Journal of the Turkish Academy of Dermatology
eISSN 1307-394X

Case Report
DOI: 10.6003/jtad.1261c2

Atypical Fibroxanthoma

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Published:
J Turk Acad Dermatol 2012; **6 (1)**: 1261c2
This article is available from: http://www.jtad.org/2012/1/jtad1261c2.pdf
Key Words: skin neoplasm, atypical fibroxanthoma

Abstract

Observation: Atypical fibroxanthoma (AFX) is a rare tumor that usually arises in sun-damaged skin of the head and neck in elderly people. A 50-year-old male patient presented with a crusted bump on the ear, which had appeared and begun to grow in the course of the previous year. Dermatological examination revealed a dome-shaped, crusted, erythematous, 8 mm in diameter, painless, solitary nodule on the left ear antitragus. Our prediagnoses were basal cell carcinoma, cutaneous leishmaniasis and angiolymphoid hyperplasia. The punch biopsy specimen obtained from the lesion showed that large eosinophilic cells had vacoulated cytoplasms, large nuclei and pleomorphism in the papillary dermis. Giant cell formations and a great number of mitoses were seen in the lesion. Immunohistochemically, it stained positive with S100, but it stained negative with keratin, EMA and desmin. The lesion was diagnosed as AFX and totally removed by excision.

Introduction

Atypical fibroxanthoma (AFX) is a rarely seen skin tumor, which generally occurs on the sundamaged skin of the head and neck of the elderly as a solitary ulcerated nodule. Because of histopathological and immunohistochemical similarities, AFX is recently described as a superficial form of malignant fibrous histiocytoma [1]. We hereby report a case of AFX on the left antitragus.

Case Report

A 50-year-old male patient presented with a crusted nodule on the ear, which had appeared and started to grow during the last year. Physical examination revealed an 8 mm diameter, well-circumscribed, dome-shaped, crusty, erythematous, solitary nodule with a central ulcer on the antitra-

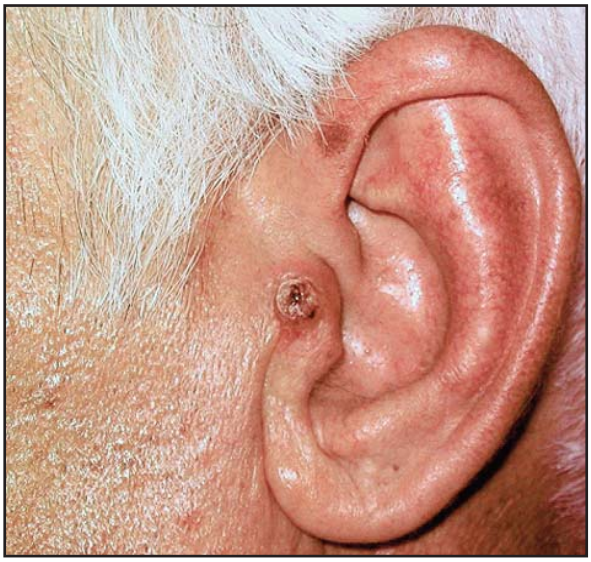

Figure 1. Ulcerated, dome-shaped, solitary nodule on the antitragus of the left ear

gus of the left ear (**Figure 1**). A few papules resembling opaque pearl were detected around the ulcer with a diameter of 1 mm. Multiple actinic keratotic lesions on the facial region and cutis romboidalis nuchae with actinic damage on the neck were observed. The skin examination was normal in the palm-plantar region, hairy skin, oral mucosa and all the nails. Systemic physical findings were normal. There were no lymphadenopathies. Complete blood count, erythrocyte sedimentation rate, and blood biochemistry values were all within the normal range. Punch biopsy specimen obtained from the edge of nodular lesion on the left antitragus rendered the prediagnoses of basal cell carcinoma, cutaneous leishmaniasis and angiolymphoid hyperplasia.

Histopathological examination showed that the lesion had occupied the whole dermis.

Neoplasic cells were irregularly arranged and the fibroblasts were spindle-like and round cells. Their cytoplasms were wide, eosinophilic, and scarcely vacuolated. There were extensively pleomorphic with prominent nuclei. Multilobular nuclei and giant cells were seen. Great numbers of mitotic figures were observed. Scattered lymphocytes were observed along with dilated vascular structures in between (**Figure 2, Figure 3**). Immunohistochemically, neoplasic cells stained diffusely and strongly positive by vimentin, whereas only focal positivity was found with CD-68 staining. Some of them showed positivity with S-100 protein. Pancytokeratin, EMA, HMB-45, actin, desmin and LCA were negative. The solitary lesion was diagnosed to be atypical fibroxanthoma with clinical, histopathological and immunohistochemical findings and the lesion was totally excised. Histopathological examination of the totally excised material revealed no tumor cells at the surgical margin. There was no local recurrence and no regional lymphadenopathy for one year. There was no pathology detected on thoracic and abdominal CT.

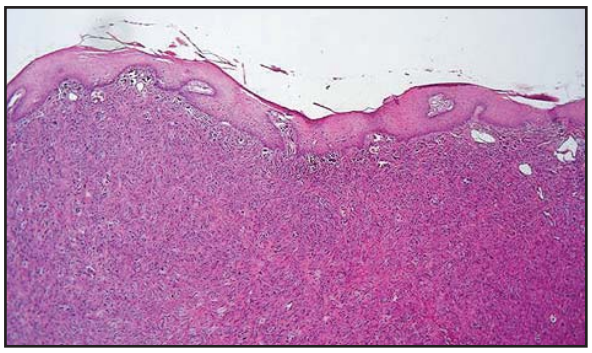

Figure 2. Tumor tissue filling the dermis underneath the squamous epithelium (HE x 100).

