INTRODUCTION

Epithelioid sarcoma is an uncommon slowly growing soft tissue sarcoma, first described as an independent entity by Enzinger.[1] It mostly occurs in dermal or subcutaneous area of the distal extremity of young adults.[1,2] The tumor poses diagnostic difficulties clinically, cytologically as well as histologically thus leading to misdiagnosis. The cytopathologic appearance of epithelioid sarcoma in fine-needle aspiration cytology (FNAC) has not been extensively described.[3] In this communication we are describing the important cytopathologic features of epithelioid sarcoma as FNAC is often the first line of investigation.

CASE REPORT

A fifty year old male presented with recurrent slow growing swelling over lateral aspect of right ankle since five months. There was no other associated symptom. Previous excision biopsy done seven months earlier from other hospital was reported as malignant fibrous histiocytoma. On examination, a 3×3 cm partially ulcerated nodule was present on the lateral malleolus of the right ankle. Complete blood picture and X-ray chest were within normal limits. X-ray right ankle revealed a soft tissue shadow. The lesion was subjected to fine needle aspiration cytology (FNAC).

The FNAC revealed dispersed round to polygonal cells with interspersed spindle cells which showed mild to moderate pleomorphism. Nuclei of the cells were large and eccentrically located giving the cells a plasmacytoid appearance. Cytoplasm was abundant and at places showed a vacuolated peripheral rim. Necrosis was present in the background [Figure 1]. The diagnosis of malignant soft tissue tumor was made.

Subsequent incisional biopsy from the lesion yielded multiple grey white soft tissue pieces together measuring 2×1×1 cm. On Hematoxylin and eosin (H and E) staining nodular pattern of the tumor showed mixed proliferation of oval to polygonal cells with moderate to abundant eosinophilic cytoplasm with vesicular nucleus, moderate atypia and small nucleolus [Figure 2]. Tumor cells were arranged around geographic areas of necrosis. Epithelial cells blended with spindle cells especially at the periphery of the tumor, which also showed inflammatory cells. 3-4 mitoes per high power field were noted. A diagnosis of malignant soft tissue tumor was made and various differentials were considered including epithelioid sarcoma, monophasic synovial sarcoma, epithelioid MPNST, epithelioid hemangioendothelioma/
angiosarcoma, epithelioid leiomyosarcoma, metastasis, MFH and pleomorphic rhabdomyosarcoma. Special histochemical stains were carried out using a panel of IHC markers. These included cytokeratin [Figure 3], EMA, vimentin [Figure 4], HMB-45 and S100. In our case cytokeratin, EMA and vimentin were found to be positive whereas HMB-45 and S-100 were negative. Thus the final diagnosis of epithelioid sarcoma was made.

**DISCUSSION**

Epithelioid sarcoma is a rare soft tissue sarcoma which was described as a distinct entity by Enzinger in 1970.[1] It accounts for less than 1% of the soft tissue neoplasms.[2] Median age of presentation is 26 years with male predominance.[2,4] Distal extremities are commonly involved.[2] The proximal-type variant, first described in 1997 as a rare aggressive form of sarcoma, usually arises more proximally.[5] The lesion poses diagnostic difficulties both clinically and histologically. It presents as a slowly growing painless nodule with ulceration easily mistaken for inflammatory process like indurated ulcer, infected wart, ulcerated squamous cell carcinoma or granulomatous process.[4] In comparison with a granuloma, cells in epithelioid sarcoma are more sharply defined, larger, more eosinophilic and less mature. The natural history of epithelioid sarcomas has shown a recurrence rate of 50% to 80% with metastasis documented in 40-67% cases.[6,7] Prognosis is dependent on the depth of the lesion in relation to deep fascia, local recurrence and regional lymph node involvement.[4,8,9] The size of the primary lesion is not a reliable indicator of the prognosis but smaller tumors are associated with significantly better metastasis free survival.[10]

**Role of cytology**

Role of FNAC in diagnosis of soft tissue tumor has been fairly documented as well as debated. Cytological evaluation plays an important role in detection of recurrent as well as metastatic soft tissue tumors.[11] Reaching a
diagnosis of epithelioid sarcoma can be concurrently helped by cytological assessment which shows cellular smears comprised of dispersed sheets or three dimensional clusters of round/polygonal/epithelioid to spindled cells showing mild to moderate pleomorphism. Nuclei of the cells are large and eccentrically located with a plasmacytoid appearance. Cytoplasm appears eosinophilic with at places showing a vacuolated peripheral rim. Granuloma like structures may be seen. However with these features morphological overlap can be seen while subtyping the lesions especially epithelioid type and round cell type thus limiting the utility of cytology and giving rise to both malignant and benign lesions in differential diagnosis.

Cytomorphologically the most common malignant differential diagnosis are squamous cell carcinoma, amelanotic melanoma, adenocarcinoma, epithelioid schwannoma, and leiomyosarcoma. Further the epithelioid nature of the cells may not be cytologically obvious and the lesion can be confused with a benign epithelial neoplasm or a reactive histiocytic process. This further brings up the role of immunocytochemistry in evaluation of the above lesion which can serve as a good adjunct to cytology.

Thus identification of epithelioid sarcoma can be challenging, on cytology however with a good clinicopathological correlation and immunocytochemistry are invariably useful.

Treatment of the primary disease is wide local excision, followed by adjuvant radiotherapy. Amputation is required in recurrence. Chemotherapy is recommended for metastasis includes ifosfamide and doxorubicin.

REFERENCES


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