巨大細胞膠原纖維瘤

—硬化性纖維瘤一罕見變異型報告—

李美青　郭承統*　林靜怡　曾興隆

長庚醫院 皮膚科　病理科*

Giant Cell Collagenoma

—A Rare Variant of Sclerotic Fibroma : A Case Report—

Mei-Ching Lee　Tseng-Tong Kuo*　Jing-Yi Lin　Heng-Leong Chan

Sclerotic fibroma (Circumscribed storiform collagenoma) is a rare tumor. It may occur sporadically or in the context of Cowden's disease. A new rare variant of giant cell collagenoma was recently described. The new variant showed scattered multinucleated giant cells in an otherwise typical circumscribed storiform collagenoma. Herein, a case of this rare variant occurring in subcutis is reported. (Dermatol Sinica 20：115-118, 2002)

Key words: Sclerotic fibroma, Circumscribed storiform collagenoma, Giant cell collagenoma

硬化性纖維瘤(侷限性漩渦狀纖維瘤)是一種罕見之腫瘤。它可以單獨發生或是Cowden's疾病的一種皮膚表現。巨大細胞膠原纖維瘤是一種罕見的新變型，於最近被提出。在病理組織學上除了典型的侷限性漩渦狀纖維瘤的表現，亦可見到散在性的多核巨大細胞。在此，我們報告一例於皮下組織的罕見變異型。(中華皮誌 20：115-118, 2002)

INTRODUCTION

Circumscribed storiform collagenoma is a benign tumor characterized by a well-demarcated non-encapsulated dermal nodule composed of coarse hyalinized collagen bundles arranged in a storiform array with prominent clefts.[1] Giant cell variant of collagenoma was recently described by Rudolph *et al.*.[2] Microscopically, it has the similar features of a circumscribed storiform collagenoma, but in addition, scattered multinucleated giant cells are present between the fibrous tracts. We report a case of this rare variant and review the related literature.

CASE REPORT

A 34-year-old woman was seen at the dermatology clinic in October 2000 for an asymptomatic, slowly growing nodule on her right thigh for more than 10 years. No other family members had similar skin lesions. Her

From the Department of Dermatology, Chang Gung Memorial Hospital, Taipei

Accepted for publication: Octorber 8, 2001

Reprint requests: 李美青，台北市敦化北路199號長庚紀念醫院皮膚科辦公室　TEL: 02-27135211 ext. 3397　FAX: 02-27191623

medical history was not significant. Physical examination revealed a solitary 1.0x1.5cm subcutaneous firm nodule on her right thigh. There were no other papular lesions on the face, oral mucosa, or distal extremities. No signs of Cowden's disease were noted. Under the impression of a pilomatrixoma, the tumor was totally excised. Histopathologic examination showed a well-demarcated, non-encapsulated subcutaneous nodule composed of hypocellular hyalinized collagen bundles arrayed in a storiform pattern separated by prominent clefts with scattered multinucleated giant cells (Figs. 1 & 2 & 3). Immmunohistochemical study showed that both the fibroblasts and the giant cells were strongly reactive for vimentin (Fig. 4), but were negative for epithelial membrane antigen (EMA), smooth muscle actin, CD34, BCL-2, S-

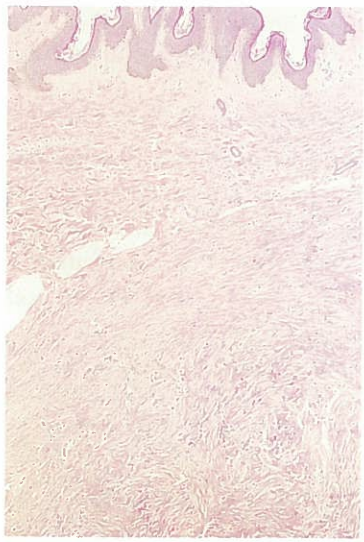

Fig. 1
A well demarcated nodule located in subcutis (H & E, 20x).

