CASE REPORT

Malignant Small Blue Round Cell Tumor- A Case Report

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Abstract

Introduction: Malignant small round cell tumors are a heterogeneous group of malignant neoplasms presenting predominantly in children and adolescents. They include Ewing sarcoma/peripheral neuroectodermal tumor or Ewing family tumors, lymphoma, mesenchymal chondrosarcoma, and small cell osteosarcoma.

Case presentation: 50yr old patient presented with swelling and pain on right side of the lower jaw since 1 month.

Management and Prognosis: treatment of Ewing's sarcoma is a multidisciplinary approach comprising of chemotherapy, radiotherapy and surgery.

Conclusion: The reported case describes clinical, radiological and histological findings of malignant small round cell tumor.

Keywords- malignant small round cell tumor, undifferentiated tumor, immunohistochemistry

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Introduction

Malignant small round cells tumors (MSRCT) is a term used for tumors composed of malignant round cells that are slightly larger or double the size of red blood cells in air dried smears.[1,2] These cells are undifferentiated in nature which makes the differential diagnosis of small round cell tumors is particularly difficult.[3] Also called as small round blue cell tumors because cells are blue, large hyperchromatic nuclei and a thin rim of cytoplasm. Major group of MSRCT includes lymphoma, leukemia, neuroblastoma, Rhabdomyosarcoma. Overall incidence is 3-5% of all cancers in United States.[4]

Case Report

A 50 year old female reported with complaints of swelling and pain on right side of lower jaw since 1 month. Patient gave a history of burning sensation in the right side of the lower lip. Extra-orally, facial asymmetry was evident on lower one third of the face on the right side. On soft tissue examination we found diffuse swelling on right side of mandibular arch with vestibular obliteration extending from 44 to retromolar area on buccal aspect. Mucosa overlying the swelling appears normal (Figs. 1 and 2). On palpation there was tender bony swelling with diffuse margins on body of mandible and angle of mandible of right side of jaw. Provisional diagnosis as bony tumor of mandible was made with differential diagnosis ameloblastoma, odontogenic keratocyst, central giant cell granuloma, intra alveolar carcinoma of jaw. OPG reveals radiolucency with ill defined borders in right 44,45,46,47,48 extending upto angle of mandible. Thinning out of lower margin of mandible on the right side. The anterior margin of the lesion involves the mental foramen (Fig. 3). Incisional biopsy was done and specimen was sent for histopathological examination (Fig. 4). The histopathology of the soft tissue showed highly cellular tissue showing starry sky pattern consisting of blue round cells, with little intercellular connective tissue stroma. Most of the round cells are plasmacytoma interspersed with few plasmablastic type of cells containing vesicular nuclei with chromatin rim at the periphery and prominent nucleoli. Plasma cells have eccentrically placed nucleoli. Final diagnosis was given as MSRCT (Figs. 5, 6 and 7).

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Discussion

Accurate diagnosis and classification of small-round-cell tumors of childhood is of great importance. Small round cell tumors comprise heterogeneous neoplasms composed of relatively small, round to oval, closey packed undifferentiated cells with high nuclear-cytolasmic ratio, scant cytoplasm, and round nuclei with evenly distributed, slightly coarse chromatin and small or inconspicuous nucleoli.[5]

In spite of a similar light microscopic morphology, SRCTs include pathologic entities from vastly different lineages, including

(1) Epithelial tumors, for example, small cell carcinoma (SmCC) (poorly differentiated neuroendocrine carcinoma);

(2) Mesenchymal tumors encompassing malignant solid neoplasms of childhood and other small round cell sarcomas; and

(3) Tumors with overlapping features, such as lymphoma and melanoma.[6]

Ewing's tumor may arise from long bones and soft tissue. Soft tissues origin of Ewing's is called "extraskeletal Ewing's tumor" (EES). PNET, similar to Ewing's tumor is a round cell tumor originating from neuroectodermal crest. EES and PNET are closely related

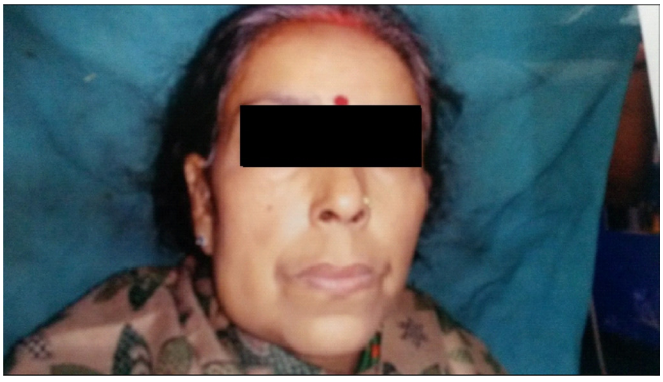

Fig. 1: Extraoral findings - facial asymmetry with diffuse swelling on lower third of face (right side)

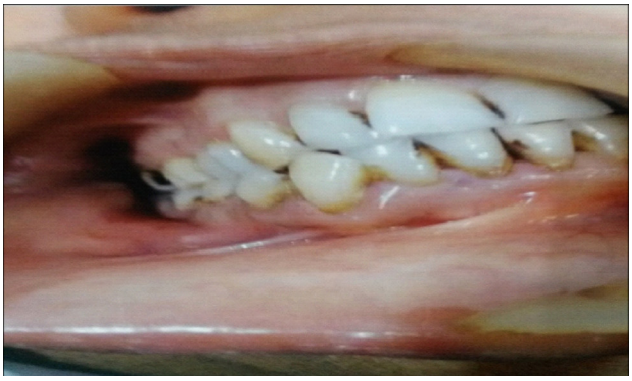

Fig. 2: Intraoral findings - ill-defined swelling on right side of mandibular arch extending till retromolar area.

