Lipidized Fibrous Histiocytoma Presenting as a Keloid-Like Lesion

-A Case Report

Feng-Ling Lin  Mei-Chin Ho  Shu-Lin Hu

A 46-year-old man presented with a gradually growing nodule on the left dorsal foot, which had been present for over 10 years. In the beginning, the lesion was treated as a keloid. Because of poor response to treatment, biopsy was arranged. Sections showed a dermal tumor composed of numerous foamy cells, hyalinized wiry collagen, spindle cells and entrapped collagen. The foamy cells were positive for KP1(CD68) and negative for S-100 protein and HMB-45. The histological and immunohistochemical results were consistent with the diagnosis of lipidized fibrous histiocytoma. Lipidized fibrous histiocytoma is a rare and underrecognized variant of fibrous histiocytoma. The atypical presentations and histological features make the diagnosis difficult. It should be differentiated from conditions with clear cell or xanthomatized cells. We hope our observation will lead a better understanding of this rare disease for clinicians and avoid misdiagnosis and mismanagement. (Dermatol Sinica 26: 252-257, 2008)

Key words: Dermatofibroma, Fibrous histiocytoma, Lipidized

INTRODUCTION

Fibrous histiocytoma (FH) is the most common fibrohistiocytic skin tumor which is a group of mesenchymal lesions with fibroblastic, myofibroblastic and histiocytic differentiation. Typical FHs occur most commonly in the middle aged women on the lower extremities. FH is usually a single, red-brown, round and firm nodule measured from 0.5 cm to 1 cm in diameter. When squeezing the lesion, a “dimpling” sign is produced. There was controversy on its true nature: a neoplasm or a reactive process? Most people agreed with the concept that it is a local reaction precipitated by local trauma or insect bite.1 Numerous clinicopathologic variants of FHs have been reported (Table 1). The different variants may present atypical patterns, which may cause substantial diagnostic problems and mismanagements.2 We reported a case of an unusual histological variant of FH presenting as a keloid-like lesion clinically.

CASE REPORT

An otherwise well 46-year-old man presented with a slowly enlarging tumor on his left dorsal foot for ten years. He denied any traumatic history or any previous skin lesions on the same location. Physical examination revealed a 2 x 2 cm, dark-red, firm nodule on his left dorsal foot (Fig. 1). The tentative diagnosis was a keloid. The patient underwent one session of cryotherapy and five sessions of intralesional triamcinolone.
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acetonide (10 mg/ml) injection. However the response to the treatment was unsatisfactory. An incisional skin biopsy was performed.

The histology showed a dermal tumor which abutted the epidermis and extended into the subcutis. There were epidermal hyperplasia with hyperkeratosis, focal ulceration and crust formation (Fig. 2). The tumor was composed of proliferating fibrous and histiocytoid cells (Fig. 3). Vascular proliferation and sclerotic collagen bundles were noted.

Higher magnification revealed a densely infiltration of numerous foam cells. These cells were oval to polygonal in shape with large hypochromatic nuclei, prominent nucleoli and abundant vacuolated cytoplasm (Fig. 4A, 4B). Conspicuous hemosiderin deposition in the cytoplasm of foam cells were observed. There are binucleated or multinucleated giant cells but no Touton giant cells. The foam cells showed diffuse cytoplasmic staining for CD68 (KP1) (Fig. 4C). The tumor was negative for periodic acid-Schiff stain, S-100 protein, and HMB-45. Lipidized FH was diagnosed and an en bloc excision was performed. The patient was followed up for one year and remained well without recurrence.

DISCUSSION

Lipidized FH was identified in 1994 as an exophytic yellowish nodule around the ankle that composed predominantly of foam cells. It is a rare and underrecognized variant of fibrous histiocytoma. Few case series were reported so far. Iwata reviewed 22 cases of lipidized FH. The mean age of these patients was 50 years. The ratio of male to female was 2.7. All but two cases of unknown site presented on the lower limbs. It is characterized that it usually arise, around the ankle, as the old name “ankle-type FH” implied. The size of the tumor arranged from 1 cm to 8 cm with a median of 2.5 cm in diameter. Fifteen lesions were exophytic in which eight of them were polypoid or pedunculat-
as fibrohistiocytic lesions clinically. It was concluded that in comparison to ordinary FH, lipidized FH occurred most frequently around the ankle, in the older males and in large size. However, Wagamon proposed that there was no difference in the age and distribution of the lesion between lipidized FH and ordinary FH.

