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Ewing Sarcoma of The Mandibular Ramus: Case Report

Mandibular Ramusun Ewing Sarkomu: OlguSunumu

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ABSTRACT

Objective: Since Ewing sarcomas are highly aggressive tumors, early diagnosis is very important for the prognosis of these patients. The aim of this case report is to raise awareness among pediatricians and dentists about the clinical, radiological, and histopathological features of these aggressive tumors. A 10-year-old child patient referred to our clinic from the ENT (Ear-Nose-Throat) outpatient clinic was found to have a sore throat that had been persisting for about 2-3 months. In addition, it was observed that there was an extraoral swelling in the right mandibular ramus region that had started to be noticed extraorally for the last 15 days. Panoramic radiography showed a multilocular lesion involving the mandibular ramus. The lesion was also evaluated by conventional CT (Computed Tomography), MRI (Magnetic Resonance Imaging) and ultrasonography. A biopsy was taken from the patient and a diagnosis of Ewing's Sarcoma was made and chemotherapy treatment was started by the Pediatric Oncology Department. The oral findings of ewing sarcomas, which rarely affect the mandibular region, should always be considered for the possibility of confusion with periodontal diseases, odontogenic and pericoronal infections. Therefore, additional and appropriate radiographic examinations should be used to make a definitive diagnosis.

Keywords: *Computed tomography, Ewing sarcoma, Mandible, Panoramic radiography, Ultrasonography*

ÖZET

Giriş: Ewing sarkomları oldukça agresif tümörler olduğundan, erken tanı bu hastaların prognozu için çok önemlidir. Bu olgu sunumunun amacı, çocuk doktorları ve diş hekimleri arasında bu agresif tümörlerin klinik, radyolojik ve histopatolojik özellikleri hakkında farkındalık yaratmaktır. KBB (Kulak-Burun-Boğaz) polikliniğinden kliniğimize sevkedilen 10 yaşındaki çocuk hastanın yaklaşık 2-3 aydır devam eden boğaz ağrısı olduğu tespit edildi. Panoramik radyografide mandibular ramusu tutan multiloküler bir lezyon görüldü. Lezyon konvansiyonel BT (Bilgisayarlı Tomografi), MRG (Manyetik Rezonans Görüntüleme) ve ultrasonografi ile de değerlendirildi. Hastadan alınan biyopsi sonucu Ewing Sarkomu tanısı konularak Pediatrik Onkoloji Bölümü tarafından kemoterapi tedavisi başlandı. Nadiren mandibular bölgeyi etkileyen Ewing sarkomlarının ağız içi bulguları periodontal hastalıklar, odontojenik ve perikoronal enfeksiyonlarla karışabilme olasılığı açısından her zaman gözönünde bulundurulmalıdır. Bu nedenle kesin tanı koymak için ek ve uygun radyografik incelemeler kullanılmalıdır.

Anahtar kelimeler: *Bilgisayarlı tomografi, EwingSarkomu, Mandibula, Panoramik radyografi, Ultrasonografi*

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INTRODUCTION

Ewing sarcoma (ES) is a skeletal malignant bone tumor that originates from neurons. ES is also one of the most aggressive bone tumors in childhood (Balkaya et al., 2017). These tumors originate from undifferentiated cells of the bone and are more common in young adults and adolescents (Ahuja et al., 2019). It is more common in the pelvis, tibia, femur, and costae. ES is also defined as a soft tissue tumor seen in the chest wall, pleural cavities, gluteal and cervical muscles. While treatment efficacy has been observed to be higher in cases with local course, survival rate has been observed to be quite low in metastasized cases (Gaspar et al., 2015). ES, which is usually seen in white children and adolescents, is the most common bone tumor after osteosarcoma. It is also the most undifferentiated form of neuroectodermal tumors (Soni et al., 2019). It has been reported that mandibular ES are extremely rare and only 1% of cases show mandibular development (Hatim et al., 2022).

In this case report, ultrasonographic (US), computed tomography (CT), positron emission tomography (PET), panoramic radiography and magnetic resonance imaging (MRI) findings of ES extending from the mandibular ramus to the condyle will be discussed.

