

CASE REPORT

Subcutaneous Dermatofibroma

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Dermatofibroma (DF) is usually confined to the dermis and the overlying epidermis is usually hyperplastic. Although DF with deep subcutaneous extension is commonly encountered, purely subcutaneous DF is uncommon. In this review, we describe a case of a 41-year-old male patient who presented with a painless, subcutaneous, hard papule on the left thigh. After the skin had been incised the lesion was totally removed, and histopathology revealed a subcutaneous dermatofibroma. (**Ann Dermatol 23(2) 254~257, 2011**)

-Keywords-

Dermatofibroma, Fibrous histiocytoma, Subcutaneous

INTRODUCTION

Dermatofibroma (DF) is perhaps the most common mesenchymal growth of the skin. It has a predilection for the lower legs of women in their early 20s. This tumor most frequently occurs in the dermis with or without extending into the subcutaneous tissue¹. The benign DF is usually confined to the dermis, whereas the occurrence of this lesion purely in the subcutaneous tissue has uncommonly been reported²⁻⁵. According to the WHO, a deep location is found for less than 1% of all benign fibrous histiocytomas⁵. Herein, we report an uncommon case of a purely subcutaneous DF.

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CASE REPORT

A 41-year-old male presented to our dermatology out-patient clinic, with an apparently asymptomatic palpable subcutaneous skin lesion on the left thigh, appearing 3 months prior to his visit to our clinic. There was no previous history of medication use or trauma and no family history of skin disease. The lesion was a hard, solitary, discrete, subcutaneous papule. The lesion had not been increasing in size over the last 3 months. The hard subcutaneous papule popped out when an incision for biopsy was made into the skin above the lesion. Histologic findings showed a well-circumscribed, subcutaneous papule composed of fibrohistiocytic proliferation (Fig. 1A). Storiform arrangement of spindle cells, trapping normal collagen bundles at the periphery of the lesion were consistent with subcutaneous DF (Fig. 1B). There was no nuclear pleomorphism or hyperchromasia (Fig. 1C). Immunohistochemical staining demonstrated positivity for factor XIIIa (polyclonal, dilution 1:200, Biogenex, Fremont, CA, USA) (Fig. 2A), and negativity for CD34 (monoclonal, clone QBEND 10 dilution 1:50, Dako, Carpinteria, CA, USA) (Fig. 2B), desmin (monoclonal, Clone D33 dilution 1:20, Dako) (Fig. 2C). CD68 (monoclonal, clonePG-M1 dilution 1:50, Dako) staining showed negative reactions in the spindle cells (Fig. 2D). Microscopic and immunohistochemical examinations confirmed the diagnosis of subcutaneous DF. The patient is asymptomatic with no evidence of recurrent disease at the 3-year follow-up.

DISCUSSION

Our case showed strong immunohistochemical positivity for factor XIIIa (polyclonal, dilution 1:200, Biogenex) and negativity for CD34 (monoclonal, clone QBEND 10 dilution 1:50, Dako), desmin (monoclonal, Clone D33 dilution 1:20, Dako) and CD68 (monoclonal, clonePG-M1

