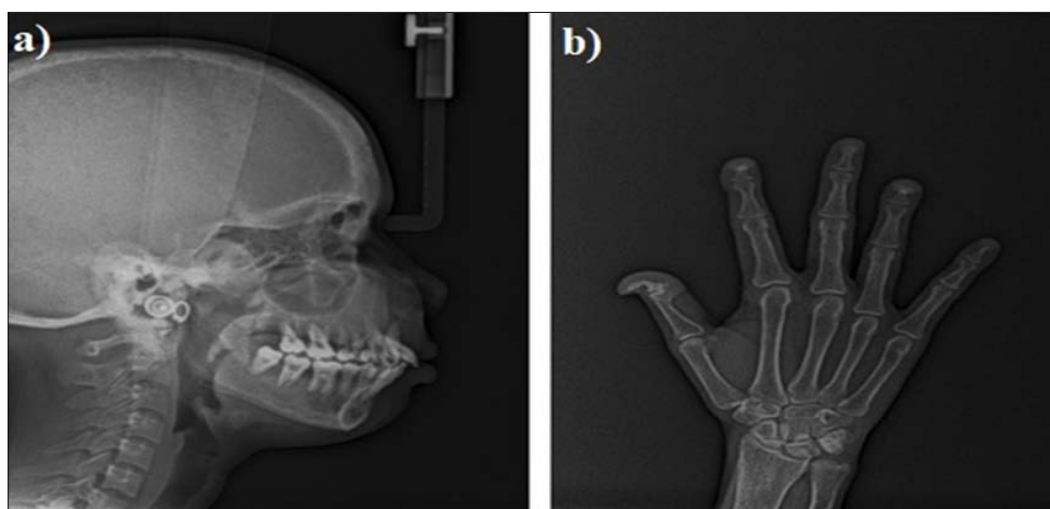


Fig 2: a) Lateral cephalogram showing multiple tiny osteolytic changes over the entire skull calvaria; b) Hand wrist radiograph showing bending of terminal phalanges of the thumb, index, and ring finger

The CBCT report of the patient shows a well-defined unilocular radiolucent lesion of soft tissue density about the left maxillary region. The lesion is roughly round in shape and size (5x5x5cm) seen extending anteroposteriorly from the left maxillary central incisor to the maxillary tuberosity region and superior-inferiorly from the infraorbital rim to the alveolar crest of the involved tooth along with buccal and palatal cortical plate expansion. Medially, the lesion seen

extending into the left nasal cavity involving middle and inferior turbinate causing partial obliteration of the left ethmoidal sinus and laterally eroding the zygomatic arch. Superiorly, the lesion extending into the left maxillary sinus causing complete obliteration. Displacement of tooth and root resorption was present (Figure 3). Overall features are suggestive of the benign osteolytic lesion about the left maxillary region.

