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Case Report

Ewing's sarcoma of the mandible- A case report

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ABSTRACT

Ewing's sarcoma is a rare aggressive variant of small round cell tumors and is an uncommon malignancy that occurs usually in childhood. It constitutes 10–15% of all primary malignant tumors and represents the second most common malignant bone tumor occurring in children and young adults after osteosarcoma - with an estimated incidence of 2.93 cases/million inhabitants under 20 years of age/year. This paper presents a case of 10-year-old male child who reported with a rapidly progressing swelling in the right mandibular posterior ramus region. The patient was diagnosed with Ewing's sarcoma on the basis of history, clinical, radiological and confirmed with histopathological & immunohistochemical examination. Since the exact diagnosis is hard to achieve before biopsy, the condition poses a difficult diagnostic dilemma for the clinician.

1. Introduction

Ewing's sarcoma/primitive neuroectodermal tumor (ES/PNET) usually arises in long bones of extremities. It is uncommon to find ES in the head and neck region, particularly in gnathic bones. It comprises 6-8% of all primary bone malignancies.[1] Only 1% of cases are reported with jaw involvement and has mandibular predilection. Even with early intervention, patients with metastasis have approximately 20% chance of 5-year survival. James Ewing described this tumor initially in the year 1921.[2] The mean age of occurrence in head and neck region is 10.9 years. ES generally affects white population and has male predilection with M: F ratio = 1.3-1.5:1.[3] Here we report a case of Ewing sarcoma in 20 year old male patient highlighting clinical, radiological, histological and immunohistochemistry findings.

2. Case Report

A 10-year-old boy presented with swelling and pain involving the right side of the lower jaw. History revealed presence of the swelling since 2 months, which gradually increased in size.

2.1. Clinical findings

On clinical examination, solitary swelling of 4 cm in diameter was noted involving right middle and lower third of face, overlying skin was stretched (Figure 1). Intraorally, a solitary swelling was present in right premolar-molar region extending from 84to46, with vestibular obliteration. Overlying mucosa appeared normal. Swelling was tender and firm consistency (Figure 2). Right submandibular lymph nodes were enlarged, tender and mobile. Systemic symptoms such as fever, weight loss were absent. On the basis of history and clinical examination provisional diagnosis of benign odontogenic cyst/tumour on right mandible was made and differential diagnosis of Fibro

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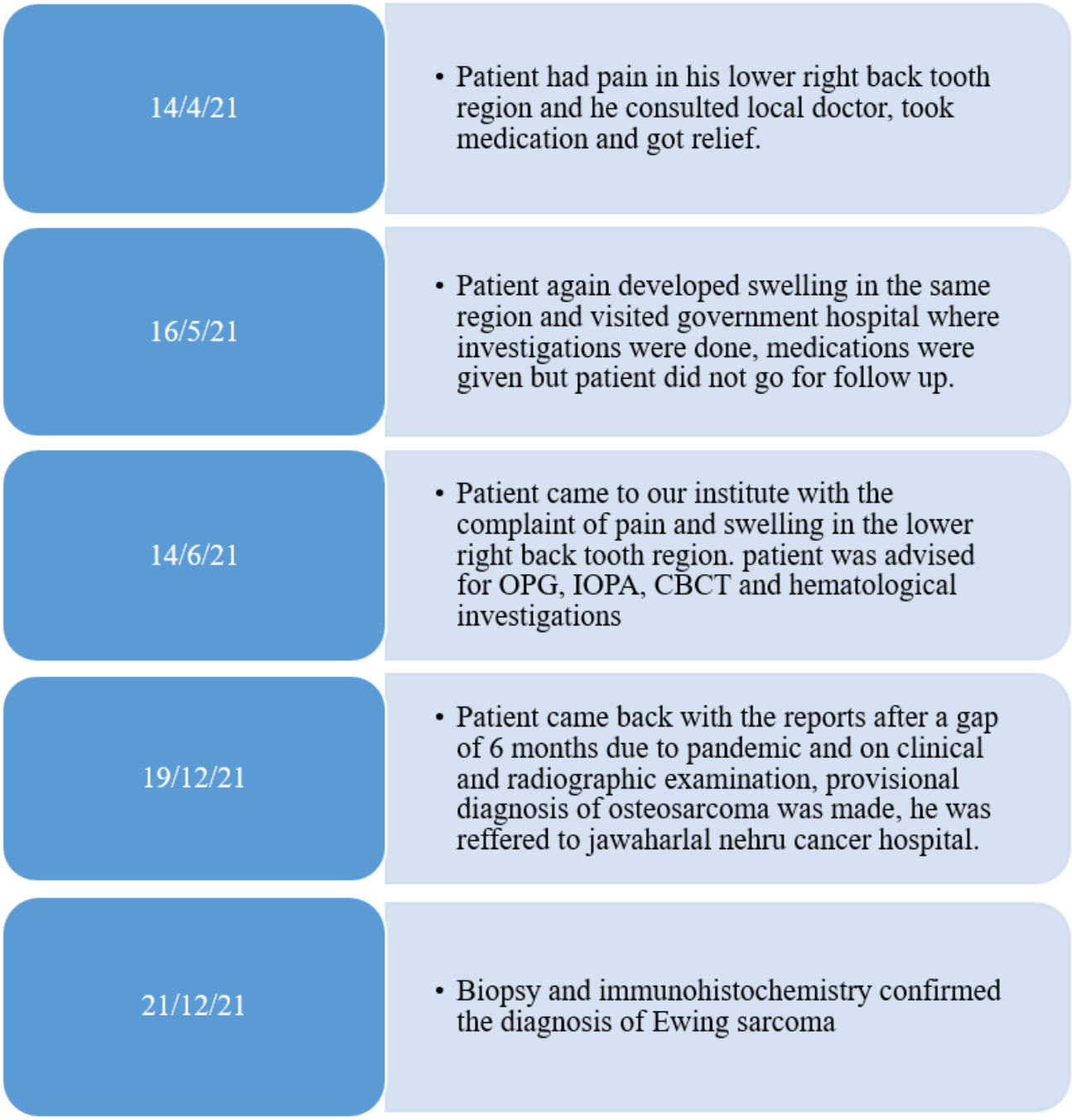

