ATYPICAL FIBROXANTHOMA

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A case of atypical fibroxanthoma is being reported in an elderly man. Though histologically it resembles malignancy, clinically it is benign.

Key words : Atypical fibroxanthoma, Pseudosarcomatous fibrous histiocytoma

Introduction

Atypical fibroxanthoma (AFX) is a tumour of the dermis, seen in adults, that follows a biologically indolent course and is characterised histologically by a population of fusiform, epithelioid and giant cells with marked atypia, numerous cells in mitosis and frequent lipidization closely mimicking malignant fibrous histiocytoma (MFH). Although the morphological features of AFX and MFH are indistinguishable, their clinical presentation and course are quite different. AFX has a good prognosis with some tendency for local recurrence, by contrast MFH is a high grade sarcoma.

Case Report

A 69-year-old male presented with a slow growing nodule on the right temple. It measured 2 cm in diameter. No lymphdenopathy was noted. The patient was otherwise in good health. An excisional biopsy was performed. On the cut surface, the specimen had a firm, pink- gray nodule that was well-delineated from the surrounding skin. No follow - up information is availabe.

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On light microscopy a well-circumscribed dermal tumor extending from the dermoepidermal junction and involving the reticular dermis was seen. The tumor was composed of fascicles of spindle cells focally forming a cart wheel pattern (Fig.1). The spindle cells showed nuclear pleomorphism and atypia.Mitoses, some atypical, was present. Bizarre tumor giant cells were also noted (fig.2).Osteoclast-like giant cells were also seen uniformly distributed within the tumor. On immunohistochemistry the tumor cells showed a positive reaction for vimentin, lysozyme, and alpha-1-an titrypsin. Based upon the characteristic his-

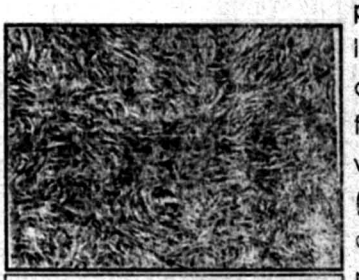

Fig-1: Microphotograph showing fascicles of pleomorphic spindle cells forming a cartwheel pattern. (HE,40X)

Fig-2: Pleomorphic spindle cells and giant cells with scattered mitoses including atypical forms. (HE,100X)

tological findings, supported by immunohistochemistry a diagnosis of atypical fibroxanthoma was offered.

Discussion

When Helwig first described AFX in 1963, it was interpreted as a benign reactive lesion. It has also been termed as atypical fibrozanthoma, dermatofibrosarcoma with monster cells, and pseudosarcomatous fibrous histiocytoma.[2] Although this lesion occurs perdominantly in elderly patients on a clnically damaged skin of the head and neck area, it may also be seen in younger individuals on non-sun-exposed sites such as the trunk and extremities.

The lesions range in size from 1-6 cms but most are in the range of 1-2 cms. If intact,the overlying skin is smooth and may have a yellow hue. The clinical differential diagnosis would include squamous cell carcinoma, melanoma and lobular capillary hemangioma.

Histopathologically it is an unencapsulated densely cellular tumour. The superficial epithelium may ulcerate but there is no direct continuity with the epidermis. Some lesions have a Grenz zone but many lesions abut upon the epidermis. The architectural growth pattern may be haphazard, fascicular or storiform. It shows atypical cytological features characterised by marked cellularity, pleomorphic spindle cells, bizarre hyperchromatic giant cells, histiocytes with large vesicular nuclei and scattered mitoses, including atypical forms.

The progenitor cell is an undifferentiated mesenchymal cell capable of showing histiocytic, fibroblastic and myofibroblastic differentiation.[4]

AFX despite apparently malignant histologic features, usually follows an indolent locally aggressive course. Local excision is curative in virtually evey case. The superficial lesions rarely recur.[4] AFX is regarded as a neoplasm of low-grade malignancy indistinguishable from MFH on histology, the more favourable prognosis being related to its size and surperficial location.[5]

To conclude it is important to be aware of this tumour so that it can be included in the appropriately ordered differential diagnosis of dermal tumors. Most important is its histologic feature that closely mimic a high grade sarcoma (MFH); however its biological progression is entirely different. Awareness of this rare entity will prevent mistaken diagnosis and unnecessary surgery.

References

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