Ewing’s Sarcoma of the Jaws with Sunray Appearance in Sonography

Vivekandh Reddy Gangili, Ravindar Puppala, Jithendar Kubbi, Ramkal Gantala, Navadeepak Korvipati

ABSTRACT
Ewing’s sarcoma (ES) is an uncommon malignancy that occurs usually in childhood. In most of the reported cases, the occurrence of this tumor is more in the mandible than the maxilla. A case of Ewing’s sarcoma of the mandible along with the maxilla is discussed. A complete clinical, radiographic, biochemical and histological evaluation was done, ultrasonography revealed a sunray pattern; microscopically, the tumor was composed of monotonous small round cells that exhibited immune reactivity for CD99 and vimentin. The patient was subjected to multi-agent chemotherapy. This case elucidates the importance of professional knowledge of the relevant aspects of malignant lesions, such as ES. Although the prognosis remains poor, an increased rate of survival is now associated with early diagnosis and combined therapy.

Keywords: Ewing’s sarcoma, Childhood tumors, Sunray appearance, CD99, Vimentin, Immunohistochemistry.

INTRODUCTION
Ewing’s Sarcoma (ES) is a primary malignant tumor affecting the skeletal system, is now thought to arise from immature reticulum cells or primitive mesenchymal cells of the bone marrow.1 James Ewing was the first to describe ES, which was named after him in 1921.

The occurrence of ES in the head and neck region is unusual and, when it occurs, it generally involves the mandible and less frequently the maxilla.2,3 It accounts for 4 to 10% of all types of bone cancer, with long bones and pelvis being the most common locations.4,5 ES rarely affects the jaws, and the incidence of jaw involvement has been reported as varying from 1 to 10%.6

In this report, we describe a case of ES showing the characteristic sunray appearance in an ultrasonography, but not in a conventional radiograph.

CASE REPORT
A 12-year-old male patient reported to our department with painless diffuse swelling near the left angle of mandible. No history of trauma, pain, paresthesia or trismus was reported. The swelling was insidious in onset. Over a period of 2 months, the entire left side of the face was involved. Medical history was noncontributory. Extraorally a gross facial asymmetry with a well-circumscribed swelling was present on the left side of face and the surface of the overlying skin is smooth and shiny (Fig. 1).

On extraoral palpation, the swelling was tender, bony hard in consistency, nonpulsatile and noncompressible. Bilateral submandibular lymph nodes were palpable and the lymph nodes were single, firm and fixed.

On intraoral inspection, there was a swelling on the left buccal mucosa, which extended anterioposteriorly from the left mandibular canine region to the retromolar area, mediially, involved the lingual vestibule, laterally the buccal vestibule and palatal, the surface of the left maxillary third molar region. There was generalized mobility of teeth (Fig. 2) upon palpation, all inspectory findings regarding the size and shape extent are confirmed, the swelling was tender and surface was lobulated prior to the investigations, a sarcoma of the mandible was suspected, and the following investigations were carried out, i.e. the complete blood picture (CBP), bone marrow aspiration (BMA), conventional radiographs, CT scan, PET scan, ultrasonography (USG) and lastly the incisional biopsy was done.

CBP and BMA were within the normal limits. Conventional radiographic findings showed the mixed radiolucent and radiopaque lesions involving the left side of the mandible and the left molar region of the maxilla, the complete resorption of surrounding structures leading to the floating teeth appearance on the left side of mandible.

Fig. 1: An extraoral swelling of size 3 × 5 cm is seen at the left side of face
with the displacement of lower left third molar distally (Figs 3A and B).

Helical CT was taken prior to the biopsy and revealed a large soft-tissue density mass lesion (6.2 × 6.2 × 10.8 cm) on left side of mandible involving the temporal, infratemporal fossa, pterygomaxillary fissure, submandibular region and the lesion was found to be herniating into the parapharyngeal space, three-dimensional reconstruction of the images revealed an irregular lytic destruction of the ramus. The body of left mandible was seen with minimal interrupted and speculated periosteal reaction and minimal erosion of coronoid and condylar process is seen.

PET CT (Tc99m-MDP tracer) revealed a hot spot in mandible and maxilla consistent with the primary tumor (Figs 4A to C).

USG shows large heterogeneously hypoechoic mass noted in the left mandible region with erosion of mandible. The mass demonstrates increased color flow in Doppler, suggestive of vascularity. Spicules noted over surface of mandible were suggestive of sunray pattern (Figs 5A and B).

Multiple enlarged lymph nodes involved all upper groups in the cervical region on left side and few nodes on right side of submandibular region.

The histological report was suggestive of small round cell tumor at the left mandibular region (Figs 6A and B) and the immunohistochemistry section shows diffuse sheets of small round cells with the cells arranged in rosettes formation. Moreover, atypical mitotic figures were seen. LCA and desmin were negative, and CD99, vimentin were positive.

