Case Report

Cytomorphology of epithelioid sarcoma: A diagnostic enigma

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ABSTRACT

Epithelioid sarcoma is an uncommon soft tissue malignancy with a known propensity for recurrence as well as metastasis. We report epithelioid sarcoma in 50 year old male with a recurrent ulcerative nodule over the right ankle since five months, initially misdiagnosed as malignant fibrous histiocytoma. The article aims at assessing the key cytological features of epithelioid sarcoma and their reliability in the diagnosis of the lesion. Also described are its histopathological and immunohistochemical features.

Key words: Cytomorphology, epithelioid sarcoma, histiocytoma, malignant fibrous

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 cytologic appearance of epithelioid sarcoma in fine-needle aspiration cytology (FNAC) has not been extensively described.[3] In this communication we are describing the important cytological 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/

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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

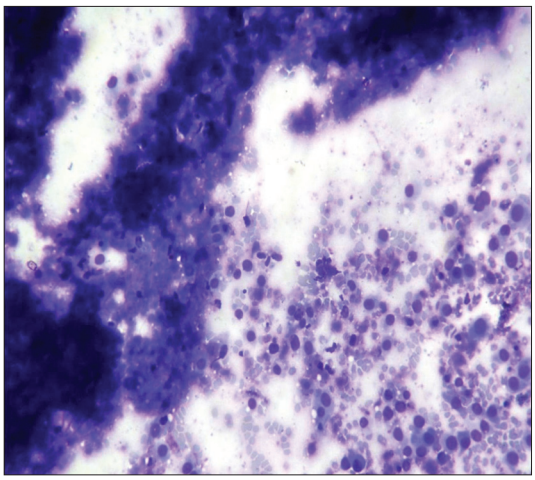

Figure 1: Epithelioid cells with necrosis in the background. Giemsa ×400

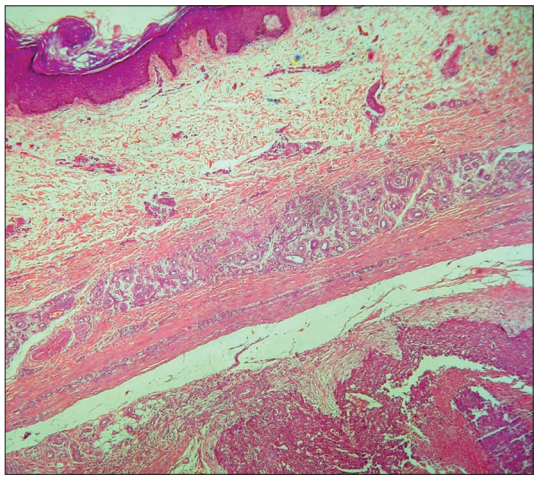

Figure 2: Section showing skin and appendages covering nodular pattern of the tumor containing mixed proliferation of oval to polygonal cells and necrosis. H and E,×100

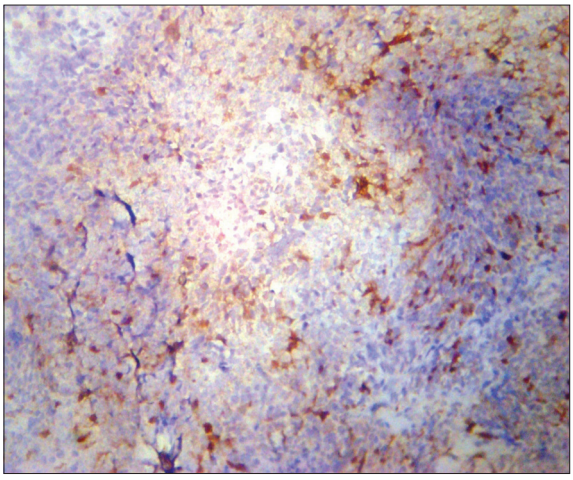

Figure 3: Immunohistochemistry showing that the tumor cells are moderately positive for cytokeratin AE1-AE3