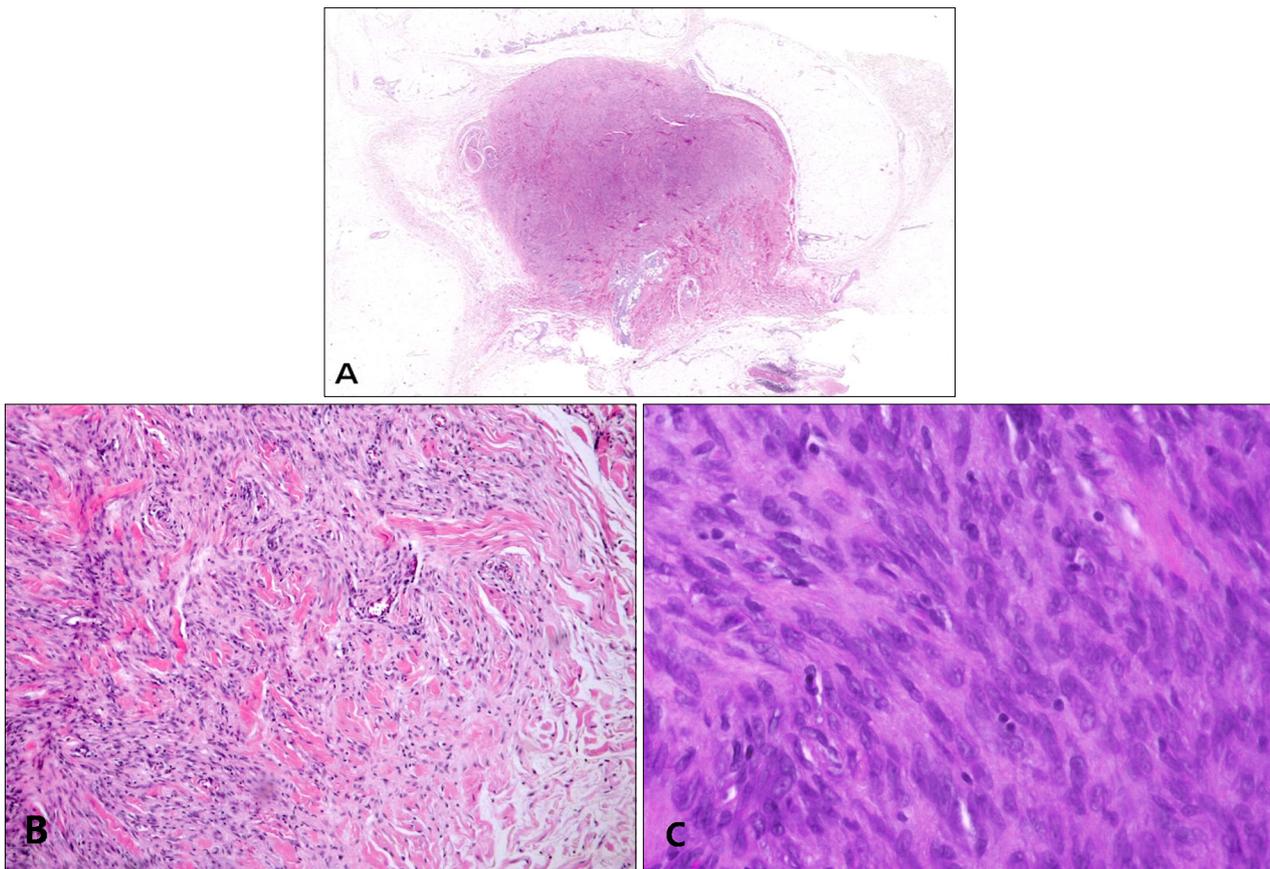


Fig. 1. Incisional biopsy specimen shows well-circumscribed, subcutaneous nodule (A: H&E, $\times 1$). The tumor cells are arranged in storiform pattern and are trapping normal collagen bundles at the periphery of the tumor nodule (B: H&E, $\times 100$). In higher magnification, tumor cells are cytologically bland and are spindle-shaped with variable amounts of pale eosinophilic, ill-defined cytoplasm. Nuclei are elongated and histiocyte-like with vesicular chromatin pattern. There is no nuclear pleomorphism or hyperchromasia (C: H&E, $\times 400$).

dilution 1:50, Dako) staining showed negative reactions in the spindle cells.

It is important to recognize the pure subcutaneous variant of DF because it may be confused with a variety of benign and malignant subcutaneous tumors⁶. Among the benign subcutaneous tumors, deep penetrating dermatofibroma (DPDF) or dermatofibroma extending into the subcutaneous tissue was described as a variant of dermatofibroma by Kamino and Jacobson⁷ in 1990. Kamino and Jacobson⁷ published the histologic criteria to distinguish DPDF from dermatofibrosarcoma protuberans (DFSP). Scanning magnification is the most helpful tool in distinguishing a regular, smooth, nodular or scalloped pattern of demarcation in DPDF from the irregular, lacelike/honeycomb or multilayered pattern of DFSP⁸. In previous reports, most cases of DPDF were positive for factor XIIIa and negative for CD34 and vice versa in cases of DFSP.

Malignant fibrohistiocytic neoplasms are composed of malignant pleomorphic sarcomatous cells, bizarre giant

cells, and frequent mitotic figures. Histological features, unfortunately, play a minor role in predicting the biologic behavior of these neoplasms⁹. The presence of certain atypical histological features, including necrosis, marked cellular and mitotic activity, does not correlate well with clinical recurrence.

As with benign fibrohistiocytic neoplasms, the diagnosis of malignant fibrohistiocytic neoplasms frequently relies upon electron microscopy and immunohistochemistry to differentiate it from other lesions. Like DF, DFSP occurs in the dermis and subcutis, but is more apt to show extensive subcutaneous involvement than benign DF. It is also characterized by a more uniform cellular population and lacks giant cells, inflammatory cells, and xanthomatous elements. Its fascicles, composed of slender attenuated cells, are longer and arranged in a distinct storiform pattern, unlike the short curlicue fascicles of DF. Its margins are infiltrative in contrast to the well-defined margins of DF^{10,11}. However, DFSP may be difficult to differentiate