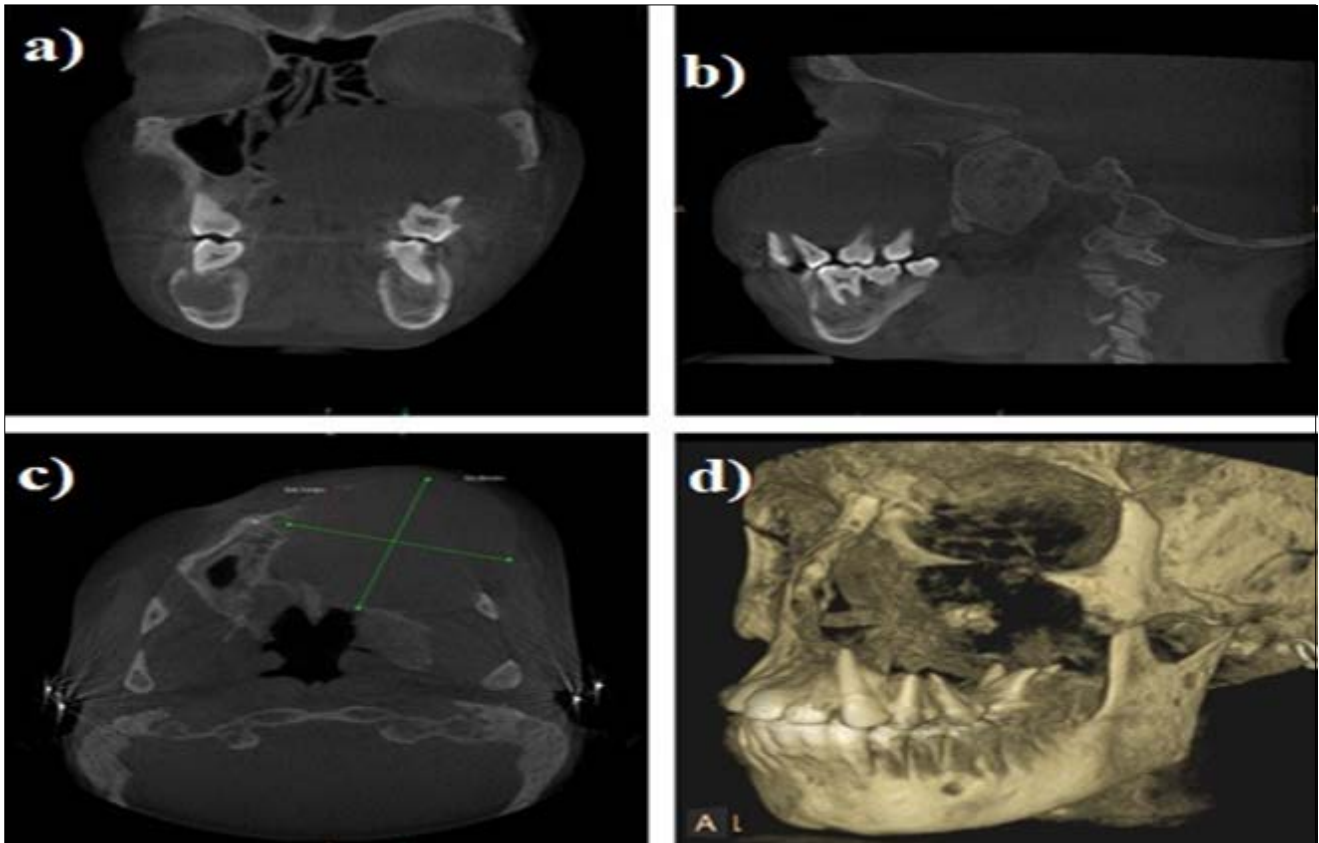


Fig 3: CBCT images showing lesion in a) Coronal section showing obliteration of nasal cavity and ethmoidal sinuses; b) Sagittal section showing displacement of tooth and root resorption; c) Axial section; d) 3D view

On CECT, a large well-defined heterogeneously enhancing lesion with peripheral calcification measuring 5.4x5.2x4.5cm is seen centred over the left palatine process of the maxilla involving the left maxillary sinus. The lesion shows multiple non-enhancing areas suggestive of cystic changes. Laterally, the lesion is seen eroding the zygomatic arch on the left side and overlying subcutaneous tissue. The lesion is also involving the buccal mucosa-buccinator complex and superior and inferior gingivobuccal sulcus. Medially, the lesion is seen extending into the left nasal cavity involving the middle and inferior turbinate, nasal septum. The lesion is also causing subtle erosion of the vomer bone on the left side. Inferiorly, the lesion is seen protruding into the oral cavity and is seen involving the alveolar process of the left side of the maxilla. Superiorly, the lesion is seen eroding the floor of the left orbit involving the infraorbital canal. Anteriorly, the lesion is eroding the anterior wall of the left maxillary sinus, and nasal spine and is reaching up to alar fibrofatty tissue on the left side. Overall features are suggestive of primary neoplastic etiology. Multiple punched-out lytic lesions are noted involving the entire bony calvaria, left occipital bone,

pterygoid fossa, medial and lateral pterygoid plates, and greater and lesser wing of the sphenoid, squamous part of temporal bone suggestive of metastasis. A soft tissue density random pulmonary nodule measuring 2x3 mm is noted in the apical segment of the right upper lobe suggestive of pulmonary metastasis. Multiple subcentimeter-sized lymph nodes are noted in bilateral level 1b. Multiple lytic sclerotic areas are noted involving ribs, vertebral bodies, humeral head, and scapula suggestive of metastasis.

