Letter to Editor

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Atypical Fibroxanthoma of Scalp: A Paradoxical Benign Tumour

Pathology Section

RITESH SACHDEV[1], RUCHIKA GOEL[2], SMEETA GAJENDRA[3]

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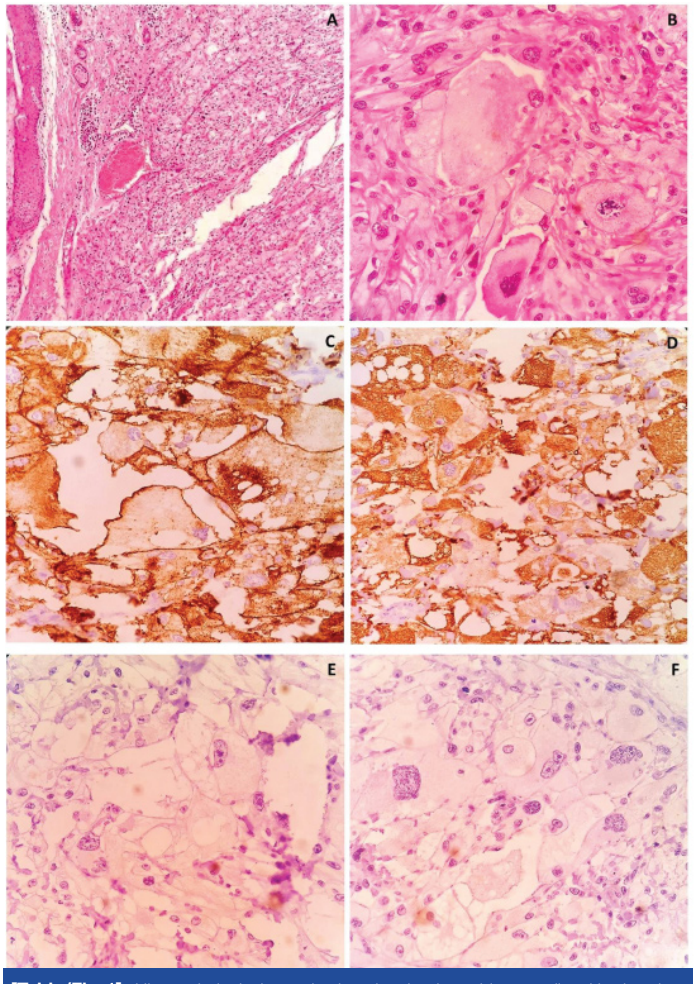
Atypical fibroxanthoma (AFX) is a recently described soft tissue tumour by World Health Organization as a benign tumour of uncertain differentiation, occurring on sun exposed skin in elderly individual [1]. The bizarreness as well as marked atypia seen in the cells raises a first impression of malignant tumour, whereas it is rarely so. It has a male predominance (70%) with an average age of presentation of 71.9 years, ranging from 29–91 years [2].

An 81-year-old male presented with hard nodular scalp swelling since 7 to 8 years with a rapid size increase since 3 months. On examination there was a nodulo-ulcerative lesion measuring 5cm x 3cm on scalp with bleeding edges. He did not have any other cutaneous lesion or lymphadenopathy. Excisional biopsy done outside was reported as sebaceous carcinoma. The patient was referred to our hospital for further management. On review of biopsy slides, the tumour cells were spindled, plump to epithelioid with marked pleomorphism, arranged in haphazard, vaguely fascicular or storiform patterns [Table/Fig-1a]. Tumour cells had hyperchromatic and multilobulated bizarre nuclei with scattered multinucleate tumour giant cells. Some cells of the lesion contained vacuolated and lipid-containing cytoplasm similar to xanthoma raising doubts of sebaceous carcinoma, as diagnosed outside [Table/Fig-1b]. On extensive IHC work up, the tumour cells were positive for Vimentin (Dako;V9) [Table/Fig-1c], CD68 (Leica;514H12), CD10 (Dako;56C6) [Table/Fig-1d] and negative for Cytokeratin (Biogenix;AE1+AE3) [Table/Fig-1e], EMA (Dako, E29), SMA (Dako;1A4), S 100 (Dako;IS504), Desmin (Dako;D33), Caldesmon (Dako; H-CD) and HMB 45 (Dako;HMB45) [Table/Fig-1f]. A final diagnosis of atypical fibroxanthoma was made.



[Table/Fig-1]: Histopathological examination showing large bizarre cells with abundant foamy cytoplasm and atypical mitosis (H&E: a) 100x; b) 400x). Immunohistochemistry showing positivity for Vimentin (c), CD10 (d) and negativity for Cytokeratin (e), HMB45 (f)

Atypical fibroxanthoma	Malignant fibrous histiocytoma	Atypical Fibrous Histiocytoma / Dermatofibroma with Monster Cells	Pleomorphic Fibroma
Commonly involves scalp and neck	Commonly involves extremities	Commonly involves extremities and trunk	Extremities and trunk> scalp and neck
Usually abuts epidermis (commonly ulcerated) Minimal involvement of subcutis. No muscle or vascular invasion	Extension to deep cutaneous tissue, fascia, muscle with vascular invasion	Grey zone between lesion and epidermis	Grey zone between lesion and epidermis
Have xanthoma cells	Lacks xanthoma cells	Lacks xanthoma cells	Lacks xanthoma cells
CD68 positive	CD 68 positive	CD68 negative	CD68 negative
Mitotic figure numerous, often atypical	Mitotic figure numerous, often atypical	Mitotic figure rare	Mitotic figure rare

[Table/Fig-2]: Differential diagnoses of atypical fibroxanthoma

Making a diagnosis of atypical fibroxanthoma is challenging and the diagnosis should be made by exclusion of other differential diagnoses after applying the stringent histological criteria and a broad panel of immunostains. Cutaneous carcinomas and melanoma must be ruled out by immunohistochemistry. Absent immunostaining for cytokeratins, S100 and HMB45 in AFX are helpful for excluding sebaceous carcinoma along with squamous cell carcinoma and malignant melanoma. Other differentials are malignant fibrohistiocytoma, atypical Fibrous Histiocytoma / dermatofibroma with monster Cells and pleomorphic fibroma [Table/Fig-2]. By definition, in AFX tumour cells lack expression of S100, cytokeratin, CD34, desmin and h-caldesmon. Previously our case was misdiagnosed as sebaceous carcinoma due to pleomorphism, abundant clear to foamy cytoplasm and bizarre pleomorphic nuclei. CD10 is a useful marker for AFX and is

positive in 95%–100% cases of AFX [2]. A histiocytic marker CD 68 is positive in more than half of cases [3]. AFX shows a very good prognosis after surgical excision [4]. Reports of metastasis are very rare, and recurrence is uncommon [5]. Distinction from high grade sarcoma is crucial to prevent inappropriate aggressive therapy.

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PARTICULARS OF CONTRIBUTORS:

1. Senior Consultant, Department of Pathology and Laboratory Medicine, Medanta- the Medicity, Sector 38, Gurgaon, Haryana. India.
2. Consultant, Department of Pathology and Laboratory Medicine, Medanta- the Medicity, Sector 38, Gurgaon, Haryana. India.
3. Associate Consultant, Department of Pathology and Laboratory Medicine, Medanta- the Medicity, Sector 38, Gurgaon, Haryana. India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Ritesh Sachdev,
Senior Consultant, Department of Pathology and Laboratory Medicine,
Medanta-The Medicity, Sector – 38, Gurgaon, Haryana-122 001, India.
E-mail: sachdev05@gmail.com

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