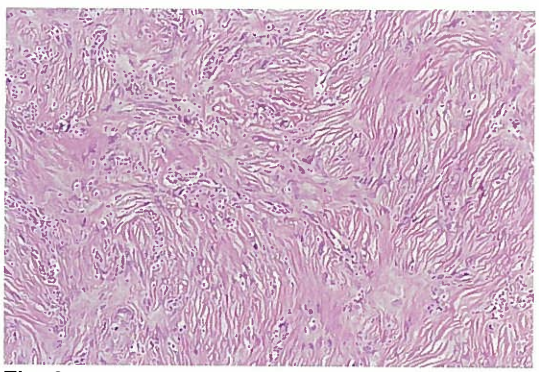

Fig. 2
Hypocellularized hyalinized collagen bundles arranged in a storiform pattern with prominent clefting. Note the presence of scattered hyperchromatic cells (H & E, 100x).

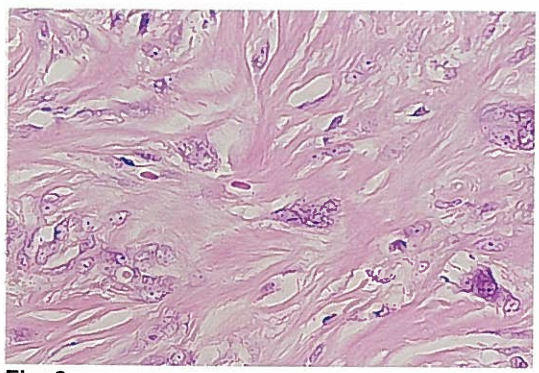

Fig. 3
Higher magnification to show multinucleated giant cells and pleomorphic cells between the collagen fibers (H & E, 400x).

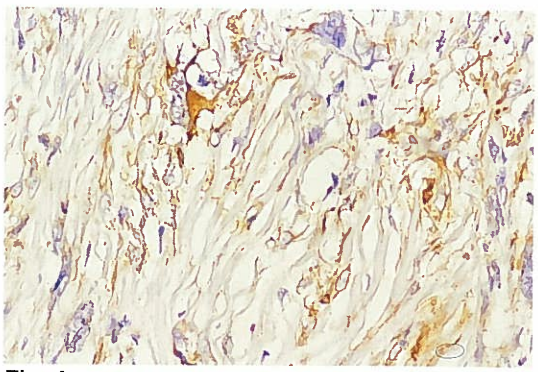

Fig. 4
Both the fibroblasts and the multinucleated giant cells are positive for vimentin (Immunostaining, 400x).

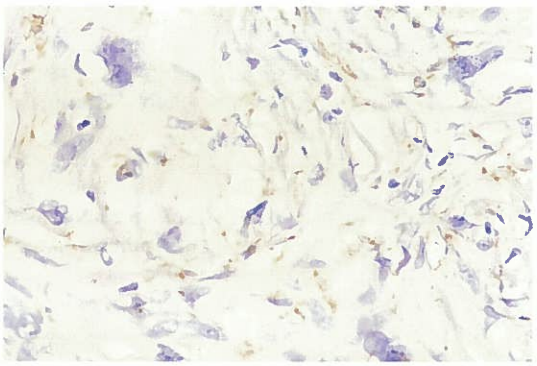

Fig. 5
The multinucleated giant cells are not reactive for KP-1. Only scattered cells are positive for KP-1 (Immunostaining, 400x).

100 protein, factor XIIIa, and macrophage markers such as KP1(Fig. 5), Ki-M1P, and Mac387. Only scattered dendritic cells were positive for factor XIIIa and macrophage markers. The diagnosis of a giant cell collagenoma was made based on the above distinctive histopathologic features and immunohistochemical findings.