Incisional biopsy of the mass shows large pleomorphic spindle to oval tumour cells with large pleomorphic hyperchromatic nuclei and foci of osteoid seen. Areas of necrosis, tumour giant cells, and abnormal mitosis are seen as suggestive of Giant cell-rich tumours. Excision of mass along with the extraction of 12, 11, 21, 22, 23, 24, 25, 26, 27 was done under general anaesthesia and sent for pathological evaluation. Histopathologically, the tumour comprised the spindle to oval cells with regular spindle-to-oval nuclei with plenty of osteoclast-like giant cells distributed uniformly throughout suggestive of giant cell tumour (Osteoclastoma), (Figure 4).

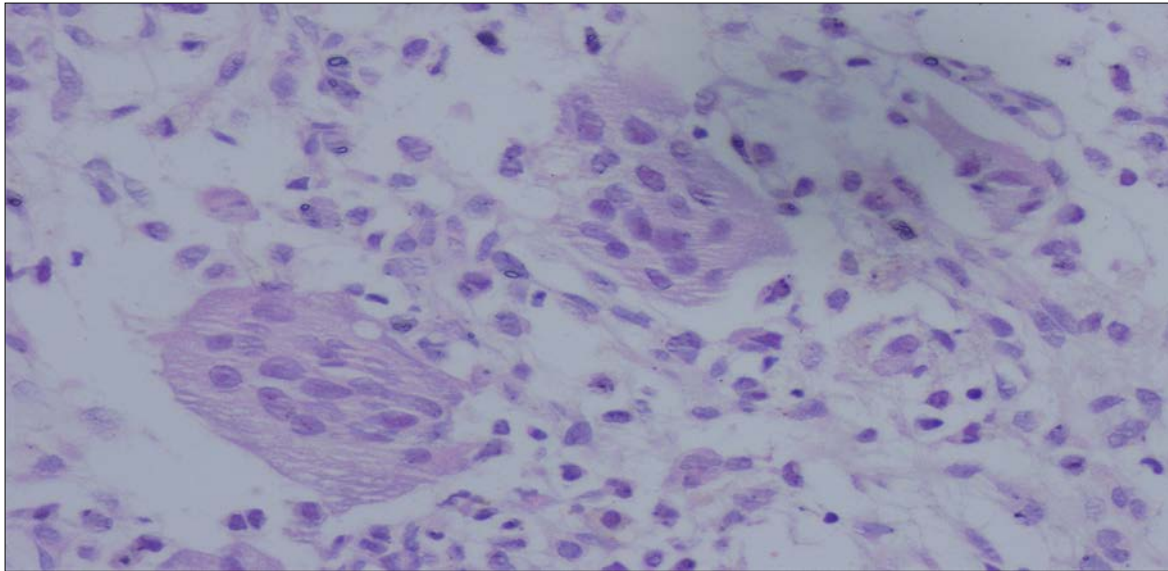


Fig 4: Histopathological photograph showing large pleomorphic spindle to oval tumor cells with large pleomorphic hyperchromatic nuclei, foci of osteoid, areas of necrosis, tumor giant cells, and abnormal mitosis

The block was sent for immunohistochemistry which shows a positive CD68 marker. The patient was under observation for 2 weeks after surgery and antibiotics, analgesics, iron supplements, and a protein diet were given as post-operative

medications. After 2 months of follow-up, the surgical site healed completely and the patient is under rehabilitation procedure at present (Figure 5a and 5b).

