Ewing’s Sarcoma Of The Mandible In 11 Year Old Female

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Citation

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Abstract

Ewing's tumor or Ewing's sarcoma is a malignant tumor of bones that primarily affects children and young adults. The true origin of this small round cell lesion still remains controversial .It was originally described by James Ewing in 1921 as arising from undifferentiated osseous mesenchymal cells, however, recent studies suggests that Ewing’s tumor might be neuroectodermally derived from various degrees of differentiation of the primitive neural tissues A case of Ewing’s sarcoma of mandible in a 11 year old female is presented here.

INTRODUCTION

The small-round-cell tumors of childhood include neuroblastoma, the Ewing family of tumors, rhabdomyosarcoma, lymphoma, and desmoplastic small-round-cell tumor[1]. This group of small round cell tumor is malignant, highly aggressive and seen under the microscope as a monotonous proliferation of small cells with scanty cytoplasm.

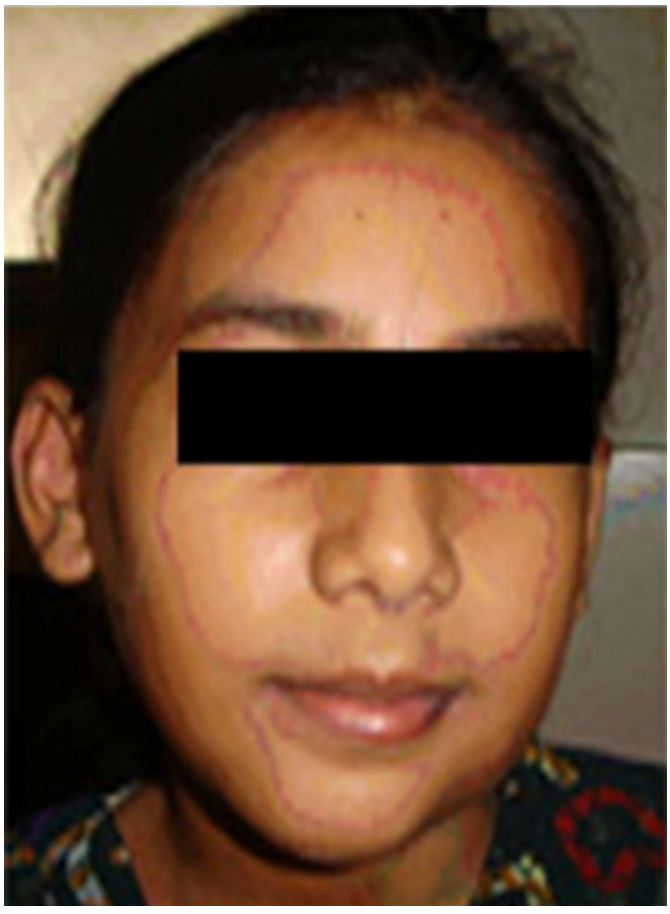
Among the tumor such as small cell osteosarcoma are more common in young patients where as tumor like metastatic small cell carcinoma are more common in adults[2].Athough classical histological features are generally highly suggestive of tumor type, on occasions these tumors may be indistinguishable by light microscopy, making a definitive diagnosis difficult[1].

CASE REPORTS

An 11-year-old girl visited Yenepoya dental college and hospital Deralakatte, Mangalore, out patient department with the complaint of swelling on the left side of the face (Fig.1).

Figure 1

Figure 1: Extra oral swelling

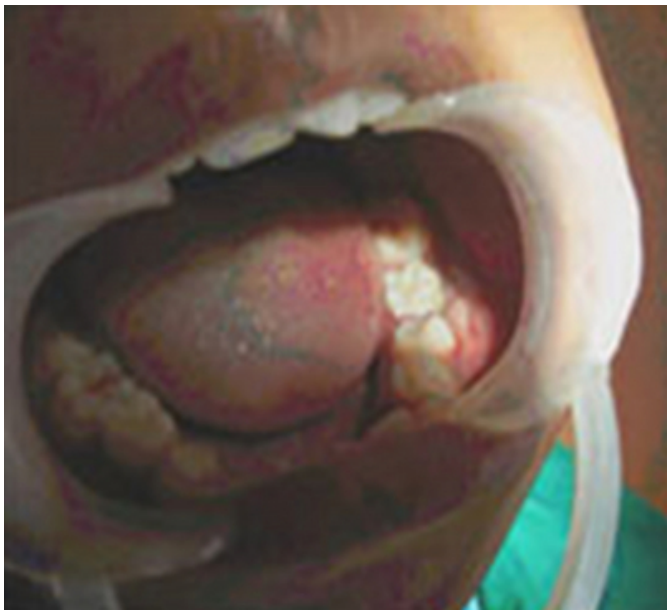


The swelling started 6 months back which gradually enlarged to present size. On examination, extra orally a diffuse swelling was seen on left side of face which extended superiorly up to ala tragus line, inferiorly up to right angle of the mandible. Intra oral examination revealed mixed

dentition and swelling extending from left mandibular canine to left mandibular second molar (Fig.2).

