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EPITHELIOID SARCOMA: CYTOLOGICAL DIAGNOSIS OF A RARE ENTITY.

Pathology

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ABSTRACT

Epithelioid sarcoma is a rare and aggressive soft tissue tumour exhibiting morphologic and immunophenotypic epithelioid differentiation and molecularly characterised by the loss of expression of SMARCB1, a key member of the SWItch/Sucrose Non-Fermentable (SWI/SNF) chromatin remodelling complex. Very few cases of Epithelioid sarcoma have been reported in literature and has been poorly studied, resulting in limited treatment options. The cytologic appearance of epithelioid sarcoma in fine-needle aspiration cytology (FNAC) has not been extensively described. In this article we are describing the important cytological features of epithelioid sarcoma as FNAC is usually the first line of investigation in soft tissue lesions.

KEYWORDS

Epithelioid sarcoma, Cytology, SMARCB1.

INTRODUCTION

Epithelioid sarcoma is a malignant mesenchymal neoplasm with morphologic and immunophenotypic epithelioid differentiation, which was first defined as a distinct clinicopathologic entity by Enzinger in 1970.(1)

It is Characterized by epithelioid cytomorphology, tumor necrosis, epithelial marker expression, and inactivation of the hSNF5/SMARCB1/INI1 gene. There are two distinct clinicopathological subtypes: the classic type and the proximal type. Differentiated by typical anatomic locations, histopathologic features, and prognosis. Classic type being more common in young adults in the second to fourth decades of life in distal extremities with a male:female ratio of approximately 2:1, and the proximal type, tends to occur in older adults in the proximal soft tissues.(2)

The cytologic appearance of epithelioid sarcoma in fine-needle aspiration cytology (FNAC) has not been extensively described. In this article we are describing the important cytological features of epithelioid sarcoma as FNAC is usually the first line of investigation in soft tissue lesions.

Case

A 27 year old male patient presented to Dermatology OPD with complaints of multiple nodular swellings with ulceration and discharge involving the flexor aspect of the left arm and forearm for a duration of 6 months, which started as a single nodular lesion and progressed to multiple lesions with ulceration and discharge over 2 to 3 months, not associated with pain and tenderness. He had history of trauma to the forearm with fracture of radius 15 years back.

On CT scan multiple heterogeneously enhancing lesions were noted in the subcutaneous plane. Heterogeneously enhancing enlarged left axillary lymph nodes were seen and few heterogeneously enhancing lesions were seen scattered in the lung parenchyma.

FNAC was performed of the skin nodule which in consideration with the clinical presentation(fig-1), radiological features and cytomorphology(fig-2) was reported as epithelioid sarcoma and confirmed by histopathology (Fig-3) on biopsy.

Cytological picture shows cellular smears comprising of dispersed sheets or three-dimensional clusters of round /polygonal/epithelioid to spindle 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.

Fig-1: Multiple Nodular Swellings With Ulceration And Discharge Involving The Flexor Aspect Of The Left Forearm.

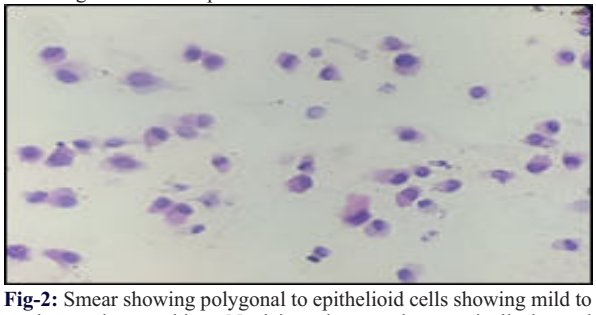

Fig-2: Smear showing polygonal to epithelioid cells showing mild to moderate pleomorphism, Nuclei are large and eccentrically located with a plasmacytoid appearance, Cytoplasm appears eosinophilic.

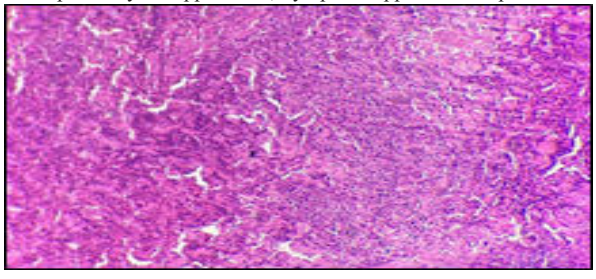

Fig-3: Low power view of the nodule shows tumour cells arranged in

nodular pattern, cells have epithelioid appearance with abundant eosinophilic cytoplasm.

DISCUSSION

We report a case of conventional type of Epithelioid sarcoma involving the left arm and forearm in a 27 year old male. Enzinger described the first case of epithelioid sarcoma in 1970 showing characteristic histologic features, tumour cells having epithelioid appearance arranged in nodular pattern with tendency to undergo central necrosis. Tumour cells were described to vary from large ovoid to polygonal with eosinophilic cytoplasm, to plump spindle-shaped cells.

Clinically, Conventional ES usually presents as painless, slow-growing, firm nodules often accompanied by superficial ulceration, hemorrhage, necrosis, and plaques.(4)

Diagnosis of this lesion is difficult both clinically and histologically due to its presentation mimicking inflammatory processes like granulomatous process, infected wart, ulcerated squamous cell carcinoma or indurated ulcer.(3)

In comparison with a granuloma, cells in epithelioid sarcoma are more sharply defined, larger, more eosinophilic and less mature. Recently, three less common variants lacking the characteristics of the granulomatous appearance have been described as fibroma like, angiomatoid and rhabdoid.

The natural history of epithelioid sarcomas have shown a recurrence rate of 50% to 80% with metastasis documented in 40-67% cases.(5,6) Prognosis is dependent on the depth of the lesion in relation to deep fascia, local recurrence and regional lymph node involvement(7,8,9).

The role of FNAC is debatable in diagnosis of soft tissue tumours ,but it is an essential modality for detection of recurrence and metastatic soft tissue tumors.(10)

Morphological overlap can be seen while subtyping the lesions, thus limiting the utility of cytology giving rise to both malignant and benign lesions in differential diagnosis. Thus making identification of epithelioid sarcoma challenging on cytology, however a good clinicopathological correlation and immunocytochemistry are invariably useful.

Molecularly epithelioid sarcoma is characterised by the loss of expression of SMARCB1, a key member of the SWItch/Sucrose Non-Fermentable (SWI/SNF) chromatin remodelling complex.

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. Epithelioid sarcoma: a sarcoma simulating a granuloma or a carcinoma. Cancer. 1970;26:1029-1040.
(2) Guillou L, Wadden C, Coindre JM, Krausz T, Fletcher CD. "Proximaltype" epithelioid sarcoma, a distinctive aggressive neoplasm showing rhabdoid features. Clinicopathologic, immunohistochemical, and ultrastructural study of a series. Am J Surg Pathol. 1997;21: 130-146.
(3) 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.
(4) Henry BA, Anil VP. Epithelioid sarcoma. Arch Pathol Lab Med. 2009; 133:814-819.
(5) Chase DR, Enzinger FM. Epithelioid sarcoma. Diagnosis, prognostic indicators, and treatment. Am J Surg Pathol 1985;9:241-63.
(6) 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.
(7) 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.
(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) 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.