

Case Report

FNAC of Extra-Skeletal Ewing's Sarcoma of the Parotid Gland

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ABSTRACT

Extra-skeletal Ewing's sarcoma is a rare soft tissue malignant neoplasm, morphologically indistinguishable from skeletal Ewing's sarcoma. The usual sites of involvement are the soft tissues of para-vertebral region, chest wall, and lower extremity. Extra-skeletal Ewing's sarcoma is rare in the head and neck region and very few cases are reported in the parotid gland. The cytological features of a case of extra-skeletal Ewing's sarcoma involving the parotid gland, an extremely uncommon site for occurrence of this tumor, are reported here. The significance of the fine needle aspiration cytology lies in the early diagnosis and hence better prognosis of this lesion.

Keywords: Ewing's Sarcoma, Parotid Glands, India

Introduction

Non-lymphoid mesenchymal tumors of salivary gland origin are uncommon and primary parotid extra-skeletal Ewing's sarcoma is extremely rare (1). Extra-skeletal Ewing's sarcoma is an aggressive small round cell tumor of neural crest origin. With a change in the modality of treatment by pre-operative chemotherapy, fine needle aspiration cytology can play a useful role in the pre-operative diagnosis with obvious advantage over open biopsy (2). The

cytomorphology of Extra-skeletal Ewing's sarcoma of the parotid gland is presented here.

Case report

A 20-year-old male patient presented with a painless swelling in the region of the left parotid gland of one-month duration. The swelling was firm in consistency and measured 5X3 cm. The swelling was fixed with well-defined edges. The general physical and systemic examinations were normal. The

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routine investigations were within normal limits. Computerized tomography of the region revealed a well-defined low-density lesion, replacing the superficial lobe of the left parotid gland. There was no evidence of osseous or lymph node involvement.

Fine needle aspiration was done with a 24-gauge needle and 10 ml syringe. Hemorrhagic aspirate was obtained. Smears were stained with hematoxylin-eosin (H and E) and May- Grunwald-Giemsa (MGG) stain. Smears were highly cellular showing a mixture of single cells and clusters of loosely

cohesive cells (Fig. 1). A double cell pattern with larger and smaller round cells was noted (Fig. 1). Larger cells had rounded nuclei with finely granular chromatin and distinct small nucleoli. They had abundant pale cytoplasm with vacuoles (Fig. 2). Smaller cells had irregular hyperchromatic nuclei and scant cytoplasm. Many naked nuclei and focal crush artifact were seen. Capillary sized vascular channels with adherent tumor cells were seen (Fig. 2). Cytological diagnosis of extra-skeletal Ewing's sarcoma of parotid gland was suggested.

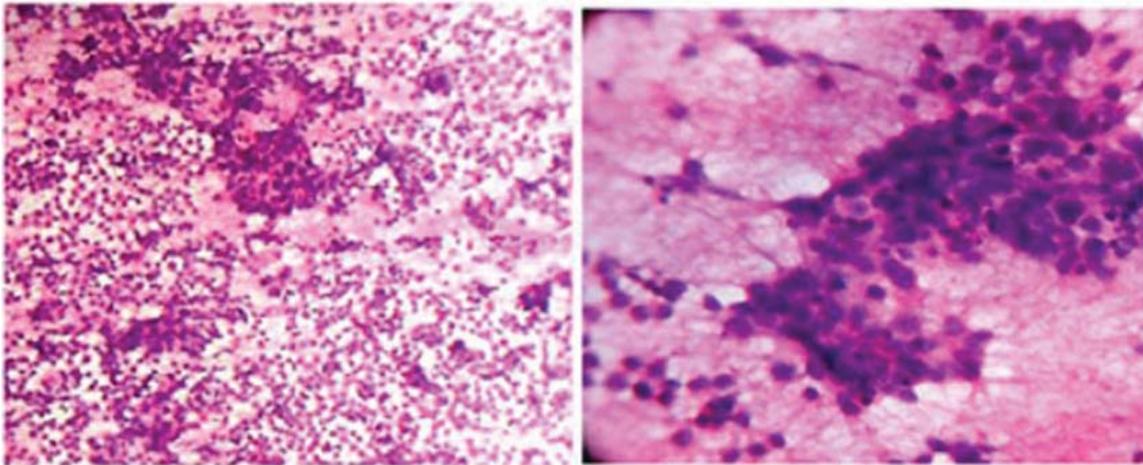


Fig. 1- Left: cellular smear showing mixture of single cells and clusters of loosely cohesive cells. (H and E, $\times 100$) and Right: smear-showing cluster of dual cell pattern (H and E, $\times 400$)

