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Ewing sarcoma of the mandible mimicking an odontogenic abscess: A case report

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ABSTRACT

Ewing Sarcoma (ES) of the mandible is a rare malignancy with poor prognosis which can be misdiagnosed as odontogenic inflammatory lesions or infections. We present a 7-year old male child which had been previously misdiagnosed and treated as an odontogenic abscess. This reported case describes the clinical, radiological and histopathological findings of oral ES with difficulties in management.

Key words: Ewing's Sarcoma, Malignancy, Mandible, Odontogenic Infections.

Introduction

Ewing's Sarcoma (ES) is an uncommon malignant neoplasm which primarily affects the skeletal system [1]. ES accounts for 4 to10% of all bone tumors, and commonly involves the lower skeleton, i.e. long bones and the pelvis [2-4]. ES involving the facial skeleton is quite rare (less than 3% of all ES), with the mandible being the most commonly affected facial bone [5-10]. The mean age of occurrence of ES in head and neck region is 10 years, generally in white population and male sex [1, 2]. It affects mainly adolescents and young adults and is rarely seen before the age of 5 [7]. The common presenting signs and symptoms of ES such as swelling, pain and sensory disturbances are rather unspecific and can sometimes be misleading [11, 12]. This paper reports a rare case of ES of the mandible in a 7-year-old boy, which had been previously misdiagnosed and treated as odontogenic abscess.

Case Report

A 7-year-old boy presented with a 3-weeks history of pain and rapidly progressive swelling of his right mandibular angle and buccal space. Assuming that the tumor was an acute odontogenic abscess of buccal space due to caries of tooth 4E the attending dentist incised the tooth and

prescribed antibiotics. However, the pathologic process of the right mandible enlarged constantly (increasing of pain and swelling) and the patient was referred to our department for further investigations. On admission the patient was in good general condition. Patient had also no history of fever, dehydration, weakness, local or general lymphadenopathy, or change of body weight.

Physical examination revealed a solid 6-cm-diameter asymmetric swelling of the buccal cortex area of the right first mandiblular molar extended into mandibular buccal vestibule (Fig. 1 and 2). However, maximum intraincisal opening was normal (about 32 mm). There were no evidences of trismus, trigomandibular/ parapharyngeal, retropharyngeal spaces involvement, dysphasia and ovula deviation. Despite the second mandibular right molar suprairruption and its floating, there was no evidence of vascular lesions such as Thrill and Bruit.

Panoramic radiograph taken before the tooth extraction by the attending dentist showed E caries, and a diffuse radiolucency lesion with ill-defined borders extending from distal of right deciduous first premolar to distal of right second molar follicle. A poorly defined osteolytic lesion in the right side of the mandible was also seen. The most important panoramic finding was superimposition of the first molar, seventh dental follicle, and the floating in air appearance which confirmed the clinical findings of mobility and pumping of the first molar (Fig. 3 A, B and C).

To more accurately depict the lesion, sonography and computed tomography (CT) scan were performed. CT showed a marrow destroying mass of approximately 4 ×2.5 cm of size associated with cortical plate destruction of the right buccal of the mandible and penetrating the lesion into the buccal and vestibular soft tissue (Fig. 4).

Laboratory examinations showed normal WBC count (9700 cell/mm^3), significant elevated C-reactive protein (CRP) (2.7 mglL), elevated erythrocyte sedimentation rate (ESR) (80 mm/hr) and elevated high-density lipoproteins (HDL) (305 /L). Incisional biopsy identified ES (grading according to FNCLCC: grade 3; diagnosis confirmed by Prof. Leuschner, sarcoma reference centre, Kiel and Prof. Jundt, bone tumour reference centre, Basel).

Histologically, the regular sheets was composed of uniform small round cells arranged in a fibrovascular Struma. The cells had hyperchromatic nuclei with scanty cytoplasm without mitotic forms or necrosis (Fig. 5). An immunohistochemical examination was performed and revealed positivity for CD99, pancytokeratin and vimentin. Histopathological and immunohistochemical findings supported the diagnosis of ES. Additional CT-,neck MRI- and CRX- and abdomen/ CT-scans showed no evidence of metastasis or primary lesion (Fig. 3). After complete examinations and having differential diagnosis of mesanschimal sarcoma, osteogenic sarcoma, LCH and PNET, the patient underwent general anesthesia in order to incisional biopsy.

