International Journal of Pediatric Otorhinolaryngology Extra 13 (2016) 33–39

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology Extra

journal homepage: http://www.ijporlextra.com/

Case report

Ewing's sarcoma of the jaws: An institutional study of four cases

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ARTICLE INFO

Article history:
Received 7 January 2016
Received in revised form
25 March 2016
Accepted 27 March 2016
Available online 27 April 2016

Keywords:
Ewing's sarcoma
Jaws
Multimodal therapy

ABSTRACT

Ewing's sarcoma (ES) is a rare malignant small round cell tumour that primarily affects the skeletal system. It accounts for less than 4–10% of all types of bone malignancies, with long bones and pelvis being involved most commonly. Clinically, ES can mimic odontogenic inflammation/abscess. Integration of clinical, radiographic, histologic and immunohistochemical information is essential for prompt diagnosis. Aggressive multimodal therapy and continuous follow up results in better prognosis of patient diagnosed with Ewing's sarcoma. Through this paper we reiterate the importance of promptly diagnosing and treating this rare tumour entity that can help in improving the prognosis.

1. Introduction

Ewing's sarcoma (ES) is a rare malignant small round cell tumour that primarily affects the skeletal system. It accounts for less than 4–10% of all types of bone malignancies, with long bones and pelvis being the most commonly involved [1–3]. It affects mainly the adolescents and young adults and is rarely seen before the age of 4 and after the age of 30 years. Males are twice more commonly affected than females. The occurrence of this tumour in head and neck region is unusual and when it occurs in the jaw, mandible is more frequently affected than maxilla [4]. Only 10% of mandibular ES are metastatic lesions, whereas the other 90% are primary tumours [5]. Prompt diagnosis is essential for good prognosis as the lesion shows aggressive behaviour characterised by rapid growth and non-specific clinical findings [1,2,6].

Histopathologically, ES is composed of small, poorly differentiated cells with medium sized round or oval nuclei. Combined therapy including surgery, radiotherapy and chemotherapy is the best approach for treatment of the tumour.

The aim of the present study was to perform an in depth clinical, radiological and histological analysis of cases of Ewing's sarcoma that presented to a tertiary care dental hospital over a seven year period.

2. Materials and method

Archival data of the patients diagnosed histopathologically as Ewing's sarcoma was retrieved from the year 2007 to 2015 from the records of Department of Oral Pathology, Maulana Azad Institute of Dental Sciences, New Delhi, India. The clinical, radiographic and histopathological features of all the cases were analysed. Special stains and immunohistochemistry were performed to facilitate correct diagnosis.

3. Results

Case #1

A 17-year-old male reported to the outpatient department with the chief complaint of a slowly growing swelling on left cheek since 4 months. He gave a history of trauma to the upper jaw prior to appearance of the swelling. He also complained of nasal obstruction and slight pain in relation to the swelling. On extraoral examination, there was a firm, tender, well defined, swelling approximately 6 × 6 cm in size, in the left canine space causing obliteration of the left nasolabial fold. The overlying skin was normal and adherent to the underlying swelling. Intraorally, buccal and palatal cortical plates were expanded from maxillary left central incisor to left first premolar. The maxillary left lateral incisor and canine were displaced but non tender and immobile. The overlying mucosa was non ulcerated with bluish tinge on the buccal aspect.

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http://dx.doi.org/10.1016/j.pedex.2016.03.005
1871-4048/

The radiographic findings (OPG and CT scan) have been described in Table 1.

An incisional biopsy was performed under local anaesthesia via the intraoral approach which was followed by uneventful healing. **The tissue specimen was sent for histopathological analysis. The microscopic findings along with immunohistochemistry have been elaborated in Table 2.** Based on the microscopic picture and immunohistochemistry findings a final diagnosis of Ewing's sarcoma of rhinomaxillary complex was made and the patient was referred to the Head and Neck Oncology Centre where he received radiotherapy followed by chemotherapy. The patient is on regular follow-up since 8 years and no recurrence is observed to date.

Case #2

An 18-year-old male reported with an 8 month history of a painless, gradually increasing swelling in the left posterior mandible. Extraorally, bony hard swelling measuring 4 × 3 cm was present along the mandibular body on left side extending up to the angle region covered with normal overlying skin. Intraorally, swelling was bony hard, non-tender and causing expansion of the buccal cortical plate and extending from second premolar to second molar. The teeth were immobile and asymptomatic. The patient was apparently healthy with no signs of paresthesia/lymphadenopathy. **Fine needle aspiration cytology (FNAC) and computed tomography (CT) scan (Fig. 1) have been elaborated in Table 1.**

The patient underwent incisional biopsy under local anaesthesia. **The microscopic picture and immunohistochemistry results have been discussed in Table 2.** Hence, a final diagnosis of Ewing's sarcoma of the mandible was made. The patient was referred to higher centre for further treatment where surgery and chemotherapy were done. Patient is undergoing chemotherapy intermittently since 2 years.

