

Atypical Fibroxanthoma Arising Within Erosive Pustular Dermatitis of the Scalp



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PRACTICE POINTS

- Atypical fibroxanthoma predominantly occurs in older men on the head and neck.
- Erosive pustular dermatosis may be a benign entity, but if it does not resolve, continue to rebiopsy, as rare tumors may mimic this condition.

We describe a painful atypical fibroxanthoma (AFX) arising in a setting of erosive pustular dermatosis of the scalp. Complete excision was curative and also was associated with resolution of pain and clearance of the erosive pustular dermatosis of the scalp. We review the diagnosis and management of AFX and discuss the role of actinic damage in this process.

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Atypical fibroxanthoma (AFX) is a low-grade dermal malignancy comprised of atypical spindle cells.¹ Classified as a superficial fibrohistiocytic tumor with intermediate malignant potential, AFX has an incidence of approximately 0.24% worldwide.² The tumor appears mainly on the head and neck in sun-exposed areas but can occur less frequently on the trunk and limbs in non-sun-exposed areas. There is a 70% to 80% predominance in men aged 69 to 77 years, with lesions primarily occurring in sun-exposed areas of the head and neck.³ A median period of 4 months between time of onset and time of diagnosis has been previously established.⁴

When AFX does occur in non-sun-exposed areas, it tends to be in a younger patient population. Clinically, it presents as a rather nondescript, firm, erythematous papule or nodule less than 2 cm in diameter. Atypical fibroxanthoma most often presents asymptotically, but the

tumor may ulcerate and bleed, though pain and pruritus are uncommon.⁵ Findings are nonspecific, and the diagnosis must be confirmed with biopsy, as it can resemble other common dermatological lesions. The pathogenesis of AFX has been controversial. Two different studies looked at AFX using electron microscopy and concluded that the tumor most closely resembled a myofibroblast,^{6,7} which is consistent with current thinking today.

Atypical fibroxanthoma is believed to be associated with p53 mutation and is closely linked with exposure to UV radiation due to its predominance in sun-exposed areas. Other predisposing factors may include prior exposure to UV radiation, history of organ transplantation, immunosuppression, advanced age in men, and xeroderma pigmentosum. The differential diagnosis for AFX encompasses basal cell carcinoma, squamous cell carcinoma, Merkel cell carcinoma, adnexal tumor, and pyogenic granuloma.

Case Report

A 93-year-old man was referred to our clinic for treatment of erosive pustular dermatosis of the scalp with photodynamic therapy (PDT). He had a more than 20-year history of multiple skin lesions including basal cell carcinoma, squamous cell carcinoma, and actinic keratoses (AKs). For one year prior to the current presentation the patient had concerns of pustules, scaling, itching, and scabbing on the scalp. The patient admitted that the pruritus caused him to pick at the scabs on the scalp. He had previously been treated with lactic acid 12% neutralized with ammonium hydroxide, tacrolimus, and halobetasol, all to no avail.

On physical examination, the lesions appeared erosive with crusting and granulation tissue (Figure 1A). The presentation was consistent with erosive pustular

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dermatosis of the scalp. Biopsy revealed granulation tissue. The patient underwent PDT and prednisone treatment with improvement. Additional biopsies revealed AKs. His condition improved with 2 PDT sessions but never fully cleared. During the PDT sessions, the patient reported intense unilateral headaches without visual changes. The headaches were intermittent and not apparently related to the treatments. He was referred for a temporal artery biopsy and rebiopsy of the remaining lesion on the scalp. The temporal artery biopsy was negative. The lesion that remained was a large nodule on the vertex scalp, and biopsy revealed AFX.

Immunohistochemical marker studies for S-100 and cytokeratin were negative. Invasion into subcutaneous fat was encountered (Figure 2A). Highly atypical spindle cells and mitoses were present (Figure 2B). Neoplastic cells were noted adjacent to nerve (Figure 2C). Excision of the lesion was curative, and his symptoms of pain and erosive pustular dermatosis resolved weeks thereafter (Figure 1B). The area of erosive pustular dermatosis

was not excised, but symptoms resolved weeks following excision of the AFX.

Comment

Our case of AFX is unique due to the patient's atypical presentation of severe pain. Because AFX usually presents asymptotically, pain is an uncommon symptom. Based on the histologic findings in our case, we suspected that neural involvement of the tumor most likely explained the intense pain that our patient experienced.

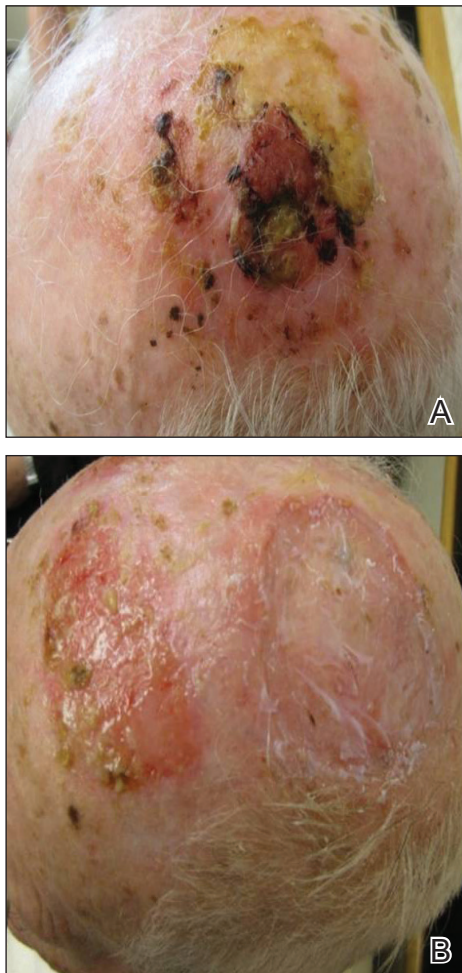


FIGURE 1. Atypical fibroxanthoma arising within erosive pustular dermatosis with evidence of erosion with crusting and granulation tissue before (A) and after excision of the lesion (B).

