

Atypical Fibroxanthoma in a 115-Year-Old Patient

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Atypical fibroxanthoma (AFX) is a rare, low-grade malignant mesenchymal neoplasm of the dermis. It arises on sun-damaged skin in primarily older male patients as a solitary ulcerated nodule on the head and neck. Clinically, it grows rapidly without any pain or local invasion, and metastasis is rarely seen. Its appearance may be similar to that of other skin cancers, meaning that histopathology is necessary to confirm the diagnosis. In spite of its highly malignant histopathological features, it is widely considered as benign if strict criteria for diagnosis are followed to distinguish it from pleomorphic dermal sarcoma, which is currently considered a different entity, although both neoplasms were previously grouped together as the same pathological diagnosis [1,2]. Herein, we present a case of AFX that initially appeared malignant on the nose of a 115-year-old man. The aim of this report is to highlight the strict criteria involved in diagnosing AFX and to discuss the differential diagnosis of this benign lesion in contradistinction to the former definition as well as histopathological diagnosis, which might be of interest to clinicians.

A 115-year-old man presented to our outpatient clinic with an elevated, ulcerated, reddish, and shiny nodule on the right lateral side of the nose. The nodule was 3 cm × 3 cm in diameter (Fig. 1). The lesion was asymptomatic and had grown within one year, with no history of local trauma. No other special findings were detected, and total excision was



Fig. 1.

A 115-year-old man presented with an elevated, ulcerated, reddish, and shiny nodule on the right lateral side of the nose.

performed. A full-thickness skin graft harvested from the supraclavicular area was used to repair the skin defect. A histopathological examination of the biopsy indicated partial ulceration in the epidermis, invasion of the tumoral cells into the adipose tissue, and lymphocytic infiltration in the tumor base.

Additionally, the tumor was made up of atypical spindle cells, mixed with pleomorphic cells that had lobulated hyperchromatic nuclei. Numerous mitotic figures and spotty necrosis were detected (Fig. 2). On immunohistochemical staining (Fig. 3), the tumor cells were positive for CD 68, vimentin, focal epithelial membrane antigen (EMA), and negative for S100 protein, desmin, pan-cytokeratin (CK), smooth muscle actin (SMA), myogenin, CKS/6, melan-A, human melanoma black (HMB) 45, and CD34. Spindle cell squamous cell carcinoma was excluded because of the negative results for pan-CK, p63, and CK 5/6; malignant melanoma was excluded by the negative results for S100, melan-A, and HMB 45; smooth muscle-origin tumors were excluded by the negative results for SMA and desmin; skeletal muscle tumors were excluded by the negative results for myogenin; and neuronal tumors were excluded by the negative results for S100. The patient was diagnosed with AFX. Local recurrence was not observed in a six-month follow-up examination.

AFX is a pleomorphic spindle cell neoplasm of the

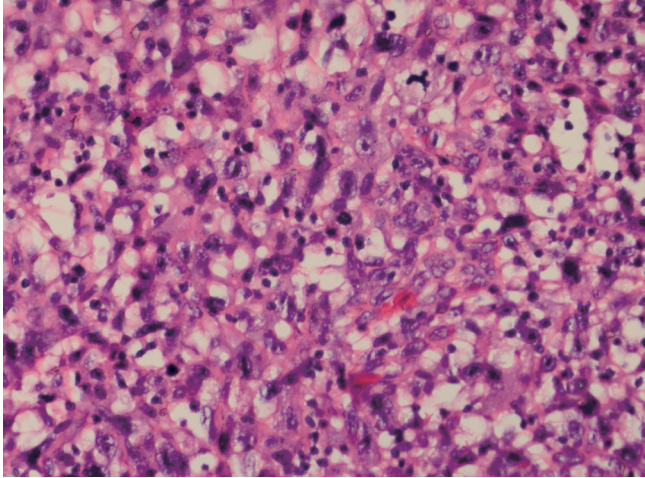


Fig. 2. The tumor cells were spindle-shaped or round, exhibiting pleomorphism and numerous mitotic figures (H&E, $\times 100$).

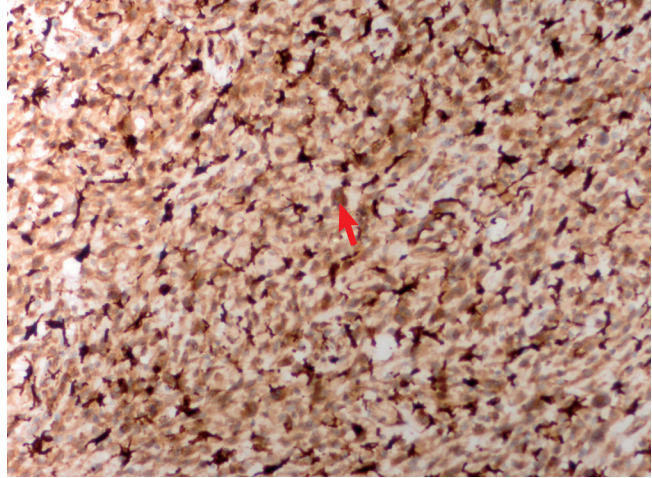


Fig. 3. The tumor cells showed strong positive immunostaining with CD68, as shown by the red arrow ($\times 200$).

dermis. It has a very low risk of metastasis or recurrence. Accordingly, it has been classified as a benign lesion despite its malignant histological features [3].