The aim is to increase the awareness of dentists in early diagnosis, although it is rare in the jaws. In our case, we present the character of this lesion, which is difficult to recognize in two-dimensional images such as panoramic radiography, in different image modalities.

CASE PRESENTATION

A 10-year-old girl was admitted to our clinic with swelling on her right cheek, which had been noticed extraorally for about 10-15 days. The patient was consulted to our ENT (Ear, Nose, and Throat) outpatient clinic because of a complaint of sore throat for several months, lip pain and recent onset of paresthesia sensation in the lip. Intraoral examination revealed a marked decrease in mouth opening and a rubbery swelling on palpation in the right buccal region. Extraoral examination revealed swelling starting from the angulus of the mandible and extending to the external auditory canal (Figure-1).

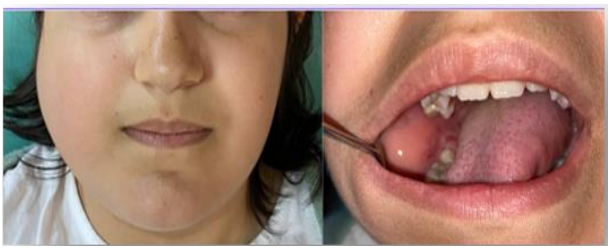

Figure 1. Extraoral-intraoral image of the patient.

MRI showed that the lesion was 6.4×4.4×4.4×5.9 cm in size and extended to the parotid and parapharyngeal area surrounding the mandibular ramus and had intense contrast uptake. The lesion extended to the condylar process and posterior corpus of the mandible and also extended to the masticator space, retromolar trigone and buccal deep fat pad. Isointense appearance on T1 sequence and intermediate signal intensity on T2 sequence was observed (Figure-2).

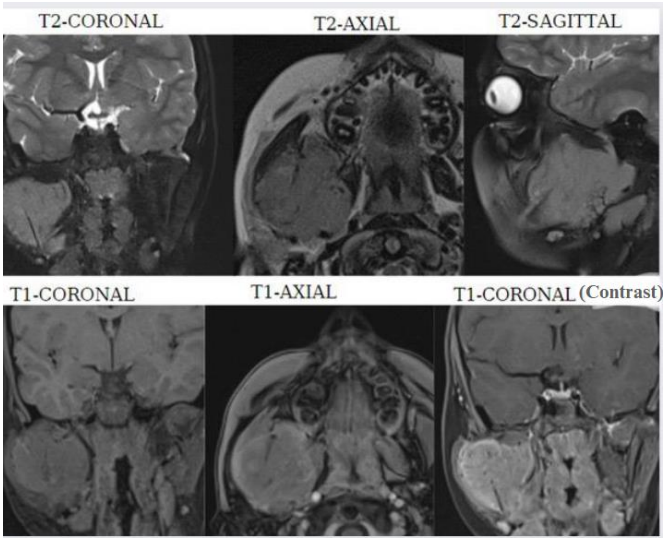

Figure 2. Coronal, axial and sagittal MRI of the lesion.

On US examination, an isodense, heterogeneous solid mass with increased vascularization was observed adjacent to the mandibular ramus and causing erosion of the bone. In addition, perisosteal reaction was clearly observed on US imaging. (Figure-3).

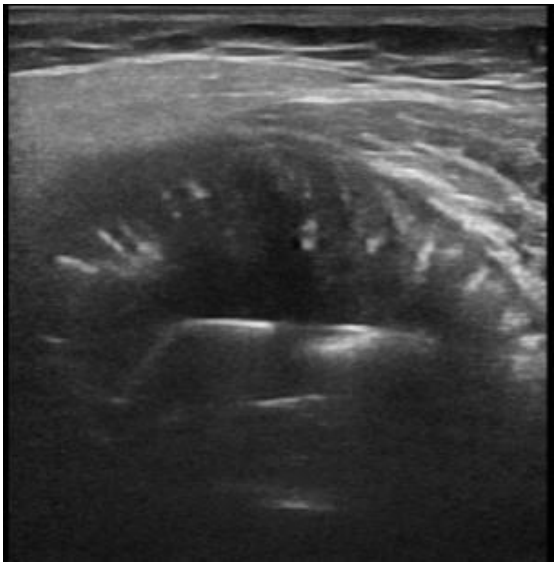

Figure 3. Ultrasonographic image of a heterogeneous solid mass with localized destruction adjacent to the mandibular ramus.

