

# Primary osteogenic osteosarcoma of the left zygomatic arch - A case report

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## ABSTRACT

Osteosarcoma accounts for 40–60% of primary malignant bone tumors. About 10% occur in the head and neck region, frequently in the mandible and maxilla. Zygomatic arch is a very rare site of involvement. We report a rare case of primary osteosarcoma of the left zygomatic arch in a 30-year-old male with a review of literature. Only six cases of primary osteogenic sarcoma of zygomatic arch have been reported in the literature. The present case is the 7<sup>th</sup> in the series of reported cases of osteosarcoma of zygomatic arch till date.

**Key words:** Left zygomatic arch, osteogenic osteosarcoma, primary

## INTRODUCTION

Osteosarcoma is a rare but highly malignant bone tumor. It is derived from primitive bone-forming mesenchymal cells with the tendency of bone formation. It is also most common primary malignant lesions of bone in patients under 40 years, excluding multiple myeloma.<sup>[1-3]</sup> It is relatively rare in the craniofacial region constituting about 6.5–7% of all osteosarcomas.<sup>[4]</sup> In the craniofacial region, mandibular tumors arise more frequently in posterior body and ramus; maxillary lesions are more common in the inferior portion (alveolar ridge, sinus floor, and palate) than the superior aspect (zygoma and orbital rim).<sup>[5]</sup> Mandible is more commonly involved than the maxilla. Zygomatic arch is a very rare site of involvement. There are only 6 well-documented cases of osteosarcoma of zygomatic arch in the literature. Average age of onset of osteosarcoma of the jaw is 10–20 years later than that reported for skeletal lesions, and survival rates are higher.<sup>[6-8]</sup>

## CASE REPORT

A 30-year-old male patient presented with left cheek swelling and pain. On examination, firm cheek swelling was subjected to fine-needle aspiration cytology. Cytosmears were fixed and stained with Giemsa stain, revealed polygonal cells in sheets and scattered singly showing pleomorphism in nuclear size, prominent nucleoli, and abundant basophilic cytoplasm. Osteoclastic giant cells and intercellular osteoid was present. On the basis of cytomorphology, diagnosis of osteosarcoma was made [Figures 1 and 2]. The patient was sent for X-ray and computed tomography (CT) scan. CT scan revealed large soft tissue enhancing mass with the new bone formation in left masticator space, arising from left zygomatic arch and mandible with the destruction of the zygomatic arch and left mandibular condyle and angle, body and involving left masseter

muscle, and deep lobe of parotid, extending up to the left submandibular region. True cut biopsy was taken from a lesion at the left zygomatic arch. H and E stained sections showed tumor cells in sheets, oval, spindly, and polygonal in shape, with ample eosinophilic cytoplasm. Marked nuclear pleomorphism and hyperchromasia were seen. Many bizarre tumor cells and occasional tumor giant cells with frequent mitotic figures were seen. Abundant osteoid was seen in most areas [Figure 3]. Thus based on cytomorphology and histomorphology, diagnosis of osteogenic osteosarcoma was confirmed. The patient was subjected to radical removal of the tumor, followed by chemotherapy and radiotherapy.

## DISCUSSION

Osteosarcoma is the most common primary bone malignancy in childhood and adolescence. Osteosarcoma has a bimodal age distribution, having the first peak during adolescence, and the second peak in older adulthood. The incidence is higher in males than in females, with a higher incidence in blacks than in whites. Osteosarcoma commonly occurs in the long bones of the extremities. Other less common sites are the skull or jaw (8%) and pelvis (8%).<sup>[9]</sup> OS accounts for about 20% of all the sarcomas of the skeleton and 6.5–7% of the craniofacial region.<sup>[4]</sup> In craniofacial mandibular tumors arise more frequently in posterior body and ramus, maxillary lesions are more common in inferior portion (alveolar ridge, sinus floor, and palate) than in superior portion (zygoma and orbital rim). As in OS of long bones, craniofacial OS often exhibits cortical destruction and extension into the soft tissue but often do not exhibit periosteal reactive bone.<sup>[10]</sup> Smith *et al.* evaluated 496 cases of the head and neck OS and observed that gnathic and skull-based OS had similar subtypes. High-grade tumors were more commonly encountered in the skull and other craniofacial bones (67%), high-grade histology in extragnathic sites compared with 53% in gnathic sites.<sup>[11]</sup>