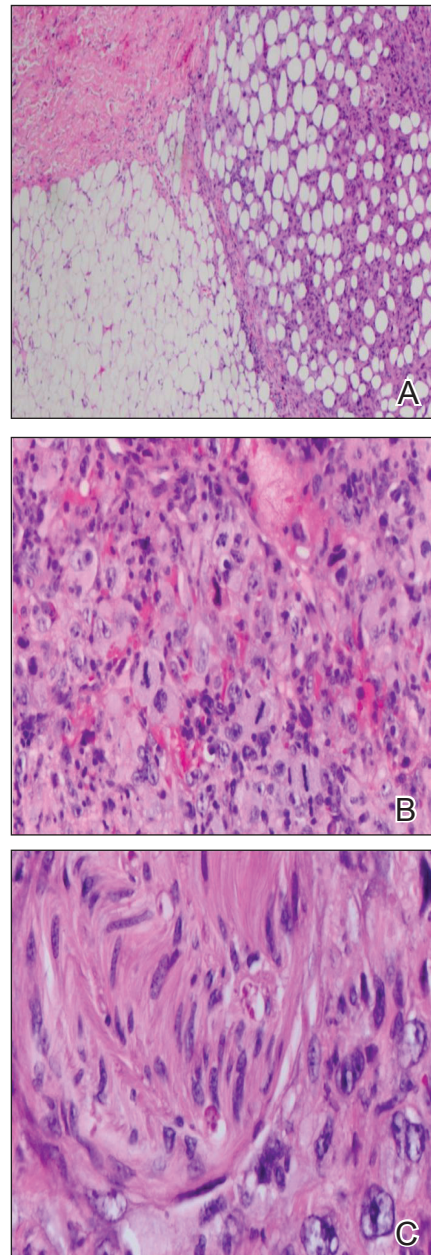


FIGURE 2. Atypical fibroxanthoma immunohistochemistry showed invasion into subcutaneous fat (A), highly atypical spindle cell neoplasm with mitoses (B), and neoplastic cells adjacent to neural tissue (C) (all H&E; original magnifications $\times 40$, $\times 200$, and $\times 400$, respectively).

The presence of erosive pustular dermatosis of the scalp also is interesting in our case. This elderly man had an extensive history of actinic damage and had reported pustules, scaling, itching, and scabbing of the scalp. It is possible that erosive pustular dermatosis was superimposed over the tumor and could have been the reason that multiple biopsies were needed to eventually arrive at a diagnosis. The coexistence of the 2 entities suggests that the chronic actinic damage played a role in the etiology of both.

Classification—There is a question regarding nomenclature when discussing AFX. Atypical fibroxanthoma has been referred to as a variant of undifferentiated pleomorphic sarcoma, which is a type of soft tissue sarcoma. Atypical fibroxanthoma can be referred to as undifferentiated pleomorphic sarcoma if it is more than 2 cm in diameter, if it involves the fascia or subcutaneous tissue, or if there is evidence of necrosis.³ Atypical fibroxanthoma generally is confined to the head and neck region and usually is less than 2 cm in diameter. In this patient, the presentation was consistent with AFX, as there was evidence of necrosis and invasion into the subcutaneous fat. The fact that the lesion also appeared on the scalp further supported the diagnosis of AFX.

Pathology—Biopsy of AFX typically reveals a spindle cell proliferation that usually arises in the setting of profound actinic damage. The epidermis may or may not be ulcerated, and in most cases, it is seen in close proximity to the overlying epidermis but not arising from it.⁸ Classic AFX is composed of highly atypical histiocytelike (epithelioid) cells admixed with pleomorphic spindle cells and giant cells, all showing frequent mitoses including atypical ones.⁹ Several histologic subtypes of AFX have been described, including clear cell, granular cell, pigmented cell, chondroid, osteoid, osteoclastic, and the most common spindle cell subtype.⁹ Features that indicate potential aggressive behavior include infiltration into the subcutaneous tissue, vascular invasion, and presence of necrosis. A diagnosis of AFX is made by exclusion of other malignant neoplasms with similar morphology, namely spindle cell squamous cell carcinoma, spindle cell melanoma, and leiomyosarcoma.⁹ As such, immunohistochemistry plays a critical role in distinguishing these lesions, as

they arise as part of the differential diagnosis. A panel of immunohistochemical stains is helpful for diagnosis and commonly includes but is not limited to S-100, Melan-A, smooth muscle actin, desmin, and cytokeratin.

Sampling error is an inherent flaw in any biopsy specimen. The eventual diagnosis of AFX in our case supports the argument for multiple biopsies of an unknown lesion, seeing as the affected area was interpreted as both granulation tissue and AK prior to the eventual diagnosis. Repeat biopsies, especially if a lesion is nonhealing, often can help clinicians arrive at a definitive diagnosis.

Treatment—Different treatment options have been used to manage AFX. Mohs micrographic surgery is most often used because of its tissue-sparing potential, often giving the most cosmetically appealing result. Wide local excision is another surgical technique utilized, generally with fixed margins of at least 1 cm.¹⁰ Radiation at the tumor site is used as a treatment method but most often during cases of reoccurrence. Cryotherapy as well as electrodesiccation and curettage are possible treatment options but are not the standard of care.

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