# BRIEF ARTICLES

## Atypical Fibroxanthoma of the External Ear: Case Report and Review of the Literature

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### ABSTRACT

Atypical fibroxanthoma (AFX) is a rare, dermal- based mesenchymal neoplasm. Clinically, these tumors are characterized by rapid, exophytic growth and epidermal ulceration. Despite striking clinical features and growth pattern, it is considered to be a tumor of low- to intermediate- malignant potential. We report a case of an 89 year old Caucasian male that had a 1 month history of a rapidly enlarging, pedunculated neoplasm on the scapha of his right ear. Histologic and immunohistochemical analysis of the lesion were consistent with atypical fibroxanthoma. After a biopsy, the patient underwent a complete resection with Mohs micrographic surgery and remains asymptomatic 6 months later. This 3.0 x 2.0 cm lesion emerged over a 4-5 week period, representing the most rapid growing AFX of the external ear reported in the literature.

### INTRODUCTION

Atypical fibroxanthoma (AFX) is a tumor first reported in 1961 to describe a dermal tumor of atypical spindle cells.[1-4] A retrospective study on 42,279 skin cancers treated using Mohs micrographic surgery revealed that .24% of these skin cancers were atypical fibroxanthomas, demonstrating the rarity of this neoplasm.[5] This tumor classically affects elderly Caucasian males and most commonly appears as an ulcerated papule or nodule less than 2.0 cm, though variations occur.[2,6] Immunohistochemical stains are required for accurate diagnosis, as AFX is a diagnosis of exclusion when ruling out more aggressive lesions. The standard of care for AFX of the head and neck is currently considered to be Mohs micrographic surgery with follow up at 6 month intervals.[2]

This case represents the most rapid growing atypical fibroxanthoma of the external ear documented in the existing literature, growing to 3.0 x 2.0 cm in 4-5 weeks. One case report of a 1.0 x 0.8 cm AFX that was excised after 2 weeks of growth, and this was the only AFX reported in the literature that could have reached similar size in 4-5 weeks.[7]



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## CASE REPORT

An 88 year old Caucasian male presented to our outpatient dermatology clinic with a 4-5 week history of a rapidly enlarging, slightly pedunculated mass on the right ear scapha (Figure 1). The lesion was asymptomatic and the patient denied local trauma or previous history of such a lesion. The lesion measured 3.0 x2.0 cm and was non-tender, pink, and friable. No regional lymphadenopathy was appreciated. After locally anesthetizing the site, the lesion was excised down to the perichondrium with half of the lesion sent to histology for permanent sections and the other half sectioned and analyzed with frozen section histology in our laboratory. Frozen sections obtained in our lab demonstrated a poorly differentiated spindle cell tumor with a large number of mitotic figures (Figure 2). The initial differential diagnosis included atypical fibroxanthoma, atypical sarcoma, and spindle cell variant melanoma. Positive margins were identified on the base of the lesion prompting Mohs surgical removal of the remaining lesion. A significant post-op defect remained after tumor removal, thus porcine xenograft was placed on the surgical site to facilitate the wound to heal by second intention (Figure 3).

Permanent section histology with immunohistochemical staining was faintly focally positive for smooth muscle actin and negative for: H-caldesmon, S-100, SOX 10, pan melanocytic cocktail (HMB-45, anti-Mart 1 and anti-tyrosinase), AE1/AE3, P63, Desmin, ERG, P63, CD31, and CD34, confirming the diagnosis of atypical fibroxanthoma. The patient had excellent healing results 3 weeks post-op and continues to be asymptomatic 6 months after removal.