CT images obtained from the patient showed a sunburst type periosteal reaction in the mandibular ramus. In addition, there were areas of bone destruction located in the center of the mandibular ramus, destroying both buccal and lingual bone and extending to the mandibular condyle (Figure-4).

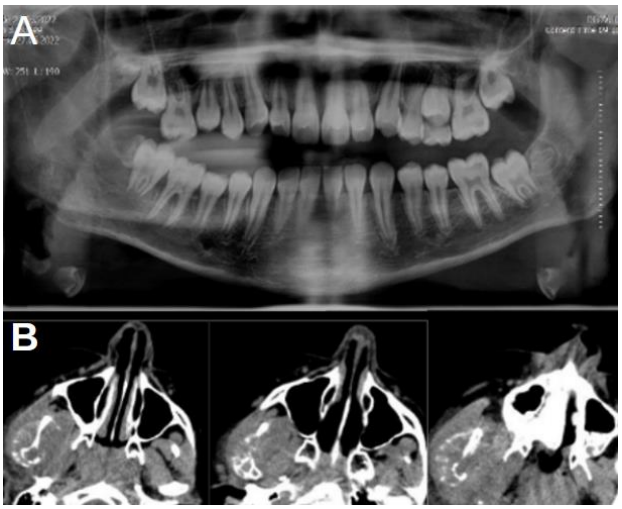

Figure 4. A-Panoramic radiograph and B-axial section CT images of the lesion showing sun-burst periosteal reaction.

In addition, two slightly asymmetric lymph nodes in the right retropharyngeal region and enlarged lymph nodes on the right and left sides were observed at Level 2. It was observed that the roundness index of the lymph node on the right side increased at Level 2 and was defined as suspicious. Panoramic radiography showed areas of lytic bone in the mandibular ramus with non-cortical borders (Figure-4). Histopathologic examination revealed cavities separated by fibrous septums infiltrating the bone tissue and neoplastic cells with open chromatin, round nuclei, narrow cytoplasm, and neoplastic cells lining these cavities. Tumor cells were observed to be solid, lining the cavities in some areas and perivascularly distributed in others. The presence of remarkable hyanilization in the stroma was mentioned. In addition, FISH examination showed that 30-35% of the neoplastic cells showed separation of red and green signals. This finding supported the presence of translocation involving the EWSR gene in neoplastic cells. This is accepted as a pathognomic finding in the histopathologic diagnosis of ES (Mishra et al., 2022). In the PET examination of the patient, pathologic FDG uptake was observed in the mass destructing the right mandibular ramus-angulus filling the infra-temporal fossa, masticator fossa and buccal area. In addition, FDG uptake was observed in bilateral cervical lymph nodes with the possibility of metastasis, with relative prominence on the right.Informed consent was obtained from the patient's parents for the photographs taken from the patient.

DISCUSSION

ES causing displacement of teeth or tooth germs are radiologically defined as poorly circumscribed osteolytic lesions (Pampori et al., 2011). ES do not give very specific clinical findings in the first stage. However, it has been observed that the most common clinical findings are swelling and pain (Davido et al., 2011). In this case report, the first clinical finding of the patient was sore throat followed by swelling. In addition, while ES affect males more frequently, a case in a female patient was described in this case report. Brazao-Silva et al. described periosteal reactions and osteolytic bone lesions with a sun-ray-like appearance as the most common radiologic findings of ES (Brazão-Silva et al., 2010). In this case report, sunburst periosteal reaction and cortical bone destructions in the mandibular ramus and lytic lesions extending from the ramus to the condylar process were noted on CT images.

