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Atypical Fibroxanthoma of the Scalp in a Young Woman: A Case Report

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Abstract

Atypical fibroxanthoma, is an uncommon tumor of fibrohistiocytic mesenchymal origin that occurs on the sun-damaged skin of elderly men. The clinical presentation is a solitary cutaneous nodule. The diagnosis is made by histology and immunohistochemistry. The large excision is the mainstay therapy and the recurrence is possible. We report a case of atypical Fibroxanthoma occurring in the scalp of a young woman.

Keywords

Atypical Fibroxanthoma; Mesenchymatous; Scalp

Introduction

Atypical Fibroxanthoma (AFX) is a rare parenchymatous tumor, it is considered by some authors as a low-grade sarcoma [1]. This cutaneous neoplasm originates from fibro histiocytic mesenchymatous tissue [2]. It affects elderly men, in photo damaged area, such are head and neck. This tumor is characterized by a favorable prognosis and a rare risk of metastasis [3].

Case Report

A young woman of 28 years old, was referred for a solitary nodule of the scalp, evolving since 2 months, without pain or pruritus, the dermatological examination showed a nodule of the frontal scalp of 1 cm size (Fig.1), erythematous and slightly ulcerated, painless. It was superficial, without subjacent involvement. The rest of the somatic examination did not show any abnormalities. In front of this clinical presentation we evoked a proliferating trichilemmal cyst, a squamous cell carcinoma, an achromic melanoma, our patient benefited from a biopsy that showed a proliferation of pleomorphic atypical spindle-shaped cells (Fig.2). In Immunohistochemistry, the HMB 45 was negative, eliminating an achromic melanoma (Fig. 3), the cytokeratin was negative too eliminating a squamous cell carcinoma (Fig. 4). While the fixation of CD68 (Fig. 5) confirmed our diagnostic of AFX. The patient benefited from cervical ultrasonography that didn't show any ganglionic involvement, then the tumor was excised with 1 cm margins. The first control was without abnormalities, the scars didn't show recurrence signs. A regular follow up is conducted for our patient (Fig. 6).

Figure 1: Solitary erythematous superficial nodule of the scalp.

Figure 2: Immunostain HES X200: non epithelial dense proliferation, made by large spindle shaped cells.

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Figure 3: Immunostain X200, negative fixation of HMB 45.

Figure 4: Immunostain X200, negative fixation of cytokeratin.

Figure 5: Immunostain X200, positive fixation of CD68 confirming the diagnosis of atypical fibroxanthoma.

Figure 6: First control after large excision of the tumor.

Discussion

Atypical Fibroxanthoma (AFX) is a rare distinctive cutaneous neoplasm of fibrohistiocytic mesenchymal origin. The oncogenetic background of this tumor demonstrates C-T mutation in P53 and TERT promoter genes which explain the location in actinically damaged skin [4,5]. Moreover, the occurrence of this tumor in patients with xeroderma pigmentosum supports this hypothesis. Radiation, burn traumatism and maybe the risk factor of the occurrence of this tumor [6]. Its pathogeny is still unclear, and it may originate from myofibroblasts or apparent fibroblast cells [7]. Although this tumor affects elderly men, there is a small subgroup of affected younger patients as was the case of our patient [1]. Clinically, AFX appears as a solitary ulcerated nodule, of 1.5-2 cm size, in a sun-damaged area, mimicking a squamous cell carcinoma or basal cell carcinoma as was the case of our patient [1]. Histological features of this tumor include the presence of dense cellularity with haphazardly ranged, atypical spindle-shaped cells just beneath the epidermis, extending to the dermis and hypodermis [2]. However, this description is not specific to AFX, it can be found in the other spindle cell tumors, especially the pleomorphic dermal sarcoma which share a clinical, histological and either immunostain characteristics with AFX [8]. The management of this tumor is a total excision, while the margins are still unprecise. The risk of recurrence is variable through the reported cases and it is ranging from 3 to 20% [8,9].

Conclusion

Atypical Fibroxanthoma (AFX) is a rare neoplasm that may occur in young patients and should be evocated in front of a solitary nodule in the sun-exposed area.

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