Pedunculated atypical fibroxanthomas of the face

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Abstract

Atypical fibroxanthomas are rare, superficial dermal tumors. Most cases are benign and only locally destructive with a low rate of metastasis. Lesions are most commonly found on sun-exposed sites of elderly light-skinned patients and present as asymptomatic nodules with irregular borders; ulcerations and friability are other key characteristics. Pedunculated lesions, however, are rarely described in the literature. We present two cases of atypical fibroxanthoma manifesting as exophytic, pedunculated lesions on the face: one in a 74-year-old man and the other in an 82-year-old woman. These tumors are very effectively treated by excision with Mohs micrographic surgery.

Keywords: atypical fibroxanthoma, surgical dermatology, Mohs micrographic surgery, oncology

Introduction

Atypical fibroxanthoma (AFX) is an uncommon atypical spindle cell neoplasm of fibrohistiocytic lineage. They are typically unlikely to metastasize, but have done so occasionally [1]. These tumors are commonly found on sun-exposed areas such as the head and neck, trunk, and upper limbs of elderly Caucasian patients [2,3]. This neoplasm presents most commonly as an asymptomatic, non-tender nodule with irregular borders that may ulcerate and bleed [2]. Once diagnosed, Mohs micrographic surgery is the treatment of choice with the lowest rates of recurrence. We report two instances of this AFX presenting as a friable, exophytic, pedunculated nodule on the face.

Case Synopsis

A 74-year-old man presented with a new, rapidly enlarging tumor on his right cheek that had begun as a small "pimple" two months prior. The lesion was asymptomatic, easily bled with minimal trauma, and sometimes expressed purulent fluid. The patient had no prior similar lesions but did have a history of squamous cell carcinoma in situ that had been successfully treated with 5-fluorouracil cream. Physical examination showed a 2cm exophytic friable red smooth plaque on a 5mm stalk on the right parotideomasseteric cheek (**Figure 1A)**. Tangential biopsy revealed an atypical spindle cell neoplasm with nuclear pleomorphism and abundant mitotic figures (**Figure 2**). Lesional cells were negative for pancytokeratin, p63, smooth muscle actin, desmin, Melan-A, S-100, and CD34 immunostains. With these findings, a diagnosis of atypical fibroxanthoma was made and patient underwent excision with Mohs micrographic surgery. At follow-up, the patient had healed well from his surgery without evidence of recurrence.

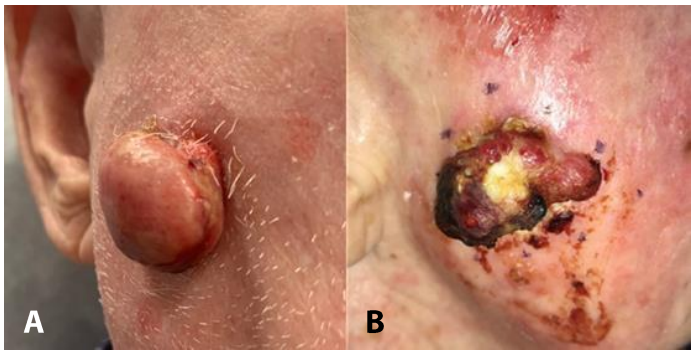

Figure 1. *Atypical fibroxanthoma presenting as an exophytic, friable pedunculated plaque on **A)** the right cheek of a 74-year-old man and **B)** the right cheek of an 82-year-old woman.*

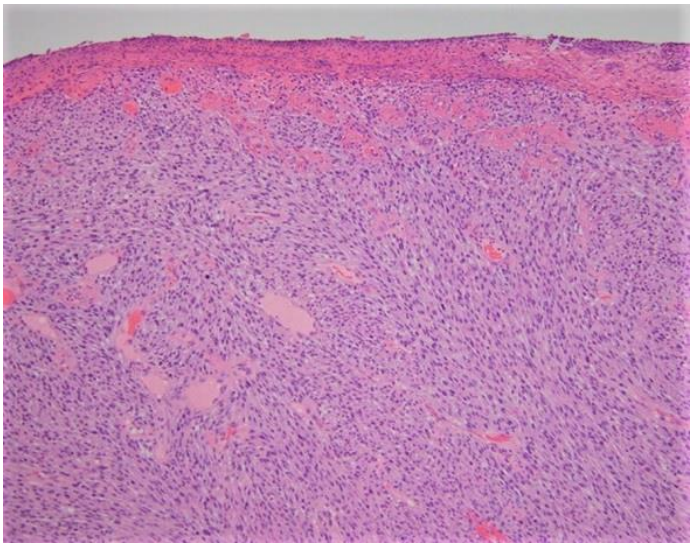

Figure 2. *Histopathology of the lesion in Figure 1A showed an atypical spindle cell neoplasm abutting the epidermis with nuclear pleomorphism and abundant mitotic figures. H&E, 10×.*