Ahuja et al. mentioned the isodense appearance of the soft tissue mass and the erosion and periosteal reactions in the bone in the US examination of a case of ES in the mandible (Ahuja et al., 2019). In this case, erosion areas and sunburst type periosteal reactions caused by the solid heterogeneous mass adjacent to the mandible on the ramus cortical bone and enlarged submandibular lymph nodes on the same side were observed.

Margaix et al. (2017) showed that 69% ES cases which are fundamentally located presented between 1960 and 2014 was the mandible (Margaix-Muñoz et al., 2017). It has been reported that the majority of cases are female and the first complaint is swelling and pain (Margaix-Muñoz et al., 2017). In this case report, our patient was female, similar to the literature. Also similar to the literature, the patient's first complaint was pain followed by swelling.

It has been reported that it is very difficult to differentiate tumors such as malignant lymphoma, metastatic carcinoma, osteosarcoma, aggressive central giant cell granuloma, osteomyelitis, histiocytosis X, Burkit lymphoma and rhabdomyosarcoma in the mandible in children (Oliveira et al., 2019; Takami et al., 2020). Osteosarcomas and other multilocular radiolucent lesions of the jaw are extremely rare and may show osteolytic bone patterns at various levels. Therefore, their differential diagnosis is difficult radiologically and histopathologic examination is required for definitive diagnosis (Chaudhary et al., 2012).

Also periosteal reaction can be seen in all of these pathologies, but intraoral soft tissue growths differentiate ES from eosinophilic granuloma and osteomyelitis. In addition, the age of the patients is also very effective in the differential diagnosis. In patients under 5 years of age, neuroblastoma is also on the list of possible diagnoses, whereas neuroblastoma is excluded in patients over 5 years of

age (Sinha et al., 2014). However, MRI and CT are considered to be the most effective imaging methods in the diagnosis of malignancies such as ES (Lopes et al., 2007). In addition, it has been reported that ES may be mostly confused with dental infections and pericoronal infections and even cases misdiagnosed as pericoronitis (Margaix-Muñoz et al., 2017, Takami et al., 2020).

Histopathologically, small round, hyperchromatic cells are observed. These cells have distinct borders and uniform nuclei and are arranged in large layers. In addition, fibrovascular septa separating the tumor into lobular areas are remarkable (Suhag et al., 2012). Huang et al. showed that 8 of 60 patients with ES had p53 gene mutation and the survival rate of patients with this mutation was lower compared to other patients. The presence of this mutation has been accepted as a very important factor in determining the prognosis of these patients (Huang et al., 2005). In addition, it has been reported that the survival rate of ES in the head and neck region (80%) was better than ES in other anatomical regions (56%) during the three-year follow-up period (Bölling et al., 2013; Yogesh et al., 2018).

ES seen in children are treated with local surgery after chemotherapy. Radiotherapy is not preferred in these patients due to possible side effects (Oliveira et al., 2019). In these patients, pre-treatment chemotherapy prevents micro metastases and reduces tumor burden (Ahuja et al., 2019). However, distant metastases of ES seen in the jaw are quite low. Species seen in other anatomical regions have been reported to be more prone to metastasis (Owosho et al., 2016; Yogesh et al., 2018). In addition, factors such as age, gender and radiation have not been found to be effective on the prognosis of these tumors (Martin et al., 2019).

ES, which rarely involve the head and neck region, are aggressive tumors. Early diagnosis of these tumors will prevent possible metastases. Since the survival rate is quite high in cases where the primary site of involvement is the head and neck region without metastasis, the mastery of clinical and radiological findings of these lesions by pediatricians and dentists will positively affect the prognosis.

Conflict of Interest: There is no conflict of interest between the authors regarding the writing and editing of this case report**.**

Ethical Approval: Informed consent was obtained from the patient's parents for the photographs taken from the patient.

This case report was presented as a poster at the 4th International Congress of the Society of Oral and Maxillofacial Radiology (October 19-23, 2022).

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