Figure 2

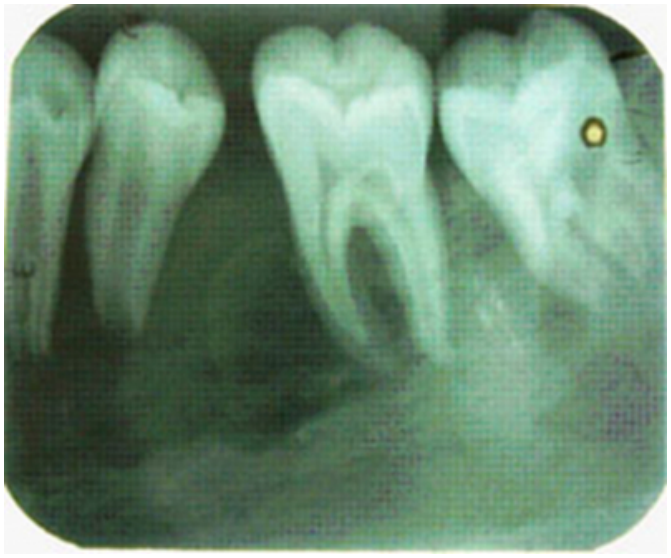
Figure 2: Intra oral swelling



On palpation the swelling was non-tender and hard in consistency. Bi-digital palpation revealed expansion of the buccal plate, no perforation of buccal/lingual cortex and no paresthesia of mucosa or lower lip were seen. Grade III mobility was seen on all the teeth from left mandibular canine to left mandibular second molar, which were non tender on percussion.

Figure 3

Figure 3: The intra oral periapical radiograph shows radioleucency near the mandibular teeth

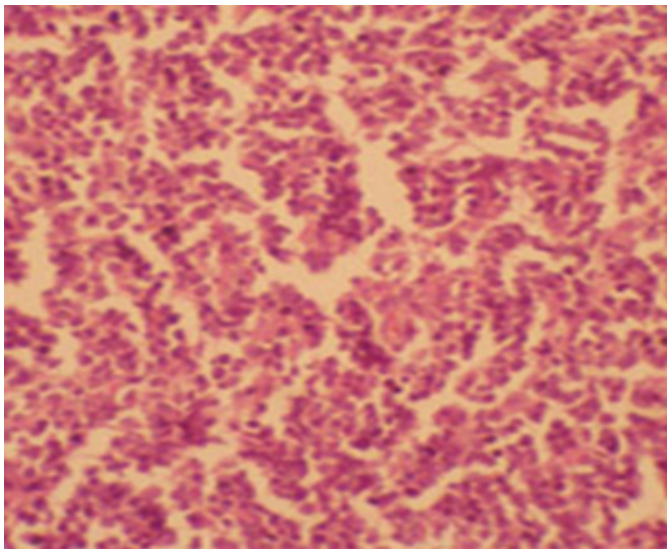


The results of hematological and biochemical investiga¬tions were within normal limits. Under Local Anesthesia, extraction of the mobile teeth left mandibular first molar was performed and the tissue sample obtained through the extraction socket was sent for histopathological examination.

On microscopic examination, H&E stained sections showed sheets of uniform, small, round cells arranged in diffuse pattern, with indistinct outline, scanty cytoplasm, and well-defined nuclear outline, with round to oval nucleus and inconspicuous nucleoli. Fig (4). Mitotic figures were rare. A diagnosis of malignant small round cell tumor was made based on the above histopathological findings.

Figure 4

Figure 4: Microscopic picture under 40 x demonstrating the sheets of monotonous, round cells containing single, round to oval nucleus.



A panel of immunohistochemical markers namely CD99, CD3, CD20, CHR, MPO, desmin, and SYN were used to rule out other small round cell tumors. Tissue sections showed positive expression for CD99 (MIC2), with characteristic membranous pattern.

DISCUSSION

James Ewing first described Ewing sarcoma (E.S) in 1921, after observing radiosensitivity in a subgroup of bone tumors. In the early 1980s, Ewing sarcoma and the peripheral primitive neuroectodermal tumor were both found to contain the same reciprocal translocation between chromosomes 11 and 22, t (11; 22). Later that decade, similar patterns of biochemical and oncogene expression were observed. These tumors were categorized as the Ewing sarcoma family of tumors (EFT) because the shared

translocation and the similar cellular physiology[3].

Ewing's sarcoma and malignant peripheral neuroectodermal tumors are the second most common primary malignant tumors of bone in childhood and adolescence[4].

Ewing's family of tumors includes Ewing's tumor of bone (ETB or Ewing's sarcoma of bone), extraosseus Ewing's (EOE), primitive neuroectodermal tumors (PNET or peripheral neuroepithelioma), and Askin's tumor (PNET of the chest wall). Studies using immunohistochemical markers, cytogenetics, molecular genetics, and tissue culture indicate that these tumors are all derived from the same primordial stem cell [5].

The median age for a patient with Ewing's family of tumors is 15 years and more than 50% of patients are adolescents [6].Major prognostic factors include site, tumor volume, and the presence of metastases [5].

In Ewing's tumor of bone the most favorable sites are distal extremities and central location (e.g., skull, clavicle, vertebrae, and ribs). Proximal extremities, and especially the pelvis, are associated with a much less favorable prognosis. Size is also significant, but the larger lesions tend to occur in the more unfavorable sites. Younger children have better event-free survival than older adolescents and young adults. Girls with Ewing's tumor of bone have a better prognosis than boys. The presence of fever, anemia, and an elevated Lactate dehydrogenase (LDH) is a poor prognostic sign for patients with Ewing's tumor of bone increased serum Lactate dehydrogenase levels prior to treatment correlate with metastatic disease and shorter disease-free survival[5].

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