EWINGS SARCOMA OF MAXILLA: A CASE REPORT

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ABSTRACT

Ewing's sarcoma is an uncommon malignancy that occurs usually in childhood. In most reported cases, the occurrence of this tumor is more in the mandible than the maxilla. A case of Ewing's sarcoma of the maxilla together with its associated structures is presented and discussed with complete clinical, radiographic, and histological evaluation. Although the prognosis remains poor, an increased rate of survival is now associated with early diagnosis and combined therapy.

Key words: *Ewings Sarcoma, Maxilla*

INTRODUCTION

James Ewing was the first to describe Ewing's sarcoma, which was named after him in 1921. Ewing's sarcoma, 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]. This tumor accounts for approximately 4-10% of all malignant neoplasms of the bone, with pelvic girdle and the long bones of the lower extremities being the most common locations. It is an uncommon malignancy, which usually occurs in childhood[2]. Ewing's sarcoma rarely affects the jaws, and the incidence of jaw involvement has been reported as varying from 1% to 10%[3].

Since the prognosis is poor due to its uncontrolled potential for metastatic spread, early and appropriate intervention is warranted

CASE REPORT

A 16-year-old female patient reported with a complaint of swelling since 6 months and occasional pain for past 2 months on the right side of the face. Swelling was insidious in onset. Over a period of 6 months, the entire right side of the midface was involved with progressive decrease in right nasal airway competence. There was no history of trauma (Figure 1).

The patient's history revealed no systemic problems. Moreover, a general physical

Figure 1: Right sided fascial swelling

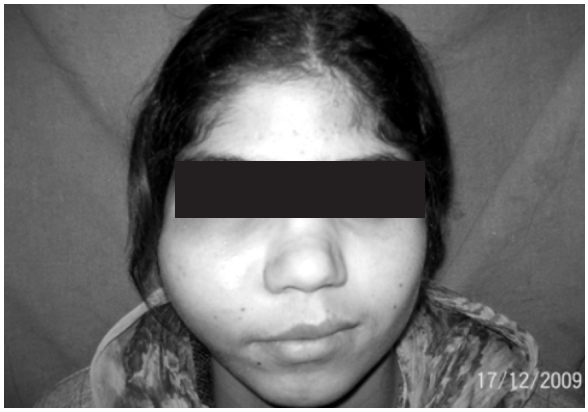
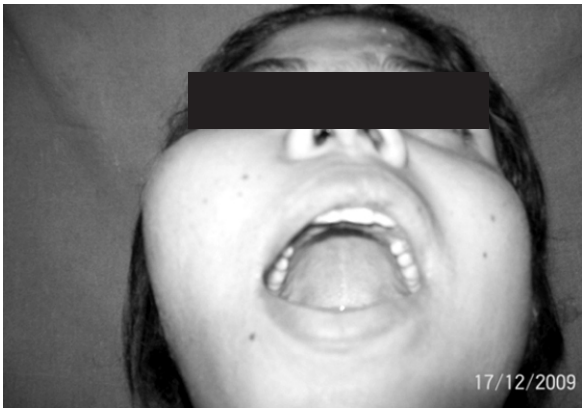


Figure 2: No bulge in palatal region



examination revealed no significant disorders. A gross facial asymmetry with a well-circumscribed swelling measuring approx.6 × 6 cm was present on the right midface. There was restricted eye opening with decreased palpebral fissure width. Visual acuity was normal with normal eye movements.

On palpation, swelling was tender, bony hard in consistency, non-pulsatile and non-compressible with no evidence of secondary changes. Intra oral examination showed no bulge in the palatal region (Figure 2).

Intraoral palpation was normal. There was grade 1 mobility of right upper premolars. Considering the patient's history and clinical observations following investigations were carried out.

1. Radiographs and a CT scan
2. Hematological and biochemical investigations
3. An incisional biopsy

CT features were suggestive of an expansile mass with erosive borders in right maxilla (suggestive of malignancy) (Figures 3 & 4).

Hematology report: Hb - 11.6 gm%, TC - 7350 cells/mm^3 , ESR - 16 mm hr^{-1} , PCV - 35%; bleeding time - 1 min, clotting time - 4 min; DC - neutrophils - 64%, lymphocytes - 33%, eosinophils - 03%, monocytes and basophils - 0%; blood group - AB, Rh type - +ve, HIV - negative (tridot), HBSAg - negative (surface line).

Biochemical investigations: Using a semiautomatic analyzer (Erbachem Pro), the following were investigated: blood sugar, urea, SGOT, SGPT, K, Na and Ca. The above parameters were within the normal range.

Histopathology: The biopsy of the lesion was taken via a sub labial incision. It was sent for histopathological examination. Sections were stained using H and E, PAS, and reticulin stains. The H and E sections revealed solid sheets and masses of small round cells with scanty stroma. Intervening between the tumor cells, few connective tissue septa were seen. The cells revealed faint cytoplasmic membrane with large hyperchromatic nuclei with scanty cytoplasm. In certain areas, the tumor cells were arranged in rosettes perivascularly. Infrequent mitotic figures were also observed. Significant necrotic areas were also seen (Figure 5).