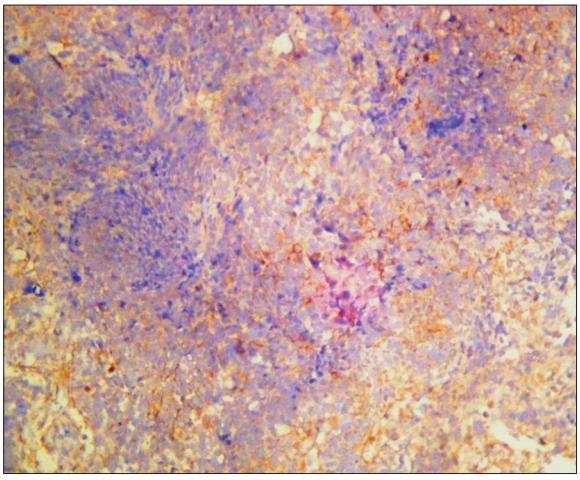

Figure 4: Immunohistochemistry showing that the tumor cells are moderately positive for viamentin

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.[12,13] 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.[14] 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.[14] 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.[15] 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

1. Enzinger FM. Epitheloid sarcoma. A sarcoma simulating a granuloma or a carcinoma. Cancer 1970;26:1029-41.
2. Chase DR, Enzinger FM. Epithelioid sarcoma. Diagnosis, prognostic indicators, and treatment. Am J Surg Pathol 1985;9:241-63.
3. Barwad A, Dey P, Das A. Fine needle aspiration cytology of epithelioid sarcoma. Diagn Cytopathol 2011;39:517-20.
4. Prat J, Woodruff JM, Marcove RC. Epithelioid sarcoma: An analysis of 22 cases indicating the prognostic significance of vascular invasion and regional lymph node metastasis. Cancer 1978;41:1472-87.
5. Woods RS, Dempsey MP, Rizkalla HF, McMenamin ME, O'Donovan D. Proximal-type epithelioid sarcoma: Case report of an unusual presentation. J Plast Reconstr Aesthet Surg 2012;65:977-80.
6. Chase DR, Enzinger FM. Epithelioid sarcoma. Diagnosis, prognostic indicators, and treatment. Am J Surg Pathol 1985;9:241-63.
7. deVisscher SA, van Ginkel RJ, Wobbes T, Veth RP, Ten Heuvel SE, Suurmeijer AJ, *et al*. Epithelioid sarcoma: Still an only surgically curable disease. Cancer 2006;107:606-12.
8. Bos GD, Pritchard DJ, Reiman HM, Dobyns JH, Ilstrup DM, Landon GC. Epithelioid sarcoma. An analysis of fifty-one cases. J Bone Joint Surg Am 1988;70:862-70.
9. Callister MD, Ballo MT, Pisters PW, Patel SR, Feig BW, Pollock RE, *et al*. Epithelioid sarcoma: Results of conservative surgery and radiotherapy. Int J Radiat Oncol Biol Phys 2001;51:384-91.
10. Evans HL, Baer SC. Epithelioid sarcoma: Aclinicopathologic and prognostic study of 26 cases. Semin Diagn Pathol 1993;10:286-91.
11. Willén H, Akerman M, Carlén B. Fine needle aspiration (FNA) in the diagnosis of soft tissue tumours; a review of 22 years experience. Cytopathology 1995;6:236-47.
12. Zeppa P, Errico ME, Palombini L. Epithelioid sarcoma: Report of two cases diagnosed by fine-needle aspiration biopsy with immunocytochemical correlation. Diagn Cytopathol 1999;21:405-8.
13. Lemos MM, Chaves P, Mendonça ME. Is preoperative cytologic diagnosis of epithelioid sarcoma possible? Diagn Cytopathol 2008;36:780-6.
14. Pohar-Marinsek Z, Zidar A. Epithelioid sarcoma in FNAB smears. Diagn Cytopathol 1994;11:367-72.
15. Cardillo M, Zakowski MF, Lin O. Fine-needle aspiration of epithelioid sarcoma: Cytology findings in nine cases. Cancer 2001;93:246-51.

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