Histologically, lipidized FH is well circumscribed and located in the deep dermis. The lesion is composed predominantly of foam cells (>75%), which are distributed between hyalinized, wiry collagen bundles. Touton-type giant cell are frequently present. Some lesions may also combine features of other FH variants.

In our case, combined histological features of hemosiderotic FH such as proliferation of small vessels, extravasation of erythrocytes and hemosiderin deposition are present. The vascular component and hemosiderin deposition contributed the dark-red appearance instead of a typical yellowish nodule.

The diagnosis of lipidized FH is established by histological features of numerous foam cells and at least focal features of FH. The immunohistochemical staining of lipidized FH is similar to ordinary FH but it failed to allow an unequivocal diagnosis. Early phases of FHs are positive for macrophages markers (such as KiM1p, PGM1, KP1) and factor XIIIa. FHs show negative staining for S-100 protein, HMB-45, and cytokeratin.

The immunohistochemical profile in our case, i.e., with reactivity for KP1 (CD68) and negative staining for S-100 protein and HMB-45, is supportive of a diagnosis of FH.

The pathogenesis of lipidized FH is still unclear. Wagamon studied the relationship between serum cholesterol and lipidized FH and found that it does not appear to be related to hyperlipidemia.

The differential diagnosis of lipidized FH included conditions or tumors with char-
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acters of vacuolated cells such as clear cell or xanthomatized cells (Table 2). Clear cell FH is an extremely rare variant of FH. They usually located on the lower extremities but the morphology was variable. It was composed mainly of clear cells (> 90%). The immunohistochemical staining is similar with lipidized FH but part of clear cells may show PAS positive. Cutaneous balloon cell dermatofibroma is a fibrous histiocytoma with balloon cell change, which was considered to represent a degenerative phenomenon. It is characterized by a gradual transition of balloon cell and clear cell into deeper spindle cell and lack of Grenz zone. Clear cell sarcoma (malignant melanoma of soft parts) is a rare malignant soft tissue tumor. It typically involves the aponeurosis and tendon sheath in lower legs in young adults. Microscopically, the tumor is composed of infiltrating nests of spindle cells with clear cytoplasm and vesicular nuclei. Clear cell sarcoma is positive for S-100 protein and HMB-45. Balloon cell melanoma is a rare variant of malignant melanoma. Such tumors have variable numbers of large, round to polygonal cells with abundant and clear cytoplasm containing fine melanin granules. Immunohistochemically, these tumors showed positive staining for S-100 protein. In non-Langerhans cell histiocytosis (such as juvenile xanthogranuloma or papular xanthoma), they show mixtures of various types of mononuclear histiocytes (xanthomatized, spindle-shaped, scalloped, oncocytic) and Touton giant cell microscopically. They are positive for macrophage markers, which sometimes are difficult to differentiate from FH. Clinically it mainly occurs on the upper part of the body and seldom happens on the extremities. Various other xanthomas (such as eruptive, tuberous xanthomas or xanthelasma) are characterized by xanthomatized histiocytes and variable degree of Touton giant cells. They are yellowish disseminated lesions with underlying increased

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Fibrous histiocytoma (FH), dermatofibrosarcoma protuberans (DFSP).
plasma lipid levels.\textsuperscript{11}

The clinical presentations of non-ordinary FH is variable, which leads to mis-diagnosis easily. We hope that our experience will make dermatologists be aware of this condition. We suggest that early arrangement of biopsy leads to a correct diagnosis and appropriate management.

REFERENCES

臨床上表現似蟹足腫之脂肪性纖維組織細胞瘤
-病例報告

林鳳玲  何美瑾  胡淑玲
國泰綜合醫院皮膚科

一位46歲男性從十年前開始於左足背上出現一個逐漸長大之結節，一開始被當作蟹足腫給予治療，因為對治療的反應不好，故安排了切片檢查，病理顯示一個由大量泡沫狀細胞，堅硬而透明之膠原纖維，梭狀細胞，膠原纖維束所構成的真皮腫瘤，免疫組織化學染色顯示KP1(CD68)陽性，S-100 protein 與HMB45陰性，綜合病理與免疫組織化學染色結果，診斷為脂肪性纖維組織細胞瘤。脂肪性纖維組織細胞瘤是纖維組織細胞瘤之罕見且不易診斷之變異型，它非典型的表現與病理特徵常造成診斷上的困難，診斷必須排除其他伴隨有透明細胞或黃色瘤細胞之皮膚疾病。希望此病例報告可以讓臨床醫師更了解脂肪性纖維組織細胞瘤，以避免誤診與不必要之處置。（中華皮誌：26: 252-257, 2008）