Atypical fibroxanthoma of the cheek—case report with dermatoscopy and dermatopathology

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ABSTRACT
We present a case report of an atypical fibroxanthoma on the cheek of a 73-year-old man. Clinical, dermoscopic and dermatopathologic images are presented.

Introduction
Atypical fibroxanthoma is an uncommon tumor of fibrous tissue, a spindle cell neoplasm. It has locally aggressive behavior and a tendency to recur after surgery. However its metastatic potential is low [1]. It is most often found on the head and neck on sun damaged skin in elderly patients. Clinically it presents as a solitary nodule often with ulceration which can grow rapidly [2,3].

Case report
A 73-year-old man presented to a primary care skin cancer clinic in Melbourne, Australia for a routine skin cancer examination. He was concerned about a deep red, slightly domed lesion on his right cheek. This appeared some six weeks earlier. There was a long history of recreational sun exposure. There was a past history of multiple non-melanoma skin cancers, both basal cell carcinoma and squamous cell carcinoma. He had received several courses of 5-fluorouracil cream field therapy to multiple actinic keratoses of the forehead, temples and nose in the last 10 years.

A whole body skin examination was undertaken with the aid of a Heine Delta 20 non-polarizing dermatoscope (Heine Optotechnik, Herrshing, Germany). Digital clinical and dermoscopic images were taken with a Medicam 800 Fotofinder non-polarizing camera (Fotofinder Systems GmbH, Aichner, Birnbach, Germany) the dermatoscopy images being at 20x magnification.

There was severe actinic damage of the scalp, temples and nose plus less severe actinic damage to the upper trunk and distal limbs with multiple actinic keratoses and solar lentigines. The lesion of concern was located on the right mid cheek and measured 15 mm x 10 mm in diameter (Figure 1).
It was non-pigmented and was composed of two separate parts, a larger deep red nodule inferiorly continuous with a white nodule superiorly.

Dermatoscopically (Figure 2) there was a red structureless area inferiorly and a large well demarcated white clod superiorly plus a polymorphous vascular pattern comprising dot and linear vessels. Based on the polymorphous vascular pattern, a preoperative diagnosis of a malignant skin tumor including amelanotic melanoma, undifferentiated squamous cell carcinoma, Merkel cell carcinoma or other malignant uncommon adnexal tumor was suspected. An excisional biopsy was performed and the specimen was submitted for assessment by a specialist dermatopathologist.

Examination of the histological sections (Figures 3-6) revealed sun damaged skin with a hypercellular proliferation of atypical spindle cells forming irregular fascicles and sheets, extending down to the subcutis. There were a few erosions and a polymorphous vascular pattern comprising dot and linear vessels. Based on the polymorphous vascular pattern, a preoperative diagnosis of a malignant skin tumor including amelanotic melanoma, undifferentiated squamous cell carcinoma, Merkel cell carcinoma or other malignant uncommon adnexal tumor was suspected. An excisional biopsy was performed and the specimen was submitted for assessment by a specialist dermatopathologist.

Examination of the histological sections (Figures 3-6) revealed sun damaged skin with a hypercellular proliferation of atypical spindle cells in the dermis. This proliferation abutted the undersurface of the epidermis but did not appear to be continuous with it. The cells were arranged in whorled nests as well as in sheets. Prominent mitotic activity was seen with a mitotic rate of approximately 2 per square millimeter.
Bizarre multinucleated giant cells were present. The lesion extended through the full thickness of the dermis into the subcutis to a depth of approximately 2.5 mm (approximate due to fragmentation). Immunohistochemical stains were performed. These were negative for S100, and cytokeratin AE1/AE3 and high molecular weight keratin, and positive for the histiocytic marker CD68 (KP-1), favoring the diagnosis of atypical fibroxanthoma. It was initially thought this lesion might be a pleomorphic dermal sarcoma, a rare and more aggressive variant of atypical fibroxanthoma [4]. However, the criteria of perineural and lymphovascular invasion were absent. In the end it was felt the lesion was more in keeping with an atypical fibroxanthoma with inflammation.

Conclusion

Until recent years there has been little published literature about the dermatoscopy of atypical fibroxanthoma.

In 2009 Bugatti et al described the dermatoscopic patterns of three cases of atypical fibroxanthoma and concluded that “atypical fibroxanthoma may be added to the list of slightly pigmented, reddish, malignant cutaneous tumors such as squamous cell carcinoma, Merkel cell carcinoma, amelanotic/hypomelanotic melanoma and eccrine porocarcinoma, displaying prominent and chaotic dermatoscopic neoangiogenetic features in more advanced stages of proliferation” [3].

In 2013 Lallas et al reported on the dermoscopic patterns of five atypical fibroxanthomas which were “typified by reddish and whitish areas in combination with a polymorphous vascular pattern consisting of various combinations of linear, dotted, hairpin and highly tortuous vessel irregularly distributed over the surface of the lesion” [5].

These descriptions fit in with the dermatoscopic appearance of the lesion we present. Any growing nodular lesion on sun damaged skin in older patients should be completely excised and submitted for specialist dermatopathological examination especially when a benign lesion cannot be confidently excluded on dermatoscopy. Dermal nevus and sebaceous hyperplasia would be the most common benign raised lesions on the face in older patients. The lesion we present did not have the typical yellow clods and crown pattern blood vessels of sebaceous hyperplasia. Neither did it have the typical brown clods and curvilinear blood vessel patterns of dermal nevus.

Application of the “EFG rule,” which recommends excision of any skin lesion with the clinical features of elevation, firmness and growth would have also ensured this lesion was removed and subjected to histopathological examination [6,7].

Atypical fibroxanthoma is currently considered an uncommon tumor. However, such uncommon tumors will present more often as the world population increases in age and has increased access to modern medicine. Dermatoscopy is a relatively new diagnostic tool. The authors feel it is important to publish such dermatoscopic images as ours to as wide an audience as possible to aid clinical diagnosis in future.

References