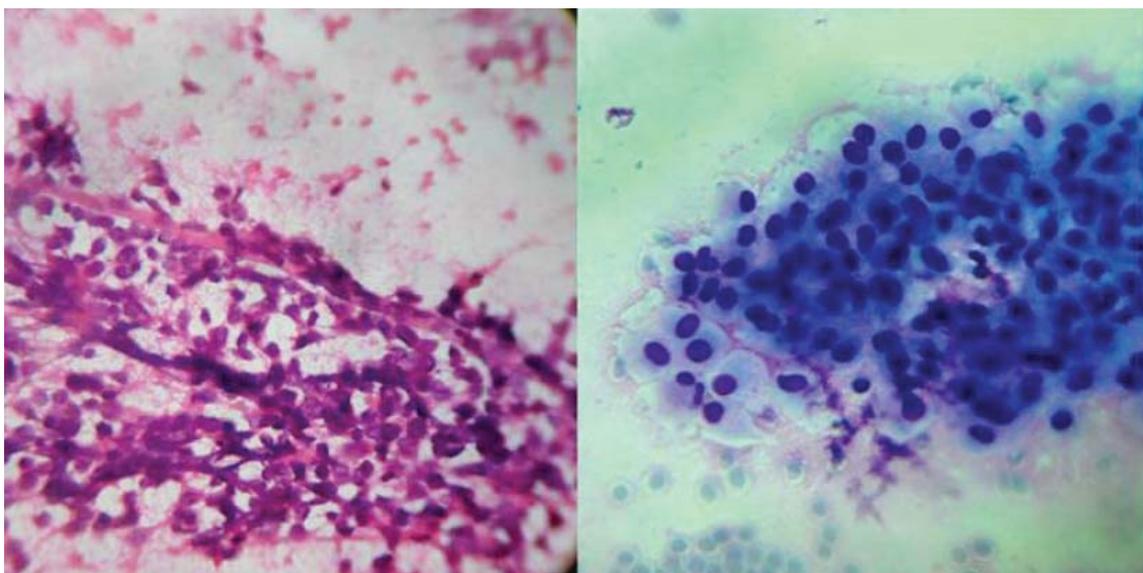


Fig. 2- Left: smear showing vascular channels with adherent tumor cells (H and E, $\times 400$) and Right: smear showing vacuolated cytoplasm (right, MGG, $\times 400$)

Grossly the resected specimen was soft and friable with a gray tan cut surface. Histopathological examination of paraffin embedded, hematoxylin-eosin stained sections showed sheets of uniform small round cells having powdery chromatin and scant pale or irregularly vacuolated cytoplasm. Focal areas of fibrosis, necrosis, and thin walled vessels were seen. Periodic acid- Schiff (PAS) stain showed intracellular glycogen deposition and immunohistochemical analysis showed strong CD99 positivity. The diagnosis of extra-skeletal Ewing's sarcoma was confirmed.

Discussion

Salivary gland tumors are uncommon and most of them are of epithelial origin. Mesenchymal tumors affecting parotid gland are extremely rare. Only a handful of cases of extra-skeletal Ewing's sarcoma affecting parotid gland are reported in the literature. Sandu *et al.* described a case of parotid extra-skeletal Ewing's sarcoma in a 16-year-old girl and the patient presented with facial nerve palsy (3). In our case, the presenting symptom is the parotid swelling and facial nerve palsy was not observed. Deb RA *et al.*, reported a case of primitive neuroectodermal tumor of the parotid gland, showing neural differentiation in the form of rosettes, which were absent in our case (4).

Extra-skeletal Ewing's Sarcoma was first reported by Angervall and Enzinger in 1975 (5). Extra-skeletal Ewing's sarcoma, Ewing's sarcoma, and peripheral neuroectodermal tumor are now considered the same tumor with variable differentiation. Ewing's sarcoma represents the most undifferentiated and the peripheral neuroectodermal tumor represents the most differentiated end of the spectrum (2). In addition to the common histogenesis of neuroectodermal origin, they show chara-

cteristic chromosomal translocation t [11: 22] and are immunoreactive to a product of MIC 2 gene (CD 99 antigen) (6).

The division into skeletal and extra-skeletal Ewing's sarcoma is relevant because the five-year survival in skeletal Ewing's sarcoma is 75% whereas for extra-skeletal Ewing's sarcoma it is 20-30%. This outcome between skeletal and extra-skeletal Ewing's sarcoma is related to the large size, poor circumscision, and deep location of the extra-skeletal Ewing's sarcoma (2). The extra-skeletal Ewing's sarcoma differs from skeletal Ewing's sarcoma in several other respects. The average age of occurrence is 20 years, in contrast to 10 years of skeletal Ewing's sarcoma. Extra-skeletal Ewing's sarcoma occurs equally in both sexes whereas skeletal Ewing's sarcoma has a 2:1 male predilection. Extra-skeletal Ewing's sarcoma involves a wider area as compared to Ewing's sarcoma. The most frequent sites of occurrence are the chest wall, paravertebral area, and lower extremity, but it has been reported in pelvis, retroperitoneum, upper extremity, lung, uterus, ovary, urinary bladder, parotid, and kidney (5).

A cytological diagnosis of extra-skeletal Ewing's sarcoma is suggested by dual population of cells and mixture of single cells and loosely cohesive cells (7), and is identical to the cytological features noted in our case. Malhotra *et al.* noted that the presence of intra-cytoplasmic vacuoles and vascular channels with adherent tumor cells could be used as additional criteria (8). Both vacuoles and vascular channels are observed in our case.

The differential diagnosis of extra-skeletal Ewing's sarcoma includes metastatic neuroblastoma, rhabdomyosarcoma, non-Hodgkin's lymphoma, metastatic small cell carcinoma.

Smears of neuroblastoma show smaller cells with high nucleo-cytoplasmic ratio and Homer-Wright rosettes. Rhabdomyosarcoma shows more pleomorphic cells with dense cytoplasm including some spindle and multinucleated cells. Presence of lymphoglandular bodies can be taken as a clue to the diagnosis of non-Hodgkin's lymphoma. Metastatic small cell carcinoma shows coarser chromatin, indistinct nucleoli and mitotic figures (2, 5).

Extra-skeletal Ewing's sarcoma is a highly aggressive neoplasm that rapidly gives rise to metastatic disease and death. Most common metastatic sites are lung and skeleton. Key prognostic factors that adversely influence the outcome of the disease are the presence of metastatic disease, large tumor size, extensive necrosis, and central axis tumor. Prognosis has improved with the use of multimodality treatment including pre-operative chemotherapy (5).

Despite the appreciable rarity of extra-skeletal Ewing's sarcoma of the parotid gland, it needs to be included in the differential diagnosis of the soft tissue tumors in children and young adults and fine needle aspiration cytology is an accurate procedure for the pre-operative diagnosis of this aggressive tumor.

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