Chart 1: History of timeline

osseous lesion, dentigerous cyst, Osteomyelitis and Mural ameloblastoma were considered.

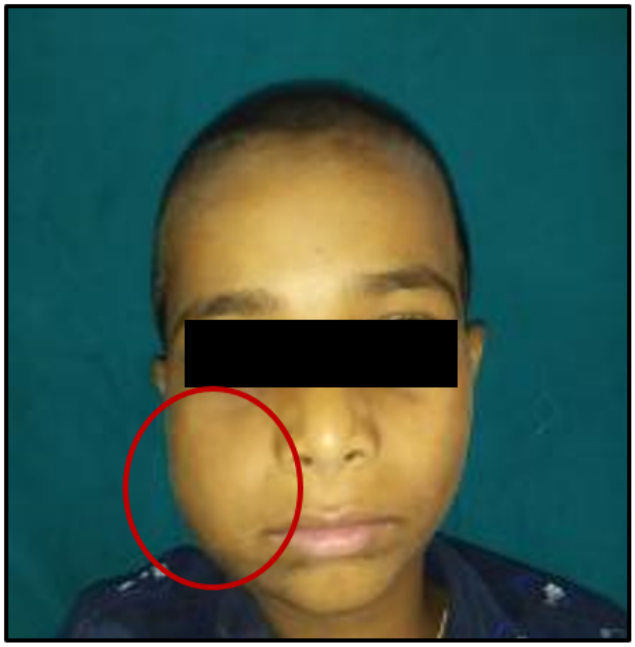

Fig. 1: Extraoral profile of the patient showing swelling on right middle and lower third of the face