Based on the history, clinical findings and investigations, a final diagnosis of ES was made, and the patient was treated with chemotherapy. Vincristine 1 ml injection (Oncovin®, Sun Spectra, India), cyclophosphamide injection 1 gm (Neophos®, Cipla, New Delhi, India) delivered over a period of 60 minutes, doxorubicin hydrochloride 5 ml injection (Adriamycin®, Pharmacia, India), ranitidine 2 ml injection (Rantac®, JB Chemicals, India) and metoclopramide tablet 10 mg (Perinorm®, IPCA, India) were administered in a 15-day-cycle for a period of 6 months, and during this period the regression of the lesion was noted. The patient is under follow-up and is being evaluated on a monthly basis till date (Figs 7A and B).

DISCUSSION

ES is a rare primary malignant tumor, affecting the skeletal system originally described by James Ewing in 1921. This tumor is found to affect children and young adults. Swelling, pain, paresthesia and loose teeth are frequent symptoms. The mandible is commonly affected then the maxilla. Radiographically, it is most often seen as destructive, expansile, mottled radiolucent lesions, which may produce a laminated periosteal reaction. Some authors point out that radiographic appearance of ‘onion skinning’ is a
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Fig. 4A and B: Helical CTs with reconstruction of the images were done and revealing an irregular lytic destruction of the ramus, and erosion of the coronoid and condylar process.

Fig. 4C: PET CT done with Tc99m-MDP tracer and it revealed a hot spot in mandible and maxilla.

Fig. 5A: USG reveals spicules noted over surface of mandible suggestive of sunray pattern.

Fig. 5B: A hypoechoic mass noted in the left mandible region with erosion of mandible.

characteristic sign of Ewing’s lesion of the bone.\(^7,8\) While it may be true in most of the cases, some dissenting authors have stated that this pattern is either uncommon or exceedingly rare in jaw lesions.\(^9,10\) In the jaw, osteolytic radiolucent area with cortical destruction was seen in most reported cases.

With respect to teeth, some radiological features were noted, which include periodontal space widening, loss of lamina dura, root resorption, displacement of teeth.\(^1\) Additionally, a sunray appearance is common in radiographic finding. A sunray appearance on USG has been described only once in the literature, i.e. in osteosarcoma of the mandible.\(^11\) This is significant because in the present case, the sunray appearance was not seen on the conventional radiograph, but was present on sonography. This can be considered to be an uncommon finding. This event could occur when the periosteum is rapidly stripped from the cortex. It is thought that a variety of substances that contact the inner surface of the exposed periosteum can cause new
bone formation. These substances include inflammatory products from infection, blood from trauma, granulomatous reactions and neoplasms.\textsuperscript{12}

Histopathologically, this tumor must be differentiated from other round cell tumors like mesenchymal chondrosarcoma, rhabdomyosarcoma, malignant lymphoma, eosinophilic granuloma, neuroectodermal tumors and metastatic neuroblastoma.

Eosinophilic granuloma, malignant lymphoma and metastatic neuroblastoma are the lesions most easily mistaken for ES and can be distinguished because of presence of ‘histolytic’ features, namely the abundance of eosinophilic cytoplasm with oval or indented nucleus. Malignant lymphomas contain lymphoid cells, intermixed with round cells components of varying size and cytoplasm contents. The PAS stain is usually negative, and reticulin stain is often positive.

The cells of metastatic neuroblastoma have a yellow fluorescence when exposed to formalin vapor and are neuron specific and enolase positive.\textsuperscript{13} The other small cell tumors, including ES, should be ruled out using immunohistochemistry. CD99 positivity has been noted in small cell osteosarcoma.\textsuperscript{14} Positive reaction of either of these: LCA, S-100, EMA, SMA, factor VIII, smooth muscle actin, neuron-specific enolase, synaptophysin, would favor the exclusion of small cell osteosarcoma.\textsuperscript{15} Most small cell osteosarcomas show vimentin positivity and occasionally minority may be muscle-specific actin (HHF-35) positive. Regarding the Mic-2 gene have confirmed the high sensitivity of the Mic-2 gene product (CD99) for all ES family tumors over 95\% of the cases showing positivity to this marker.\textsuperscript{16} Our case showed CD99 and vimentin positivity and LCA, desmin negativity.

The prognosis of small cell osteosarcoma was considered to be worse than conventional osteosarcoma and ES.\textsuperscript{17} It is known that periosteal osteosarcoma is capable of local recurrence and distant metastasis while parosteal type is considered to be a low-grade neoplasm.\textsuperscript{18}
SUMMARY

We have described a case of ES showing the characteristic sunray appearance in an ultrasonography but not seen in conventional radiographic images. We believe that this report will alert clinicians to the fact that the absence of characteristic radiographic features may not necessarily exclude a disease and might necessitate further investigations to arrive to a definitive diagnosis.

REFERENCES