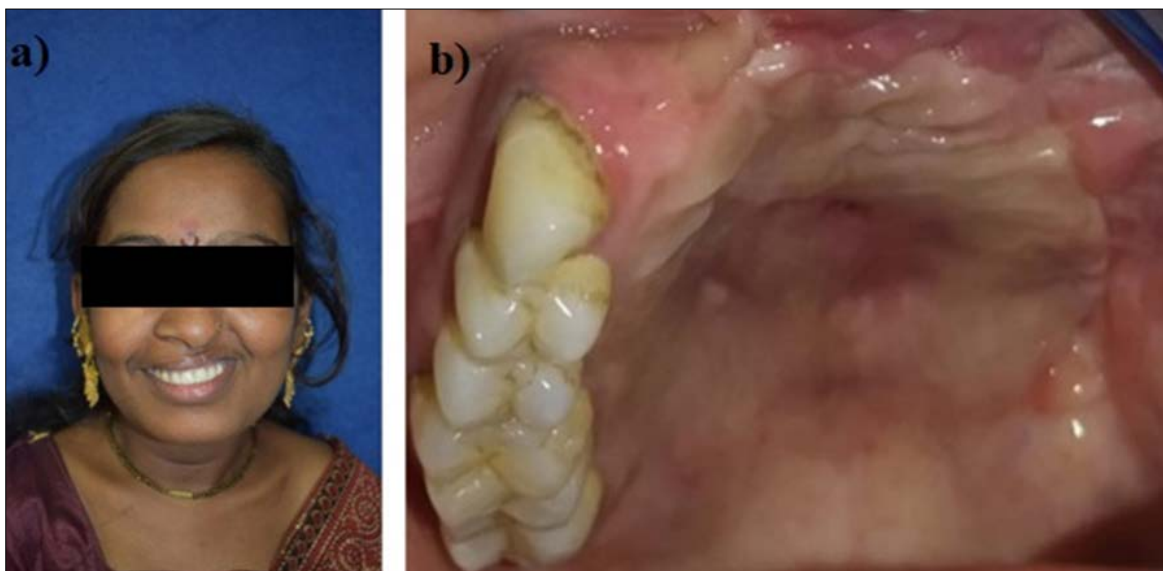


Fig 5: a) and b) Post-operative Photograph

Discussion

Osteoclastoma or giant cell tumor is a rare, benign primary osteolytic neoplasm of long bones. Even though, it is benign mostly it shows local aggressiveness. It affects the epiphysis of long bones and accounts for 5-9% of all primary bone tumours. This tumour's stromal cells are malignant osteoblastic cells that display many osteoblast cell markers like alkaline phosphatase and osteocalcin [6]. Although the pathogenesis is uncertain, it is believed to be caused by osteoblast-like mononuclear stromal cells overexpressing the Rank/Rankl signalling pathway. Monocytic pre-osteoclast cells are converted to osteoclast cells as a result of this overexpression [7]. These osteoclasts start absorbing the bone resulting in osteolysis seen with these tumours. It is usually occurring in the 30-50 years of age group with mild predominance in females. Our case also occurred in a 30-year-old young female patient.

Although there are more cases among people with Paget disease, which often originate in the pelvis or cranial bones, risk factors are not well known. These patients will exhibit high levels of alkaline phosphate as a result of aberrant laboratory readings. A Giant cell tumour of bone without Paget's disease typically manifests as a single lesion, most frequently affecting the distal femur and proximal tibia (50-65%), then the distal radius, sacrum, and vertebral body. In the head and neck region, its occurrence is very rare and accounts for only 2% of cases with the majority of them seen in the sphenoid sinus, ethmoidal sinus, and temporal bones [8]. Our case comes under this 2% rare category and the lesion was present in the maxillary bone.