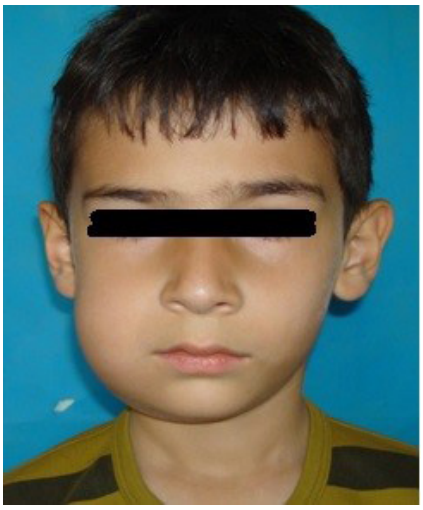


Figure 1. Extra-oral presentation of tumor, showing significant increase in angle of right mandible.

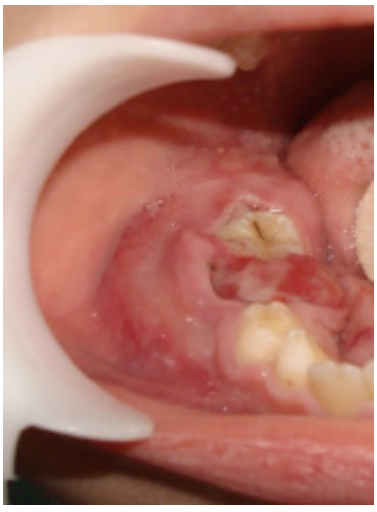


Figure 2. Intraoral aspect of the tumor, showing a tender and swollen erythematous mass on the mucosa in the molar region of the right mandible.

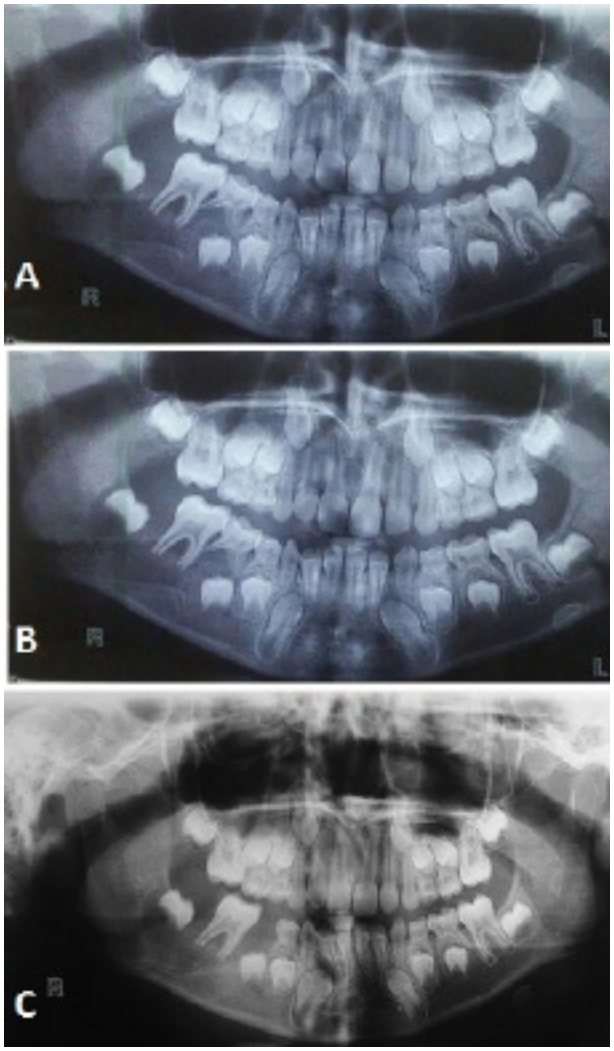
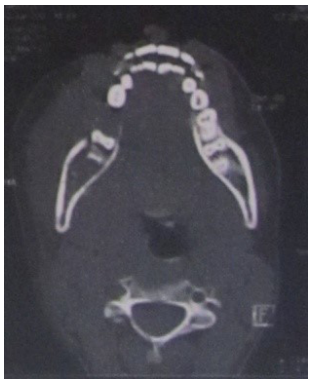
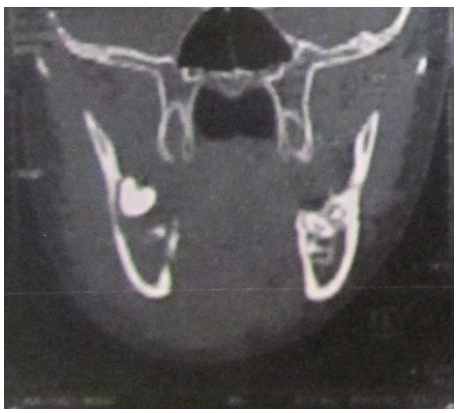


Figure 3. Panoramic radiograph.