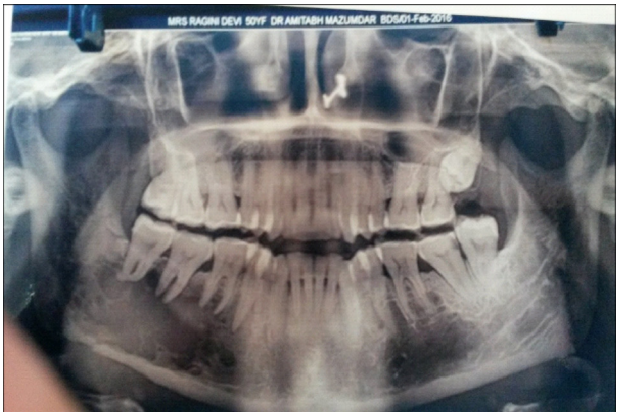

Fig. 3: Orthopantamograph - diffuse radiolucency extending from 44 involving mental foramen upto angle of mandible.

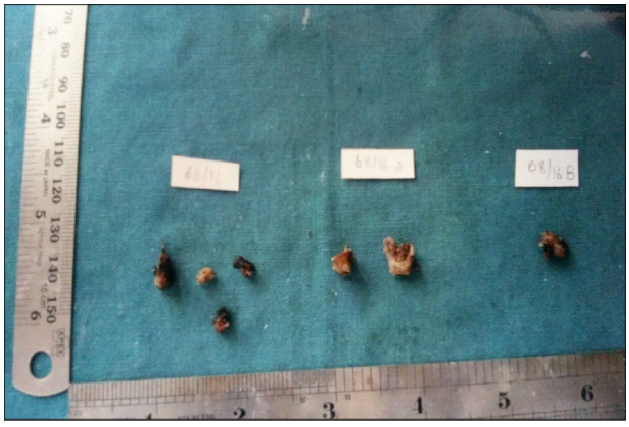

Fig. 4: Gross specimen - multiple soft tissue bits, brownish white in color

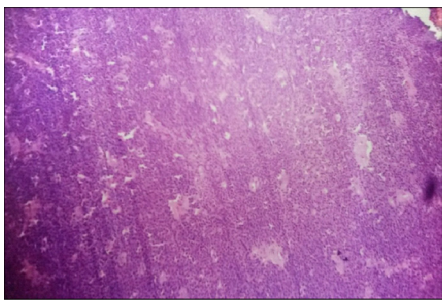

Fig. 5: Round cells with scanty intercellular matrix. (H and E stain, 10X)

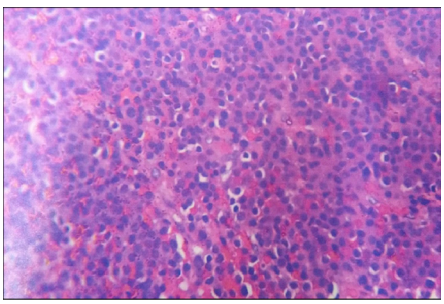

Fig. 6: Sheets of round cell with hyperchromatic basophilic nuclei. (H and E stain, 40X)

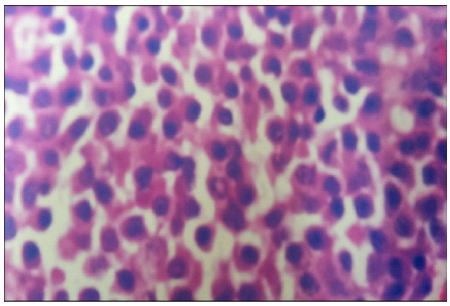

Fig. 7: Sheets of plasmacytoid cells with eccentric and vesicular nucleus. (H and E stain, 40X)

tumors, histologically and have been grouped together as Ewing family of tumors. Diagnosis is routinely based on imaging studies; however, biopsy is essential for definitive diagnosis. The most commonly utilized biopsy techniques are either open biopsy or an imaging-guided core biopsy.[4]

To distinguish between EWS/pPNET and other SRCTs is sometimes difficult by conventional morphologic methods, especially if the tumor arises in an unusual region. The typical PNET may resemble a primitive fibrosarcoma, malignant schwannoma, or malignant fibrous histiocytoma.[7] Because up to 80% of patients with apparently localized disease have occult metastatic disease, multidrug systemic chemotherapy is indicated, as chemotherapy will reduce the tumor size and clear metastasis.[8,9,10]

The present case highlights the features of malignant small round cell tumor. Differential diagnosis is importance in case of MSRCT. And the use of advance diagnostic aid is helpful to find out the lineage of cell involved for accurate diagnosis.

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