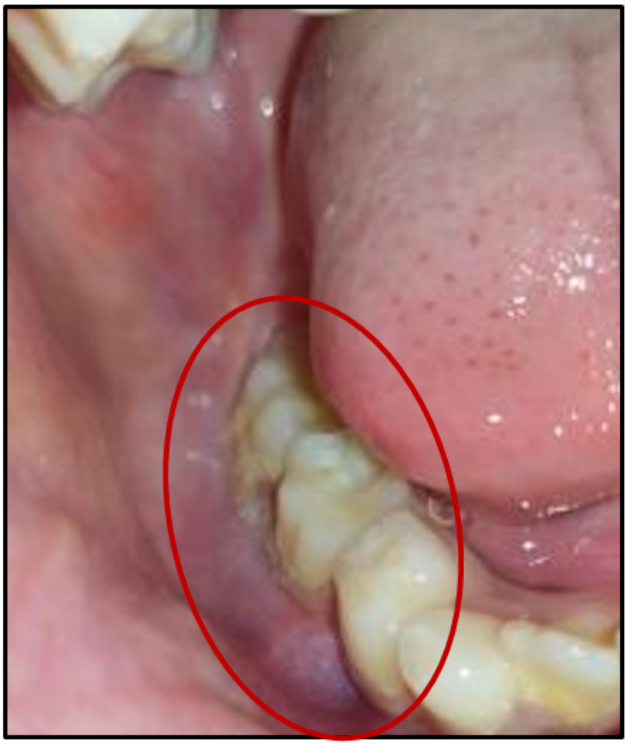

Fig. 2: Intraoral picture showing solitary swelling is present in right lower vestibular region, extending from 84 to right retromolar region of size 6 x 3 cm

2.2. Diagnostic assessment

Patient was subjected to radiographic examination, OPG revealed ill-defined lytic lesions involving tooth 84, 85, 46 on right-side body of mandible, involving angle, coronoid & condylar process. Multiple irregular radiolucent areas with thinning of inferior border of right mandible and destruction of cortical plate was evident (Figure 3). Mandibular cross-sectional occlusal radiograph revealed a mixed radiopaque-radiolucent lesion with fine bony spicules, radiating from lower border of mandible giving sunburst appearance (Figure 4). CBCT revealed irregular area of bone destruction in right mandible extending from 85 to condylar process, involving sigmoid notch, coronoid process and lesion measuring approximately 80X33 mm, Cortical plate showing irregular resorption of bone with perpendicular oriented bony spicules projecting outward giving a sun burst appearance. Tooth follicle of 45 showing loss of follicle lining with pericoronal radiolucency surrounding the tooth devoid of cancellous bone. Loss of cortication seen around mandibular canal on right side of mandible, anterior and posterior border of ramus, coronoid and condylar head showing irregular area of resorption. Thinning of the inferior border of right mandible was evident These features were giving impression of malignancy of right mandibular alveolus (Figure 5). CT face revealed mixed lytic-sclerotic area with sunray spiculation and periosteal reaction, on the right body, angle and ascending ramus of mandible giving impression of osteogenic sarcoma (Figure 6). Adjacent masticator space shows soft tissue thickening and swelling, left half of the mandible and rest of the bone were normal in outline. Haematological examination revealed, haemoglobin-10.7g/dl, RBC-3.6 mil/cmm, Packed cell volume-32.1% rest of the parameters were within normal limits. Biochemical investigation were within normal range, serum calcium-10.6 mg%, serum phosphorus-4.6mg/dl, serum alkaline phosphate-197 U/L. Histopathological examination revealed tiny bits of fibrous tissue with sheets of round to spindle cells showing hyperchromatic nuclei and ill-defined eosinophilic cytoplasm, cell nest were separated by fibrous stroma. Overlying squamous epithelium was hyperplastic which was suggestive of malignancy of round cell tumour, Ewing's sarcoma (Figure 7). Immunohistochemistry finding revealed diffusely positive vimentin, negative S-100, Diffusely positive MIC-2, diffusely positive FLI-1, and negative Desmin. These findings were in favour of malignant round cell tumour of Ewing's family. Histopathological and immunohistochemical findings confirmed the diagnosis of Ewing's Sarcoma.

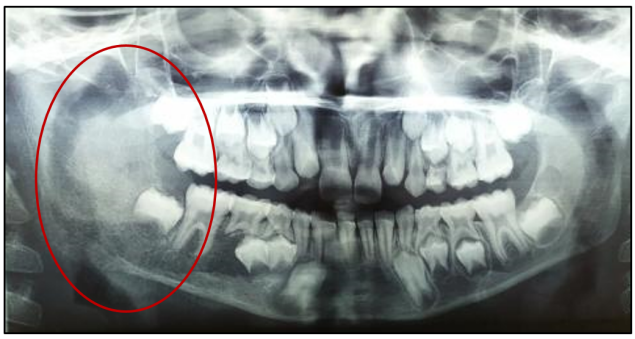

Fig. 3: OPG showing ill-defined lytic lesions involving tooth no 84, 85, 46 and region of right-side body of mandible, angle of mandible, coronoid, condylar process. Multiple irregular radiolucent areas seen, thinning of inferior border of mandible seen in right side of mandible, destruction of the cortical plate, fine bony spicules giving a sunray appearance radiating from the lower border of the mandible

