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Case Report

Ewing's sarcoma of mandible: A case report presenting as odontogenic infection

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ABSTRACT

Ewing's sarcoma is a small, round and blue cell malignancy that is primarily affecting skeletal system. Ewing's sarcoma is an aggressive tumor showing rapid growth and metastasis with complex diagnosis. Because of the rarity of tumor involving the mandible, it was usually misdiagnosed for long time as odontogenic infection. We report a case of Ewing's sarcoma involving mandible, to make the clinicians aware of this unusual clinical and histopathological spectrum of this rare tumor.

1. Introduction

Ewing's sarcoma (ES) was first described by James Ewing in 1920 as a diffuse endothelioma of bone [1]. Only 2.3% of the lesions are seen in the head and neck region [1,2]. The mean age of occurrence in the head and neck region is 10.9 years. ES generally affects population with male/female ratio of 1.3–1.5:1 [3]. Immunohistochemistry and molecular assays for chromosomal translocation are the main stay of diagnosis. It has very much unfavorable prognosis of all primary musculoskeletal tumors. Even with early intervention, patients with metastasis have approximately 20% chance of 5-year survival [4]. Here we report a case of ES involving mandible in a 12-year-old girl.

2. Case report

A 12-year-old girl patient complained of swelling on right side of face since one and a half months ago. Initially swelling was small in size and then gradually increased to present size. She experienced pain and discomfort while chewing and difficulty in opening the mouth. She gave history of hot fomentation with the swelling. She also complained of intermittent fever and weakness. Patient was on medication of antibiotics and analgesics as it was considered as an odontogenic infection.

Extraoral examination revealed that face was asymmetrical, with a single large diffuse swelling of approximately 4 cm × 3 cm on the right side of face. The swelling was extending superoinferiorly from the line passing through infra-orbital ridge to the inferior border of mandible and anteroposteriorly from the line passing through medial canthus and corner of mouth to 1 cm behind the line passing through lateral canthus of eye (Fig. 1). On palpation, the swelling was slightly tender, bony hard, nonfluctuant, noncompressible and nonreducible. Right submandibular lymph nodes were palpable. These were nontender, roughly oval in shape and approximately 1.5 cm × 1.5 cm in size.

Intraoral examination revealed an ulceroproliferative growth on the alveolar ridge extending from the distal surface of 47 to the pterygomandibular raphe, with erythematous areas (Fig. 2). It was

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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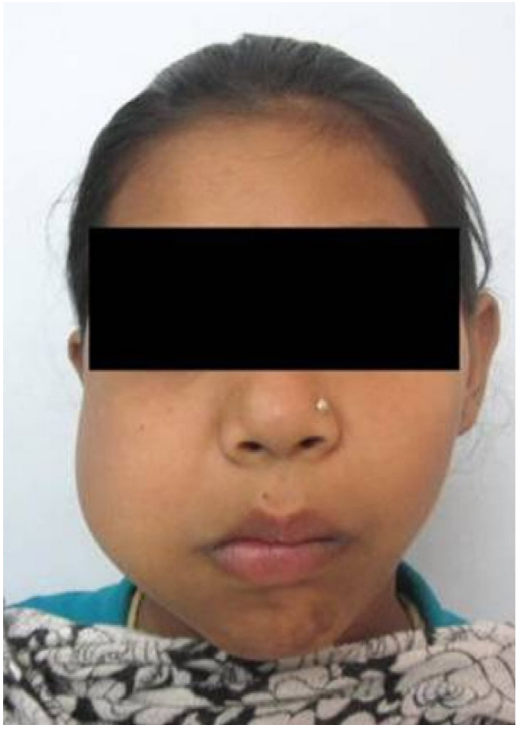

Fig. 1. Extraoral view: a large swelling on the right side of face extending from ala-tragus line to the inferior border of mandible was noted.

oval in shape, with smooth surface and well-defined margins. It was firm on palpation and slightly tender.

Routine hematological examination revealed that the patient was anemic and also showed leukocytosis and increased erythrocyte sedimentation rate (ESR).

Radiographic examination revealed an osteolytic radiolucent lesion with the right side of ramus of mandible, extending from the distal surface of mandibular second molar to the anterior ½ part of ramus of mandible, with destruction of the coronoid process. Carious 46 and developing 48 were noted (Fig. 3).