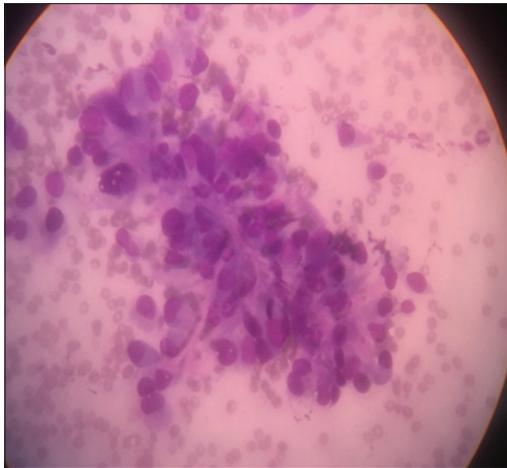
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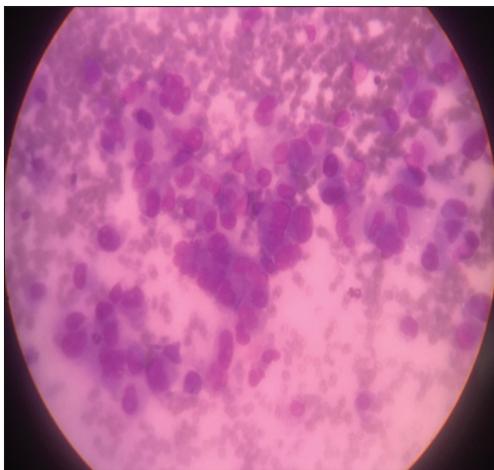
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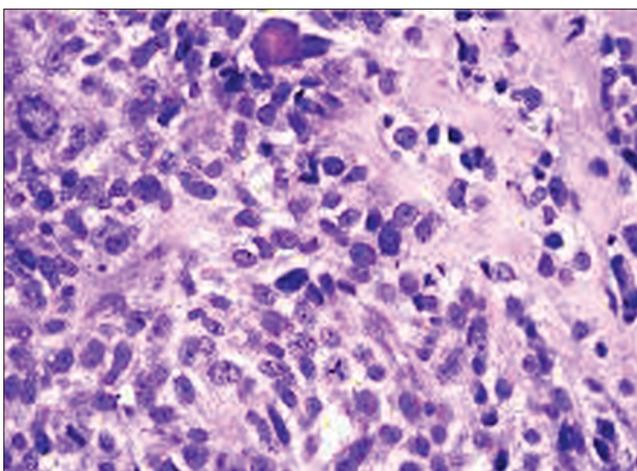
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**Figure 1:** Cytosmears show polygonal cells showing



**Figure 2:** Cytosmears, Giemsa stain,  $\times 40$  pleomorphism in nuclear size and shape, hyperchromatic nucleus, abundant cytoplasm Giemsa stain,  $\times 40$



**Figure 3:** Histology, H and E stain,  $\times 40$

Craniofacial OS are classified as primary and secondary. Primary type occurs *de novo*. Secondary type occurs in patients with

pre-existing diseases such as skeletal Paget's disease,<sup>[12]</sup> fibrous dysplasia of bone,<sup>[13]</sup> or prior regional radiation.<sup>[12]</sup>

Conventional osteosarcomas are osteolytic, blastic or mixed variety. Conventional osteosarcoma is divided into chondroblastic, fibroblastic, and osteoblastic depending on the predominant matrix formation. It is important to differentiate these tumors from chondrosarcoma, fibrosarcoma, chondromyxoid fibroma, intraosseous hemangioma, osteoma, and Ewing's sarcoma as these lesions are described affecting the zygomatic arch.<sup>[14-18]</sup>

As per the available literature, 6 cases of osteosarcoma of zygomatic arch have been reported. This is the 7<sup>th</sup> case in the series to be reported. Osteosarcoma should be considered in the differential diagnosis of lytic lesions of the zygomatic arch with associated soft tissue swelling.

## CONCLUSION

Cytology is primary, less expensive and rapid tool for accurate diagnosis of tumors. In all bony lesions of the face and zygomatic arch, differential diagnosis of osteosarcoma should be kept. Hence, it can be stated that cytomorphology and histomorphology show high accuracy in diagnosis of osteosarcoma. The patient is well after 9 months of completion of chemotherapy and radiotherapy.

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