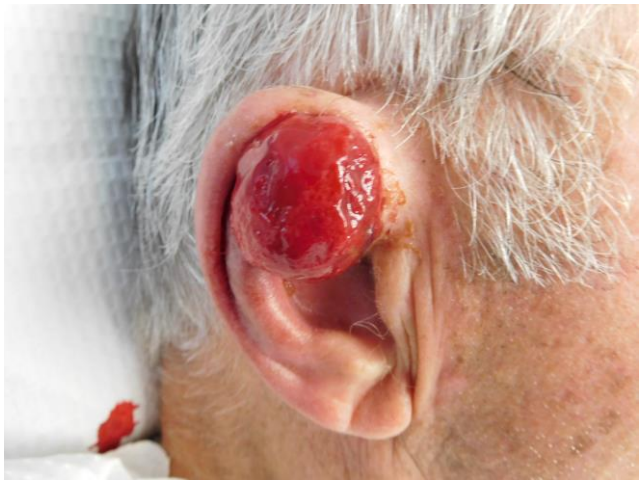

Figure 1: Atypical fibroxanthoam of the right ear

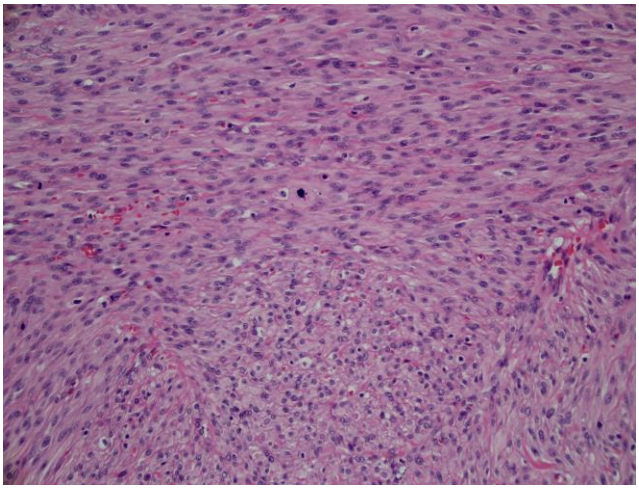

Figure 2: H&E staining revealing dermal proliferation of atypical spindle cells



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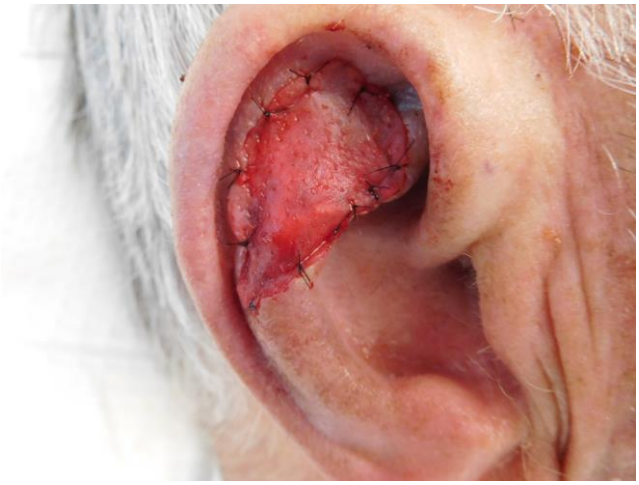

Figure 3: Reconstruction with porcine xenograft

## DISCUSSION

Atypical fibroxanthoma (AFX) is a tumor that is classically found in elderly Caucasian male patients, with most neoplasms occurring on chronically sun-damaged skin.[1-3,9] There are 2 distinct anatomic locations and demographic populations in which these tumors occur.[8] 78% of AFX occurs in the sun-exposed head and neck, with a median age of 69 among patients affected.[8] The remaining 22% of cases of AFX are present in the trunk and limbs, with a median age of 39.[8] AFX is at least twice as common in men than women.[2]

UV-induced DNA damage affecting p53 proteins are central to the pathogenesis of AFX.[1,2,10,11] These tumors have an accumulation of inactive p53 proteins on histology and are strongly correlated with sun-damaged skin.[1,2] Other risk factors for the development of AFX include X-ray exposure, xeroderma pigmentosa, and chronic immunosuppression (HIV, organ transplant).[2,10,11] The cell type that gives rise to AFX is under debate, with current sources suggesting an undifferentiated mesenchymal cell with features of fibroblastic, myofibroblastic, or histiocytic origin.[2,3,7]

The tumor may appear as a pink to red papule or nodule with superficial ulceration and bleeding.[10] Most AFX are less than 2.0 cm in diameter, with a 4 month median interval from onset to presentation.[2,9-11] The differential diagnosis includes dermatofibrosarcoma protuberans, malignant fibrous histiocytoma, spindle cell squamous carcinoma, angiosarcoma, Kaposi sarcoma, pyogenic granuloma, leiomyosarcoma, and spindle cell variant malignant melanoma.[1,9,10] Histologic exam is required for diagnosis.

H&E staining shows a well-defined dermal neoplasm with a proliferation of atypical spindle cells haphazardly arranged.[12] Though this tumor has a low risk of metastasis, histologic features associated with more aggressive cases of AFX behavior include vascular invasion, necrosis, and invasion into the subcutaneous fat.[2] A panel of immunohistochemical stains is usually required to rule out other potential neoplasms. No specific immunohistochemical markers identify AFX, but vimentin, CD10, CD68, and p53 are all likely to be positive in cases of AFX.[1-3,6,12] It is advisable to order pan cytokeratin stains, pan melanocytic cocktail (HMB-45, anti-Mart 1, anti-tyrosinase), S100, AE1/AE3, P63, smooth muscle actin, CD31, and CD34 to exclude other neoplasms.[1-3] While soft tissue sarcomas are graded based on histological appearance, this is not appropriate for AFX because the alarmingly abnormal features would suggest a high-grade tumor although it has intermediate malignant potential at most.[2]

Mohs micrographic surgery has been shown to be the standard of care in the treatment of AFX, replacing wide-margin excision.[2,11,12] Recurrence rates of AFX have been up to



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6% using Mohs surgical techniques in comparison with 16% recurrence associated with wide-margin excision.[2] AFX has an excellent prognosis, though there have been rare cases of metastatic disease.[1,2,6,10] Metastatic spread is so rare that it has been suggested that cases of metastatic AFX were misdiagnosed cases of undifferentiated pleomorphic sarcoma.[2] Despite low recurrence rates, follow up at 6 month intervals is recommended to check for recurrence, healing at the excision site, lymphadenopathy, and other skin cancers, as these patients have a history of aggressive sun damage.[2,10]

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