Similarly, an 82-year-old woman presented with a rapidly growing mass on her right cheek that had been present for one month (**Figure 1B)**. Her past history was significant for meningioma which was treated with radiation. She denied other similar skin lesions in the past. On physical exam, there was a 3.3×1.5cm red exophytic pedunculated friable plaque with a small stalk. Computed tomographic scan with contrast of the head and neck did not show invasion into the deep fat and there was no associated lymphadenopathy. Biopsy confirmed a diagnosis of AFX. Pleomorphic dermal sarcoma was unable to be completely ruled out. The patient declined Mohs micrographic surgery and was subsequently seen by a head and neck surgeon who excised the lesion. There has been no evidence of recurrence.

Case Discussion

Atypical fibroxanthoma was first described as a benign neoplasm of atypical spindle cells in the literature by Helwig and May in 1961 [1]. Risk factors for the development of AFX include photosensitivity, trauma, radiation therapy, as well as history of squamous cell carcinoma, basal cell carcinoma, and actinic keratosis [2]. Given its rarity and shared characteristics with other common conditions, the clinical differential diagnosis of AFX is broad and may include basal cell carcinoma, dermatofibrosarcoma protuberans, and lobular capillary hemangioma.

Histologically, most lesions are characterized by an abundance of spindle cells and mixed fibroblasts with atypia, nuclear enlargement, pleomorphism, and frequent mitoses [2,3]. Ionizing radiation is believed to be the driving factor in the pathogenesis of AFX. A case series of 140 patients found that 8.2% of patients with history of prior radiation therapy developed AFX in their lifetime [2]. The presence of pyrimidine dimers induced by radiation was also found to be in higher quantities in AFX when compared to other benign fibrohistiocytic tumors [4].

To our knowledge, the two cases described herein represent a novel presentation of AFX on the face as an exophytic, pedunculated friable lesion. A history of skin cancer and radiation therapy was present in these cases. However, in most instances, AFX typically presents as an enlarging dome-shaped or ulcerative nodule, without a peduncle. Although there have been a few previously published cases of pedunculated AFX, these have been limited to two ocular cases [5,6] and one auricular case [7]. Another instance of a pedunculated lesion was seen in a 71-old man as a collision tumor between amelanotic melanoma and AFX [8]. Undifferentiated pleomorphic sarcoma (UPS), the deeper malignant counterpart to AFX, is also rarely seen as a pedunculated lesion. On review of the literature, this type of presentation of UPS has been reported twice, once on the cheek and the other time on the scapula [9,10]. This demonstrates the rarity of pedunculated AFX and AFX-like lesions, as well as the need for histopathological analysis for definitive diagnosis.

Treatment of AFX is primarily surgical. Total margin control using Mohs micrographic surgery (MMS) appears to be most effective, with recurrence rates reported as low as 0% in a series of 59 patients; the majority of tumors were cleared after two stages [11]. Other options include wide local excision, electrodesiccation and curettage, and shave excision, but these are associated with increased risk of recurrence. Given a low likelihood of metastasis

and low recurrence rates with MMS, the prognosis of AFX is generally excellent [2].

Conclusion

Atypical fibroxanthoma is a low-grade atypical spindle cell fibrohistiocytic neoplasm most commonly found on sun-exposed skin. With early detection and treatment with MMS, recurrence is rare and prognosis is excellent. It is important for clinicians to be cognizant of AFX as it shares characteristics with many other skin cancers highlighting the need for biopsy and histopathological analysis. These cases highlight an unusual presentation of AFX as an exophytic pedunculated neoplasm on the face.

Potential conflicts of interest

The authors declare no conflicts of interests.

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