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Atypical fibroxanthoma and malignant fibrous histiocytoma☆

Aaron H.J. Withers [a], Nicholas D.L. Brougham [a,b], Rachel M. Barber [c], Swee T. Tan [a,b,d,*]

[a] Wellington Regional Plastic, Maxillofacial & Burns Unit, Hutt Hospital, Wellington, New Zealand
[b] Wellington School of Medicine & Health Sciences, University of Otago, Wellington, New Zealand
[c] Department of Pathology, Hutt Hospital, Wellington, New Zealand
[d] Gillies McIndoe Research Institute, Wellington, New Zealand

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Treatment

Summary *Background:* Atypical fibroxanthoma (AFX) and malignant fibrous histiocytoma (MFH) are soft-tissue tumours with variable aggressiveness. There is considerable debate about the relationship between these lesions, as histological and immunochemical differentiation is difficult.
Methods: Current opinions and evidence for diagnostic differences between AFX and MFH were reviewed. Consecutive cases of AFX and MFH were identified from our non-melanoma skin cancer (NMSC) database 1996—2007 for the Central Region of New Zealand.
Results: Of the 50 411 NMSC lesions excised surgically from 26 138 patients, there were 101 AFX and 15 MFH cases. Three MFH cases were originally diagnosed as AFX. AFX and MFH share similar patient demographics, size and location and histological and immunohistochemical features. Most diagnostic biopsies of AFX were not followed by formal excision. Incomplete excision occurred in a large proportion of patients with AFX, which often did not proceed to re-excision, resulting in local recurrence. Cases of MFH generally underwent definitive treatment including re-excision if incompletely excised, and postoperative adjuvant radiotherapy.
Conclusions: The failure to treat AFX adequately may have resulted from the lack of appreciation of its aggressiveness. Contrary to the literature, we found few clinical differences between AFX and MFH. AFX and MFH also share similar histologic features and there are no immunohistochemical markers that reliably distinguish them. AFX is best considered a distinct entity with MFH, now reclassified as an undifferentiated pleomorphic sarcoma.

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* Corresponding author. Wellington Regional Plastic, Maxillofacial & Burns Unit, Hutt Hospital, Private Bag 31-907, High Street, Lower Hutt, New Zealand. Tel.: +64 4 5872506; fax: +64 4 5872510.
E-mail address: swee.tan@huttvalleydhb.org.nz (S.T. Tan).

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Atypical fibroxanthoma (AFX) and malignant fibrous histiocytoma (MFH) belong to a group of fibrohistiocytic tumours that are considered to have a variable aggressiveness.[1] In 1961, Helwig first coined the term 'atypical fibroxanthoma' to describe an atypical dermal spindle-cell tumour that exhibited a benign course.[1] AFX is thought to be related to actinic damage and ultraviolet (UV)-induced mutations in the p53 gene have been implicated in the proliferation of dermal fibroblasts.[2]

MFH was first described in 1961[3] and then classified by O'Brien and Stout in 1964[4] as a distinct histological type of soft-tissue sarcoma showing a pleomorphic phenotype and a storiform pattern derived from histiocytes. It has been regarded a distinct entity since the 1980s, and is considered to be the most common sarcoma in adult life. In 1992, Fletcher suggested that, historically, MFH consisted of a collection of poorly differentiated malignant mesenchymal neoplasms, the majority of which can now be categorised into correct entities, based on modern immunochemistry and electron microscopy.[5] The term 'undifferentiated pleomorphic sarcoma' has been proposed for the remaining small group.[6] The cause of MFH is unknown and it does not appear to be associated with sun exposure.[1]

As a regional service, our unit has been regularly referred cases of AFX and MFH. Anecdotally, many cases were not referred or formally excised following a diagnostic biopsy in the primary and secondary health settings and these patients subsequently presented with locally advanced tumours. This study was conducted to verify our concerns that these lesions might not have been adequately managed.

Materials and methods

A literature search using Medline (1996–2010) was undertaken and recent pathological textbooks were reviewed to determine current opinions and diagnostic differences between AFX and MFH (Table 1).

In accordance to the protocol approved by Central Regional Ethics Committee, cases of AFX and MFH were identified from our non-melanoma skin cancer (NMSC) database 1996–2007 for the Central Region of New Zealand with a population of approximately 824 800.[7] This database included all cases treated by a number of clinical disciplines and original histology reported by public and private laboratories, across the region. The patient demographics were recorded and the histology reports were reviewed for the size and anatomic location of the lesion, treating clinical discipline, excision margins, any further surgical procedure(s) performed following diagnostic biopsy or incomplete excision, recurrence and time to recurrence.

Table 1 Clinical, aetiological, histological and immunohistochemical features of AXF and MFH in published literature.

Features	AFX	MFH
Clinical		
Age of onset	7th decade	5–7th decade
Gender	Unknown	Male > Female
Location		
Head & neck	75%	10%
Trunk & limbs	25%	–
Limbs	–	70–75%
Size of lesion	<2 cm	5–10 cm
Metastasis	No	Yes
Aetiological		
Related to *UV*	Yes	No
Histological		
Spindle or round cell	Yes	Yes
Pleomorphism	Present	Marked
Atypical mitotic figures	Yes	Yes
Necrosis	Uncommon	Common
Pattern	Fasicular	Storiform
Perineural/vascular invasion	No	Yes
Extension	Superficial subcutis	Deep subcutis dermal, fascial & muscle extension
Immunohistochemical		
S100, HMB45	Negative	Negative
CD34	Negative	Negative
Cytokeratin	Negative	Negative
Smooth muscle actin, vimentin	Positive	Positive
CD68, α1-antichymotrypsin, α1-antitrypsin	Positive	Positive
CD10	Positive in high % of cases	Positive in almost all cases
CD74	Weakly positive	Strongly positive
CD99	Positive in 35–75% of cases	Negative