PAS sections were positive for intracytoplasmic glycogen content. Reticulin was

Figure 3: CT Scan (view A)

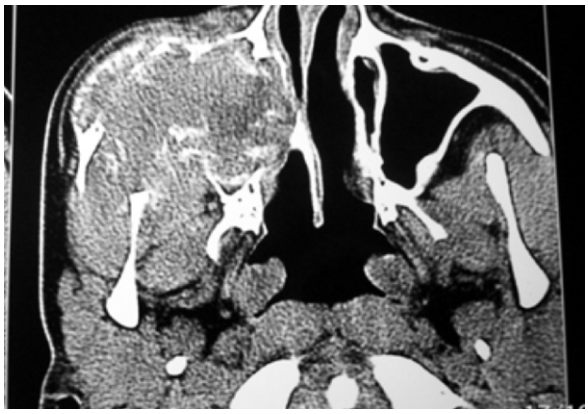


Figure 4: CT Scan (view B)

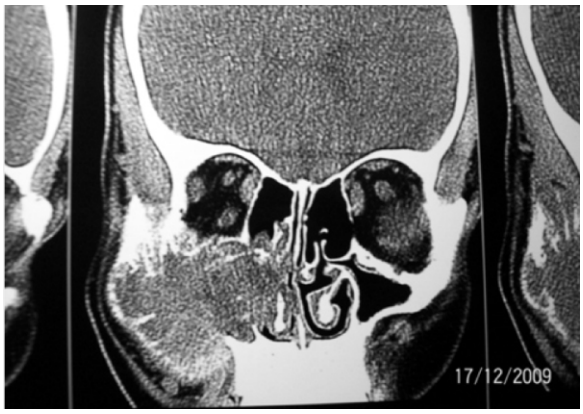
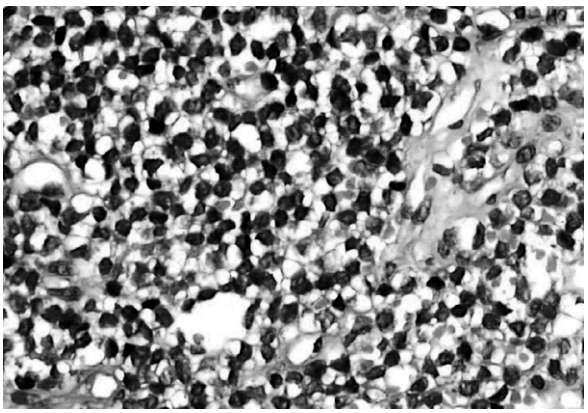


Figure 5: Histopathological Picture



positive around vessels and in collagenous septa. The typical histologic appearance with the presence of PAS-positive glycogen and the absence of any tumor osteoid was conclusive to diagnose this tumor as Ewing's sarcoma.Patient was sent for chemotherapy to the department of Medical Oncology as she denied surgery citing permanent disfigurement of face.

DISCUSSION

Ewing's sarcoma is a rare malignant tumor affecting the skeletal system. This tumor is found to affect children and young adults. Swelling, pain, parasthesia, and loose teeth are frequent symptoms[3]. Unlike the present case, mandible is found to be more commonly affected than the maxilla. Radiographically, it is most often seen as destructive expansile, mottled radiolucent lesion, which may produce a laminated periosteal reaction. [3] Some authors point out that radiographic appearance of "onion skinning" is a characteristic sign of Ewing's lesion of the bone[4,5]. 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[6,7]. In the jaw, an 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 or more commonly destruction of unerupted tooth follicles[1].

An extraskeletal form of this tumor has been described by Angervall and Enzinger, which is termed Ewing's sarcoma of soft tissues. The ultra structural characteristics of the cells of this tumor studied by Gillespie and his associates proved that these cells are identical to those of the typical Ewing's cells[8].

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

Of these, eosinophilic granuloma, malignant lymphoma, and metastatic neuroblastoma are most easily mistaken histologically for Ewing's sarcoma. Eosinophilic granuloma can be distinguished because of the presence of "histiocytic" features, that is, abundant, indistinct eosinophilic cytoplasm with oval or indented nucleus. Multinucleated cells may be present. Malignant lymphoma contains lymphoid cells, intermixed with round cell components of varying size and cytoplasmic contents. The nuclei of these cells are round or oval with a distinct nuclear membrane, which is sometimes smooth or cleaved. 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[9].

The prognosis of Ewing's sarcoma is poor because of multiple metastases most commonly to the bone, lung, lymph node, and liver, which may occur within a few months after the onset of the tumor[10]. Recently, some reports of early and prompt diagnosis with combined surgery, radiation, and chemotherapy protocols have raised the five-year survival rate of patients with Ewing's sarcoma from 16% to 74%[11].

CONCLUSION

In general, Ewing's sarcoma is a rare malignancy that may affect the facial bones of young individuals. Even with an isolated area of Ewing's sarcoma, the risk of metastasis is so great that it may warrant multiple therapy modalities. In case of suspected cases, an evaluation of the lesion should be carried out using plain films, CTs, MRIs, bone scan, and biopsy. After treatment, it is mandatory to provide suitable prosthesis, so that these young patients lead a quality life.

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