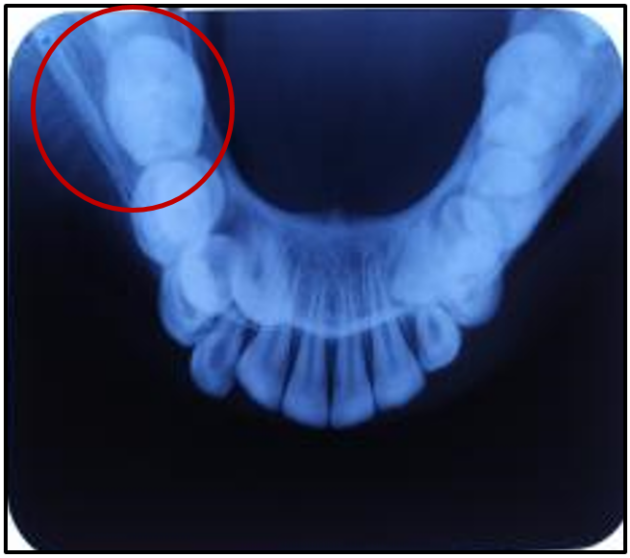

Fig. 4: Mandibular cross-sectional occlusal radiograph showing a mixed radiopaque radiolucent lesion with Fine bony spicules giving sunburst appearance, radiating from lower border of mandible

3. Discussion

Ewing's sarcoma family tumors (ESFT) include Ewing's sarcoma, peripheral primitive neuroectodermal tumors (PNET), and Askin tumors. These tumors are undifferentiated small blue round cell tumors that mainly appear in bone and less frequently in soft tissues involving the right side of the mandible.[4] Clinically, majority of the jaw tumors present as enlarging swelling causing facial asymmetry commonly involving the mandible. Very few cases which are reported in the maxilla. Most common complaint of the patient at the time of presentation are

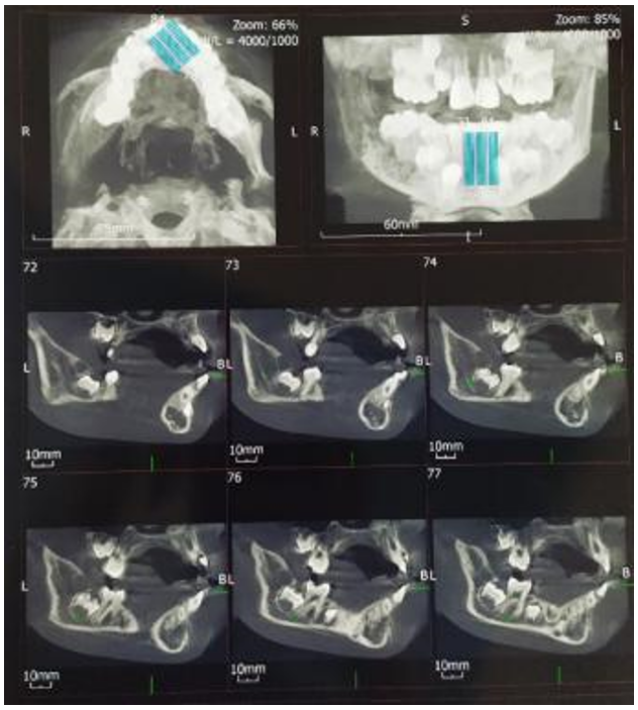

Fig. 5: CBCT giving impression of Malignancy of right mandible alveolus 45, ramus, condylar region

swelling, rapid growth and pain. Adjacent teeth may become loose as a result of permeative growth of the tumor causing destruction of alveolar bone.[5] In the present case systemic symptoms such as fever and weight loss were absent. Patient was complaining of pain and swelling was tender on palpation. Where as earlier case report had no such symptoms.[1] These presentations are not specific but confirmatory examination is needed for adequate diagnosis. In contrast, cases reported in the literature in head and neck region displayed these symptoms.