Osteoclastoma patients frequently exhibit gradually worsening swelling, either with or without pain. More than three-fourths of the patients had noted swelling of the affected region. The presenting symptoms are otalgia, headache,

hearing loss, and dysfunction of cranial nerves. Less common symptoms are weakness, and joint motion limitation and about 12% of the affected reveal pathological fracture at the time of diagnosis. If a tumor cause weakening of the bone, it leads to pain and pathological fracture. In our case patient came with the complaint of progressive swelling without the association of pain and showed a sign of bending of terminal phalanges. A plain radiograph can be used to differentiate it from other odontogenic cystic lesions that commonly occurred in the maxilla. CT scan is very useful to assess the extent of the disease. Campanacci graded both primary and recurrent tumors radiographically as follows:

- **Grade I:** Tumor whose border is a thin rim of mature bone, with an intact cortex or slightly thinned out without deformation.
- **Grade II:** Tumor with well-defined margins but no radiopaque rim. The cortex along with re-active bone is thin or moderately thickened. Grade-II lesions with a fracture are graded separately.
- **Grade III:** A tumor with fuzzy borders caused by rapid growth leading to the soft tissues swelling clinically which is not bordered by reactive bone.

According to this classification, our case lies under Grade III. The confirmatory diagnosis of this tumor is made based on a biopsy. The histology of osteoclastoma reveals three types of cells i.e., interstitial fibroblasts, monocytes or macrophages, and multinucleated giant cells. The basic proliferating cell has a round-to-oval or even spindle-shaped nucleus in the field, which is diagnostic of a true giant cell tumor. The giant cells are typically distributed uniformly throughout the lesion, with the nucleus surrounded by an ill-defined cytoplasmic zone and the absence of a discernible intercellular substance. Mitotic figures can be seen, sometimes in great numbers. They usually contain 40-60 nuclei. Areas of infarct-like necrosis are common in giant cell tumors. Some tumors are almost completely necrotic. There is no inflammatory reaction connected to the necrosis. Small clusters of foam cells are typical. In our case, the histopathological report revealed spindle-to-oval cells with regular spindle-to-oval nuclei with plenty of many osteoclast-like giant cells distributed uniformly throughout the lesion.

If the tumor presents in an accessible site, then wide resection surgery is the treatment of choice. Curettage and Pharmacotherapy are other commonly used techniques. Pharmacotherapy for osteoclastoma includes bisphosphonates such as Zolidronate, which induce apoptosis in the MNGC (multi-nucleated giant cell) fraction, thus preventing tumor-induced osteolysis. Recently, humanized monoclonal antibodies such as Denosumab are used to target the RANK ligand (RANKL) as increased expression of Rank ligands by stromal cells plays a role in tumor pathogenesis^[9]. The recurrence rates of this lesion are reported as high as 50%. To reduce these recurrence rates, local adjuvants such as aqueous zinc chloride, PMMA, phenol, argon beam coagulation, or cryotherapy have been utilized. The overall prognosis of osteoclastoma is good, though it shows local recurrence. It often has a poorer prognosis when it shows malignant transformation^[10]. Metastasis varies between 0% and 9%. The lung is the most common site. Other less common sites include the brain, kidneys, bone, skin, and lymph nodes^[11]. Pulmonary metastasis was first reported by Flinch and Gleave in 1926. A combination chemotherapy regimen with Ifosfamide, Adriamycin, and dacarbazine can be given, considering the aggressive nature of the tumor. Even if

complete remission of pulmonary metastatic disease has been achieved in some studies after chemotherapy, prophylactic whole lung irradiation and chest computed tomography (CT) monitoring is recommended in all patients^[12]. Our patient is under observation and oral rehabilitation procedures. Usually, a CT scan of the primary site is performed three months after treatment, twice a year for the following two years, and then once a year for the following five years. Based on clinical findings, follow-up frequency and length should be decided.

Conclusion

Even though the occurrence of Osteoclastoma is rare in the oral and maxillofacial region, it has to be considered as one of the differential diagnoses of swelling of the maxillofacial region and the diagnosis should be made depending upon the proper history, clinical examination, systemic conditions, radiographic and histopathological features. On account of the high recurrence rate and chances of metastasis, regular surveillance has to be done depending on the prognostic condition.

Conflict of Interest: Not available

Financial Support: Not available

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