AFX arises on sun-damaged skin of the head and neck, and it predominantly affects elderly (> 70-year-old) male patients. The typical presentation of AFX is a rapidly growing, exophytic, ulcerated, and erythematous nodule, measuring < 2 cm in diameter. Pain and pruritus are uncommon.

Histologically, it presents as an unencapsulated tumor composed of large, multinucleated, spindle-shaped cells arranged in a haphazard fashion [2]. Squamous cell carcinoma, basal cell carcinoma, and keratoacanthoma, which are the most common skin cancers of the head and neck region, are epithelial cell-origin neoplasms and therefore must be distinguished from dermal-origin neoplasms. A diagnosis of AFX requires that the tumor be confined to the dermis with no epidermal connection, evidence of squamous or melanocytic differentiation, or vasoformative elements. A diagnosis of AFX also requires the absence of positive immunohistochemical staining for cytokeratins, S100, desmin, or CD34 in order to differentiate this tumor from other cancer types, including malignant melanoma, squamous cell carcinoma, and other non-melanocytic spindle cell tumors, such as leiomyosarcoma, rhabdomyosarcoma, angiosarcoma, liposarcoma, and dermatofibrosarcoma protuberans [2]. Focal positivity for EMA may also be seen. As stated by Tchernev et al. [4], AFX is a diagnosis of exclusion.

Historically, AFX was considered to be a superficial variant of malignant fibrous histiocytoma (MFH) [5]. However, MFH is now accepted as a more generalized term for sarcomatous neoplasms of the subcutaneous tissue [1]. Additionally, according to some authors, AFX was considered a superficial variant of undifferentiated pleomorphic sarcoma [5]. However, in recent years, it has been reported that findings of deep subcutaneous tissue invasion, tumor necrosis, lymphovascular invasion, or perineural infiltration are not compatible with a diagnosis of AFX, and these tumors are therefore better regarded as pleomorphic dermal sarcomas [1]. If these criteria are followed, AFX shows an entirely benign clinical course after complete excision. As mentioned, complete excision is generally curative, as in our case.

AFX generally has a good prognosis, with a low rate of recurrence, even so local recurrence has been described for lesions larger than 1.5–2.0 cm, as well as a low rate of lymph node metastasis [5]. Surgery is the only treatment option. Complete excision with safety margins is recommended as a treatment strategy.

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A Rare Case of Ecthyma Gangrenosum Caused by *Proteus vulgaris* and *Candida albicans* in a Patient with Castleman Disease

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Ecthyma gangrenosum (EG) is an ulcerative pyoderma of the skin that extends into the dermis and subdermal tissue with necrotic changes, and is generally described as a round, deep, punched-out ulceration with a central black eschar surrounded by an erythematous halo [1]. Development of EG is most commonly associated with *Pseudomonas aeruginosa* and is often seen in immunocompromised patients. However, cases of EG caused by other bacterial, viral, and fungal pathogens have been reported in the literature, albeit rarely. Prompt surgical debridement with appropriate antibiotic therapy is crucial for lowering the mortality rate in EG patients [1]. We describe an interesting case of EG caused by *Proteus vulgaris* (*P. vulgaris*) and *Candida albicans* (*C. albicans*) in a patient with multicentric Castleman

disease (MCD), which involves hyperactivation of the immune system and multiple organ system dysfunction. Our patient showed extensive lesions involving necrotic ulcerative changes on the lower abdomen and on the right upper arm. To the best of our knowledge, this was a rare case of EG associated with *P. vulgaris* and *C. albicans*.

A 57-year-old woman presented to the emergency department with multiple skin lesions involving her trunk and upper extremities. Six months previously, the patient had developed a perianal fistula and a colostomy was performed. The patient noticed an erythematous skin rash on her right arm that spread to her trunk and gradually progressed into necrotic bullae. Her past history indicated that she had been diagnosed with MCD three years previously, was taking immunosuppressive agents with oral steroids, and had no history of taking antibiotics. The patient had a mild temperature of 38.0°C, and the laboratory findings showed leukopenia with thrombocytopenia. A skin examination revealed a deep punched-out ulceration on the abdomen near the colostomy site and a large, palm-sized eschar formation with an erythematous halo on her right arm (Figs. 1, 2). Under suspicion of EG, the patient received cefepime to treat the most likely cause, *Pseudomonas aeruginosa* infection. Superficial wound swabs and tissue for Gram staining and culture were obtained before antibiotic



Fig. 1.
A large palm-sized eschar formation with erythematous halo changes on the patient's right arm.