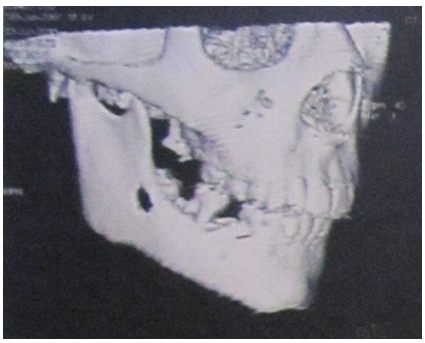


Figure 4. CT scan.

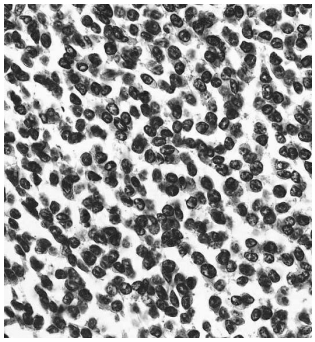


Figure 5. Histological picture.

Discussion

ES is a rare malignant neoplasm that comprises approximately 4-10% of the primary bone tumors. Occurrence of this tumor in the mandible is rare, but this is the primary site for this disease in the head and neck. Clinical findings of ES in the facial regions have been nonspecific and usually involve rapid growth, swelling, and pain. Moreover, when mandible is involved, loosening of teeth, middle ear infection, and paresthesia are common. Systemic symptoms such as fever, weight loss, leukocytosis, and elevated ESR and CRP are also seen in some patients. These symptoms may be the first signs of oral ES, which are also seen in odontogenic infections. In this case, the patient visited the dentist with symptoms of pain, swelling, and abscess similar to odontogenic infections, and had his tooth removed due to the wrong diagnosis.

Although osteolytic change in this tumor was seen, but this is not a pathognomonic feature because some other lesions such as neuroblastoma, osteogenic sarcoma, histiocytosis X, and osteomyelitis have the same patterns [2,9]. The presence of onion peel or sun-ray spicules of periosteal bone have been described as the commonest radiological features for ES, but neither was present in this case [13].

Although conventional Xray is still used for bone lesions, other imaging methods like CT scan and

magnetic resonance imaging (MRI) are highly recommended for accurate evaluation of lesion's progress on soft tissue and bone invasion [11]. In this case, axial and cronal CT with 3D reconstruction was useful in detecting the intraosseous tumor source.

The histopathological presentation of ES includes layers of small poorly differentiated cells with unclear boundary and little cytoplasm with round or oval nucleus. In 75% of cases, due to the presence of intracytoplasmic glycogen granules, Periodic Acid Schiff (PAS) staining is positive, which helps the diagnosis, but is not specific, as other tumors with small round cells also turn PAS positive [14]. Due to this similarities between many malignant tumors, the lesion must be differentiated from other small round cell tumors, such as small cell osteosarcoma, mesenchymal chondrosarcoma, embryonal rhabydomyosarcoma, neuroblastoma and lymphoma. The use of immunohistochemistry has helped in the diagnosis of this tumor [3]. Usually, the tumor cells are positive for vimentin, CD99 and pancytokeratin and negative for neural, skeletal, vascular and lymphoid cell markers [8,15]. In the case presente ed, demonstration of intracytoplasmic glycogen by PAS-diastase stains and immunohistochemical staining positivity for vimentin, CD99 and pancytokeratin confirmed the diagnosis of ES.

It has been reported that the best approach treatment for ES include surgery, radiotherapy and chemotherapy [16,17]. In the present patient, he received multidrug chemotherapy (i.e. Doxorubicin, Actinomycin, Vincristine, Cyclophosphamide) which has an important role in managing metastases, the main concern in this malignancy [18]. No radiation was planned as the possible effects on jaw growth. According to the reports, chemotherapy can increases survival from 10 to 75% [19]. After chemotherapy, the tumor will shrink and proved to be resectable. The patient will have the posterior half of the right side of the mandible removed and the entire tumor will be resected.

Conclusion

Ewing's Sarcoma is a rare tumor in oral cavity which can mimic odontogenic infections. Thus, it should be carefully examined clinically, and because of similarity in microscopic presentation with other small round cell tumors, the diagnosis is often difficult. For such an aggressive large lesion, early biopsy is recommended. Long term follow up is necessary to see the outcome of the lesion.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Competing Interests

The authors declare any competing interests.

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