DISCUSSION

Circumscribed storiform collagenoma,[1] also known as hypocellular fibroma or sclerotic fibroma, was first described in a patient with Cowden's disease by Weary *et al.* in 1972.[3] The hallmark of the tumor is a well-demarcated, non-encapsulated dermal nodule composed of hypocellular, hyalinized collagen bundles arrayed in an interweaving storiform pattern and separated by elongated mucin-containing clefs. Later, a sporadic form of the sclerotic fibroma of the skin was described by Rapini and Golitz in 1989.[4-7] In 1998, Rudolph *et al.*[2] reported five cases in which these tumors had the similar histopathological features of a circumscribed storiform collagenoma, but scattered multinucleated giant cells were present between the stromal fibrous tracts. Immunohistochemically, both the giant cells and the plump mononucleated cells were positive for vimentin, and negative for cytokeratin, smooth muscle actin, desmin, S-100 protein, CD34, factor XIIIa, and macrophage markers KP-1, Mac387, and Ki-M1P. Therefore, they were not histiocytes and believed to be fibroblastic in origin. The authors regarded it as a variant of circumscribed storiform collagenoma and termed the tumor a giant cell collagenoma. The present case showed similar histological and immunohistochemical features except that the tumor was located in subcutis. The multinucleated giant cells were also shown to be not histiocytic in origin.

The differential diagnosis of a giant cell collagenoma includes various fibrous proliferations with multinucleated cells, such as atypical benign fibrous histiocytoma, giant cell fibroblastoma, atypical fibroxanthoma, pleomorphic fibroma, solitary fibrous tumor, and sclerosing perineurioma. Atypical benign fibrous histiocytoma,[8] also referred to as dermatofibroma with monster cells,[9] occurs more frequently in women and is predominantly located on the extremities. It is characterized by a well-demarcated dermal nodule composed of plump fibroblasts and thick collagen fibers in a haphazard array. In addition, there are scattered multinucleated cells with hyperchromatic and large pleomorohic nuclei which are reactive with macrophage markers such as Ki-M1P. Giant cell fibroblastoma,[10,11] a rare benign tumor occurring almost exclusively in children, presents as a poorly-circumscribed and an infiltrating mass in deep reticular dermis and subcutaneous tissue. It consists of pleomorphic spindle cells and floret-type giant cells in a collagenous or myxoid stroma. Both the spindle cells and the giant cells strongly express CD34. Atypical fibroxanthoma[12] usually presents as a rapidly-growing, solitary nodule on the sun-exposed area. It is characterized by an exophytic, unencapsulated, densely cellular nodule composed of pleomorphic spindle cells and giant cells with high numbers of typical and atypical mitotic figures. Besides, hyalinized collagen bundles in a storiform pattern and prominent clefting are not found in atypical fibroxanthoma. Pleomorphic fibroma[13,14] usually presents as polypoid or high-dome shaped cutaneous fibrous lesion. It is characterized by a circumscribed dermal nodule composed of

coarse collagen bundles with sparse cellularity. Besides, scattered atypical cells and multinucleated cell are found throughout the fibrous stroma. However, its stroma does not exhibit prominent hyalinization, elongated clefts, and the uniform storifom array which is characteristic of a giant cell collagenoma. Solitary fibrous tumor,[15,16] a rare mesenchymal tumor, typically arises in the pleural cavity. It is also a spindle cell neoplasm characterized by a well-demarcated, non-encapsulated dermal nodule with diverse histologic patterns. It may display fascicular areas, storiform areas, and staghorn-like dilated vascular spaces. The main differential features of a solitary fibrous tumor is its positive CD34 staining. Sclerosing perineurioma,[17] a variant of perineurioma, has a predilection for the hands of young adults. It presents as a small, painless, dermal or subcutaneous mass that is composed of abundant thick hyalinized collagen arranged in whorls, cords, and trabeculae containing small epithelioid or short, plump spindle cells. The epithelioid cells and spindle cells of sclerosing perineurioma are positive for EMA, which are negative in giant cell collagenoma.

In conclusion, giant cell collagenoma has distinctive histologic and immunohistochemical features. Awareness of this rare variant will lead to a correct diagnosis.

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