Based on all these examinations, provisional diagnosis of carcinoma of alveolus was made. Mesenchymal sarcomas and

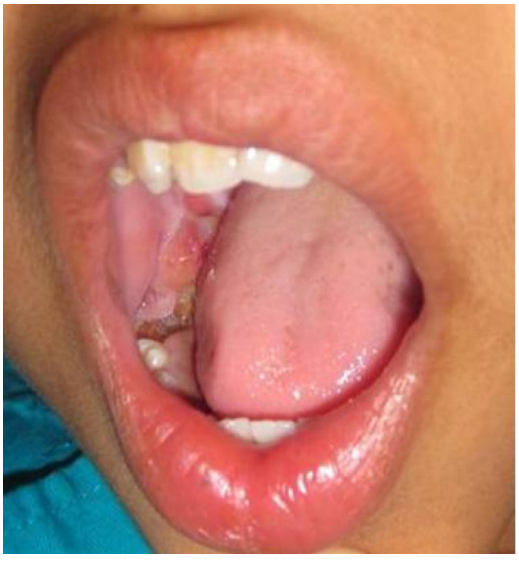

Fig. 2. Intraoral view: an ulceroproliferative growth with alveolar bone in the region of distal surface of 47, up to pterygomandibular raphe was noted.

metastatic malignant tumors were considered as differential diagnoses. Incisional biopsy of the lesion was performed and multiple tissue bits were obtained. Each bit was approximately 1 cm × 1 cm in dimension, grayish in color, and soft in consistency (Fig. 4).

H&E stained section showed the sheets of uniform small round cells arranged in the form of nests that are separated by fibrous septae (Fig. 5A). It also showed ragged resorption of bony trabeculae by tumor cells (Fig. 5B). The densely packed tumor cells have uniform, small, hyperchromatic, round nuclei with inconspicuous nucleoli, fine powdery chromatin, scanty cytoplasm and indistinct cell borders. Mitotic figures were rare (Fig. 6A). Presence of any Homer–Wright rosettes was not noted throughout the section. The Periodic Acid Schiff's (PAS) stained sections demonstrated abundant diastase-labile material, which is interpreted as glycogen (Fig. 6B). A diagnosis of malignant small round cell tumor probably Ewing's sarcoma was made based on the above histopathological findings.

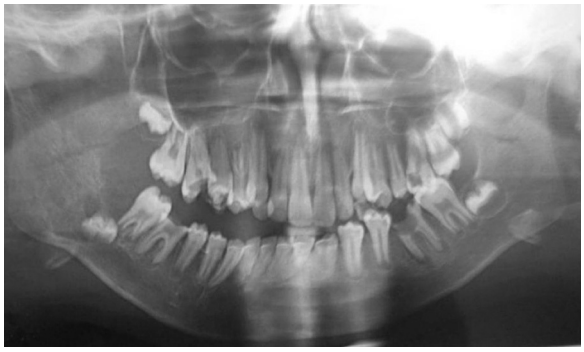

Fig. 3. Radiography: an ill-defined osteolytic lesion is seen on right side of mandible extending from distal surface of 47 to half of ramus of mandible. Destruction of coronoid process is also notable.

Immunohistochemistry is a very good ancillary to provide confirmatory diagnosis of many tumors. It is a technique for identifying cellular or tissue constituents (antigen) by means of antigen–antibody interaction, the site of antibody binding being identified either by direct labeling of the antibody, or by use of a secondary labeling method. In the present case, panel of immunohistochemical markers namely Pan Cytokeratin, LCA, CD3, CD20, myeloperoxidase (MPO), synaptophysin (SYN), chromogranin (CHR), desmin, myogenin, vimentin, CD99, and terminal deoxynucleotidyl transferase (TdT) were used to rule out other small round cell tumors.

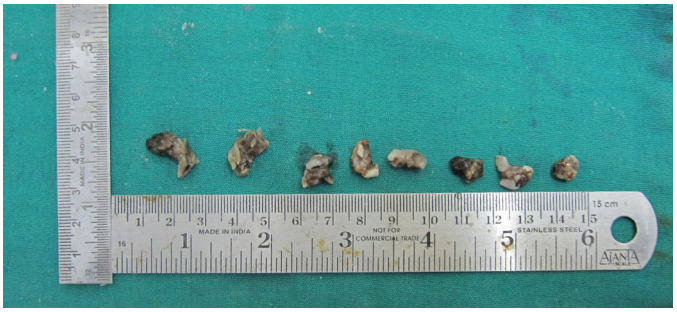

Fig. 4. Incisional biopsy: multiple tissue bits measuring approximately 1 cm × 1 cm in dimension, grayish in color, and soft in consistency were harvested.

Pan Cytokeratin is used to rule out the epithelial origin of the tumor e.g. sinonasal undifferentiated carcinoma. LCA, CD3, CD20, and MPO are used for differentiation of B cell or T cell lymphoma and myeloid leukemias. Synaptophysin (SYN) and chromogranin