Discussion

Atypical fibroxanthoma has two different clinical forms. Solitary, ulcerated nodules (that usually grow in less than 6 months) of 1-2 cm in diameter and nodules seen on sun-damaged skin of the elderly is the most common form of the disease. A less common second form has been characterized on the body and extremities of young people [**2, 3, 4**]. AFX is seen twice more frequently in males compared to females [**5**].

AFX seems to occur predominantly on sun-damaged skin, with solar elastosis present in 99% of the patients. Reflecting this, the principle sites of occurrence are areas likely to be frequently exposed to sunlight. There was a particular predilection for the head and neck (91% of cases), especially the ears [**6**]. Our case's lesion was on the ear, suggesting that sun protection of the ears by long hair in females may be of importance.

In 1963, *Helwig* defined the exact features of the tumor in accordance with the malignant and xanthomatous appearance and described it as AFX [**7**]. In 1977, *Barr* reported the fibrohistiocytic origin of AFX [**8**]. Atypical fibroxanthoma is a pleomorphic neoplasm consisting of non-epithelial, non-melanocytic and spindle-like cells. When observed in detail, spindle cells, large fibroblastic, histiocytic cells, atypical multinuclear giant cells, osteoclast-like giant cells, lymphocytes and

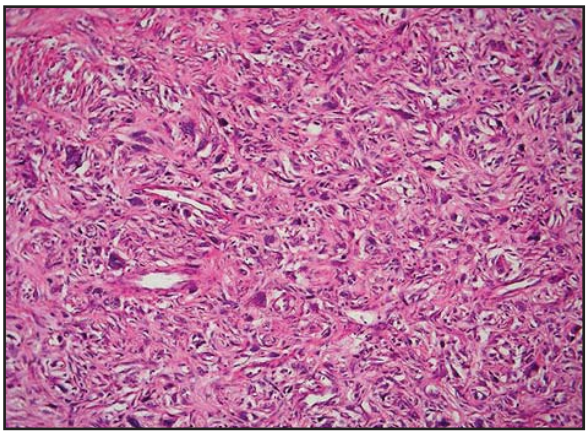

Figure 3. Spindle shaped and polyhedral epitheloid cells in the cellular tumor tissue and multilobulated tumor cells, showing prominent polymorphism in between (HE x 400).

ectasic vascular areas can be observed. The cells are randomly lined and show high mitotic activity, along with hyperchromatic and multilobulated nuclei. In particular, inflammatory cells are found more often at the tumor margin and ulcerated regions [**2, 4, 5, 9, 10**].

Light microscopy alone is insufficient in the differentiation of AFX lesions from the other cutanous spindle cell tumors, therefore immunohistochemical and ultrastructural investigations should be performed. It usually stains positive with CD68 and actin but it usually stains negative with desmin, S-100, HMB-45 or keratin markers. But keratin staining should be used at least once in every AFX-diagnosed patient in order to rule out squamous cell carcinoma. Jensen reported positive staining of 40 of 46 AFX lesions with procollagen-1 [**6, 11**].

Some primary and metastatic neoplasms can be similar to AFX, and malignant melanoma, dermatofibrosarcoma protuberance, malignant peripheral nerve sheath tumors, soft tissue sarcomas and pyogenic granuloma should be keep in mind in the differential diagnosis [**6, 9, 10, 12**]. Specific diagnosis is important because there seems to be a very low risk of recurrence or metastasis despite the frequently alarming histology [**6**].

Solar radiation is a predisposing factor in the etiopathogenesis of AFX. In recent molecular studies, there have been reports of p53 mutations in AFX cells. As in non-melanoma skin cancer, this condition is attributed to the p53 gene mutations caused by UV light and p53 immune reactivity has been found to be present in 7 of 10 lesions [**13**]. H-ras and K-ras gene mutations have been detected in 2 of 8 malignant fibrous histiocytomas. There were no H-ras, K-ras or N-ras mutations in 8 AFX patients in the absence of N-ras mutation [**14**]. Our case is in consistence with this theory with respect to the occurrence of the lesion on a sun-exposed area and at the same time, in the presence of other signs of actinic damage such as actinic keratosis and cutis rhomboidalis. However, the lesions in the non-sun exposed parts of the body cannot be explained by the UV light effect. Radiation therapy and local skin trauma are other possible factors in the etiology of AFX. Out of 642 kidney transplant patients, two malignant fibrous histiocytomas and one AFX cases were detected and immunosuppression was thought to play a role in occurrence of the tumor [**15, 16**].

Spontaneous regression rarely may occur but total excision of the lesion is the treatment of choice [**6**]. The prognosis is good following total excision of small and superficial lesions. Due to the reported recurrence rates of 5-10%, total excision of the tumor with a clean surgical margin is a must. Recurrence is related to the subcutaneous fat tissue invasion and incomplete or insufficient excision, and it usually occurs 1-2 years after the excision. Metastasis is rare and metastatic dissemination occurs to the regional lymph nodes, parathyroid, and especially to the lung. Risk factors for metastatic invasion are: prominent cellular pleomorphism, high mitotic activity, deep invasion, vascular invasion, tumor necrosis and repetitive local recurrence [**5, 9, 17, 18**].

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