Radiologically, jaw tumors present as expansile osteolytic lesions with poorly defined borders and often with cortical destruction and permeation of surrounding soft tissues. Some cases have also been reported with findings of periosteal reactions such as onion peel and sun spicule or sunray pattern, which was evident in our case.[6] Immunohistochemistry with markers such as Myc 2 (CD99), cytokeratin, desmin (specific for rhabdomysarcoma), chromogranin (specific for neuroblastoma), and vimentin helps in differentiating the type of round cell malignancy, and is thus the confirmatory investigation to arrive at the diagnosis.[7] In the present case immunohistochemistry revealed diffusely positive vimentin, negative S-100, Diffusely positive MIC-2, diffusely positive FLI-1, and negative Desmin. Similar positive results were seen in cases reported by earlier.[8–10]

Although Ewing's sarcoma is a highly radiosensitive malignancy, it rapidly metastasizes to the bone and lungs.

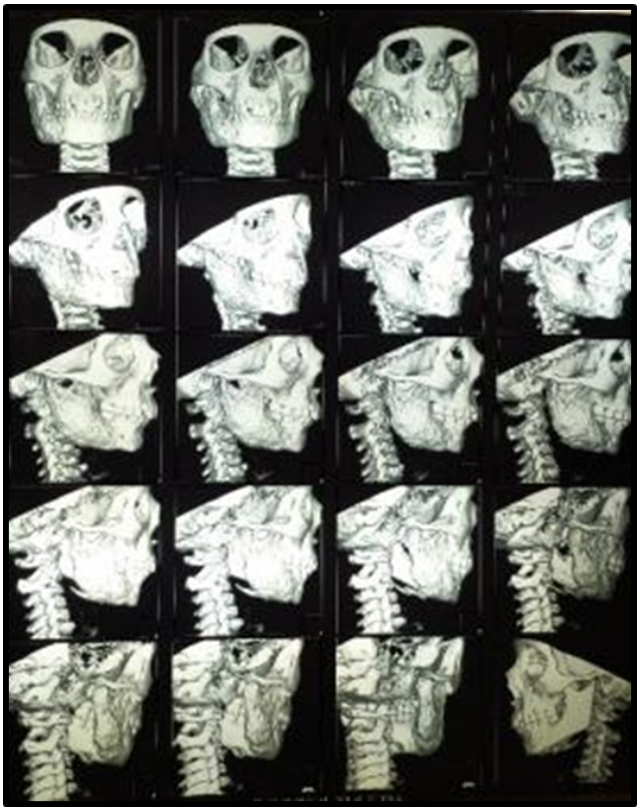

Fig. 6: CT face showing mixed lytic – sclerotic area with sun ray spiculation and periosteal reaction, Adjacent masticator space shows soft tissue thickening and swelling, left half of mandible is normal in outline. Giving impression of osteogenic sarcoma

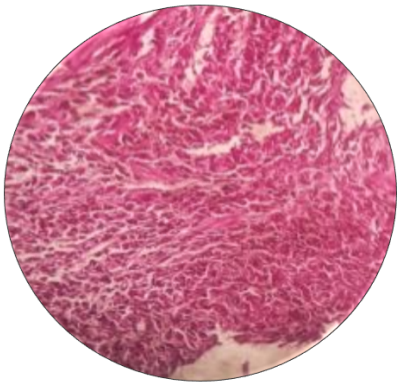

Fig. 7: Histopathological findings reveal tiny bits of fibrous tissue with sheets of round to spindle cells showing hyperchromatic nuclei and ill-defined esosinophilic cytoplasm. Cell nest were separated by fibrous stroma. Overlying squamous epithelium is hyperplastic suggestive of Ewimg's sarcoma

Combined surgery, radiotherapy and chemotherapy is the best approach for Ewing's sarcoma with an overall survival of 5 years in 4–7% of patients.[5] Due to its high lethality much of the literature suggests aggressive multidisciplinary approach which include surgery chemotherapy and radiotherapy. Most current treatment call for multidrug chemotherapy followed by surgery and radiotherapy. 30-40% cases have chances of recurrence.[4]

4. Conclusion

This paper highlights the uncommon entity in mandible which will be helpful to make dentists and pedodontists aware of a possible outcome of a rapidly growing swelling. Ewing's sarcoma of the bone is an uncommon malignancy that rarely occurs in the jaw and as it is mistaken for odontogenic infections, it should be carefully examined clinically, and because of similarity in microscopic presentation with other small round cell tumors, the diagnosis is often difficult. This tumor has the better prognosis if lesion is detected early with initiation of treatment. Evaluation of the tumor using conventional radiographs, CBCT, CT followed by histopathology and immunohistochemistry is necessary for early diagnosis and better management, as it has tendency to metastasize and for its poor prognosis.

5. Source of Funding

None.

6. Conflict of Interest

None.

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