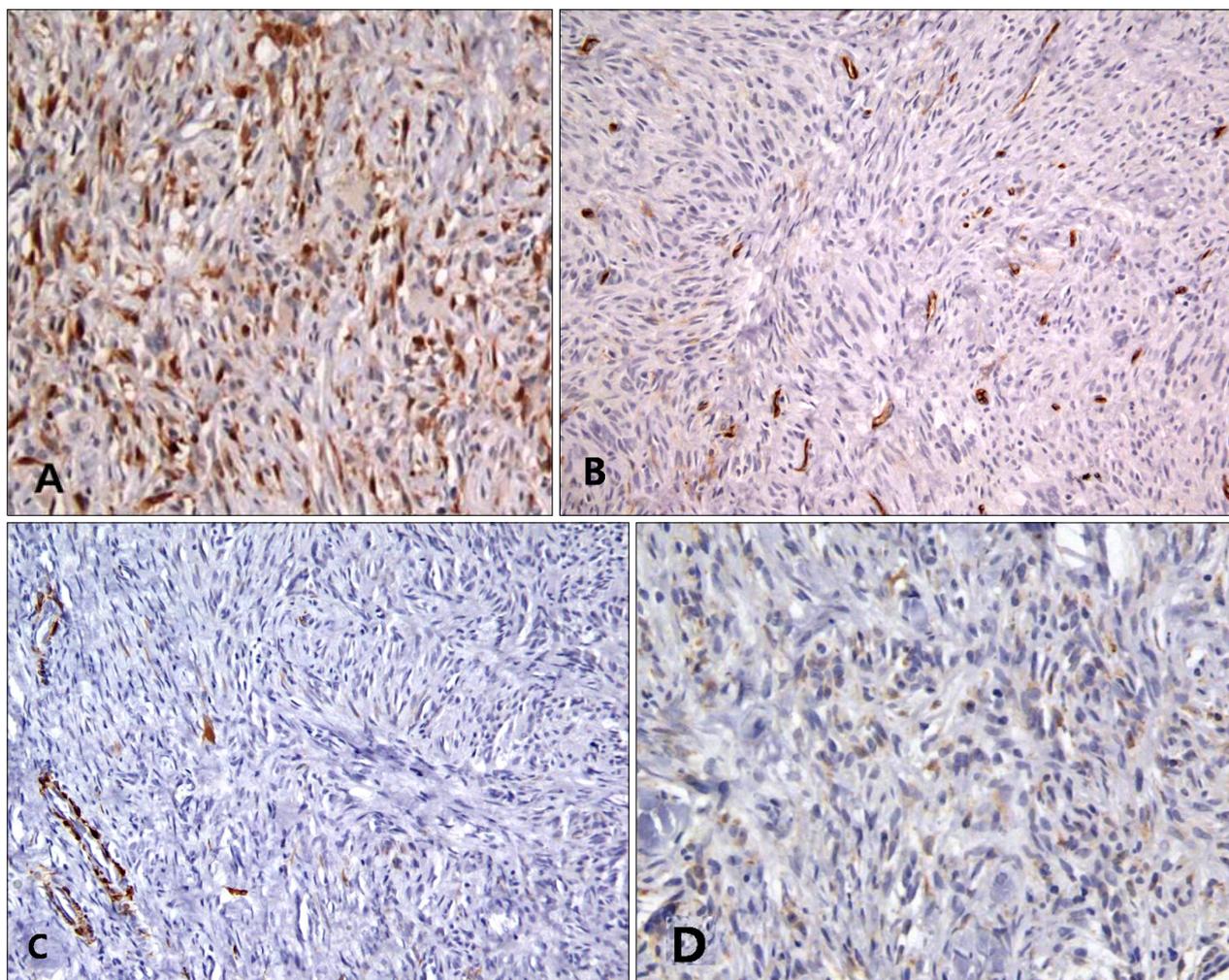


Fig. 2. On the immunohistochemical staining, there are positive reactions for Factor XIIIa and negative reaction for CD34 and desmin in the spindle cells (A: Factor XIIIa, $\times 200$, B: CD34, $\times 200$, C: desmin, $\times 200$). The CD68 staining shows negative reactions in the spindle cells (D: CD68, $\times 200$).

from DF. Immunostaining can help to reveal the distinct differences in the cellular composition of these tumors. DF contains a significant population of factor XIIIa-positive cells, although it has been debated whether these cells represent a population of neoplastic cells or a peculiar infiltrate that accompanies the tumor. In contrast, DFSP contains only scattered factor XIIIa-positive cells, but, in striking contrast to DF, it expresses CD34 in a significant portion of neoplastic cells^{12,13}. The combination of these two stains has proved to be highly reliable in distinguishing these two lesions, which often cause diagnostic problems, particularly when only a superficial biopsy specimen is available for assessment.

Usually, DF does not require treatment unless it is symptomatic. As per the literature, most subcutaneous DF's were treated by local excision without sacrificing structures that would cause major functional or cosmetic morbidity. Our

patient had not symptom but the lesion was removed when an incision for biopsy was made into the skin above the lesion.

DF almost pursues a benign course, with a recurrence rate of less than 2%, although a higher rate of recurrence has been reported for some variants, including the cellular, aneurysmal, and atypical (pseudosarcomatous) forms¹⁴. A higher rate of recurrence has been noted in the subcutaneous and deep types and in lesions located on the face, in which a recurrence rate of 15% has been reported^{6,15}. Also, a local recurrence rate of 19% (four of 21 cases) has been reported in 21 cases from the previous subcutaneous DF series². Accordingly, if the lesion has been proved to be subcutaneous DF with or without symptoms, it would seem reasonable to recommend that the lesion should be completely removed by local excision. In this paper, we have reported an uncommon case of

subcutaneous DF.

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