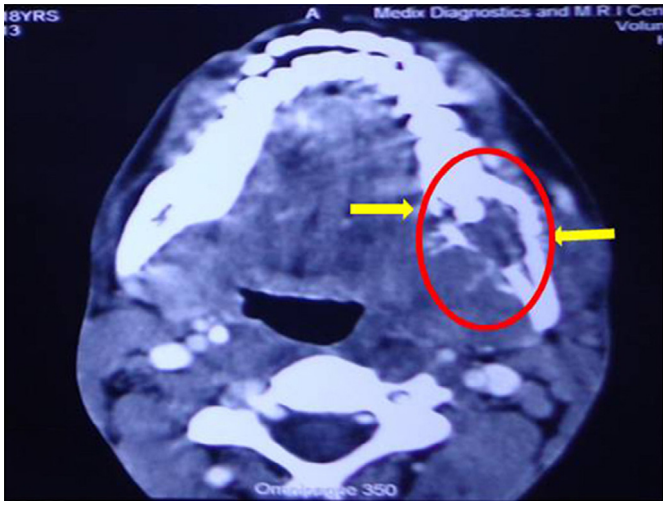

Fig. 1. CT scan revealing ill defined osteolytic lesion within body and ramus of left mandible causing erosive destruction of overlying cortices and showing periosteal new bone formation giving a sun ray appearance (case #2).

Case #3

A 4-year-old boy reported to the Department of Oral Pathology with a 4–5 months history of a painless, progressively enlarging swelling in the right mandible which mimicked a dental abscess.

Table 1
Radiographic presentation and treatment.

S. no	Orthopantomogram (OPG)	Computed tomography (CT) scan	Fine needle aspiration cytology (FNAC)	Treatment
1	Ill defined osteolytic lesion in relation to maxillary left central incisor to premolar, with non sclerotic borders displacing roots of maxillary lateral incisor and canine.	Diffuse ill defined homogenous density lesion involving maxillary sinus and reducing in size, crossing the midline with thinning of cortical plate of hard palate	NA	Radiotherapy + chemotherapy
2	NA	Irregular soft tissue density lesion within body and ramus of left mandible causing erosion of overlying cortices and showing periosteal new bone formation: **sun ray** appearance	Suggestive of small malignant round cell tumour	Surgery + chemotherapy
3	Ill defined osteolytic lesion in the right mandibular body	NA	NA	Surgery + chemotherapy
4	Well-defined multilocular radiolucency extending from 33 to 36 up to inferior border of mandible displacing roots of teeth (34, 35, 36). Thinning of the inferior border of mandible.	Well-defined multilocular expansile lytic lesion from 33 to 36 with erosion of cortices, perforation of the buccal cortex and displacement of roots of premolars and molars	Suggestive of small malignant round cell tumour	Surgery + chemotherapy

NA, not available.

Table 2
Histopathological presentation and immunohistochemistry.

Case	Histopathological presentation	Immunohistochemistry
1	Sheets of small round cells with vesicular nuclei, hyperchromatism and mitotic figures.	CD99 +++ (membranous positivity) Negative: S-100, desmin and HMB-45
2	Sheets of round cells having large, vesicular nuclei and indiscernible cytoplasmic boundary. Few cells show hyperchromatic nuclei and mitotic figures. PAS positive diastase sensitive glycogen in the cytoplasm of the tumour cells.	CD99 ++ (membranous positivity) Negative: S-100
3	Numerous clusters, cords strands of small, round cells with vesicular nuclei. Connective tissue stroma is fibrovascular. PAS positive diastase sensitive intracytoplasmic glycogen in round cells.	CD99 ++ Negative:S-100, CD 45, desmin, neuron specific enolase (NSE), synaptophysin
4	Nests, cords and sheets of dysplastic round to ovoid cells showing hyperchromatism, pleomorphism, few mitosis and vesicular nuclei. Few areas of central necrosis. Connective tissue was fibro collagenous with moderate cellularity and increased vascularity. PAS positive diastase sensitive glycogen in the cytoplasm of the cells.	CD99 +++ Focal S-100 positivity Negative: CD 45, desmin, neuron specific enolase (NSE), synaptophysin, chromagranin, HMB 45, LCA

+, intensity of positive staining.