Pachydermodactyly
-A Case Report

Yu-Lin Yeh   Horng-Shin Lin   Yu-Ping Hsiao   Hung-Chun Shih   Bor-Jeng Tsai  Jen-Hung Yang

Pachydermodactyly is a rare form of digital fibromatosis characterized clinically by asymptomatic soft tissue swelling affecting the skin of the lateral aspect of the proximal interphalangeal joints of the fingers in young boys around puberty. We herein report a 20-year-old healthy male with characteristic manifestations of pachydermodactyly. (Dermatol Sinica 26: 248-251, 2008)

Key words: Pachydermodactyly, Digital fibromatosis.

INTRODUCTION
Pachydermodactyly is a rare, benign form of digital fibromatosis characterized clinically by asymptomatic soft tissue swelling affecting the skin of the lateral aspect of the proximal interphalangeal joints of the fingers.1 It affects predominantly young boys around puberty, and only rarely affects girls.2 The exact etiology of pachydermodactyly is unknown; however, it is thought possibly relate to repetitive trauma, obsessive-compulsive disorder and Asperger’s syndrome (a pervasive developmental disorder that shares similar features of social impairment disorder, restricted interests, and repetitive behaviors with autistic disorder, usually manifests itself at preschool age).3 We herein report a 20-year-old healthy male with characteristic manifestations of pachydermodactyly.

CASE REPORT
A 20-year-old male student suffered from progressive asymptomatic bulbous swelling of proximal interphalangeal (PIP) joints over the fingers of bilateral hands for six to seven years. He is the only son and there are no known similar skin problems of his parents. He also denied having a history of repetitive trauma or having obsessive-compulsive behavior onto the affected skin. No arthralgia or limitation of movement of the PIP joints, and no morning stiffness was complained.

The skin examination revealed symmetrical swelling with light-brown close-set keratotic papules over the dorsal and lateral aspects of PIP joints of the 2nd to the 5th fingers of both hands (Fig. 1). X-ray radiographs showed soft tissue swelling over PIP joint of the fingers of bilateral hands without any bony or articular abnormalities (Fig. 2). Laboratory examinations including CBC/DC, ESR, GOT/GPT, BUN/creatinine, electrolytes were all within normal limits; in addition, ANA and rheumatoid factor were negative.

From the Department of Dermatology, Chung Shan Medical University Hospital
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Reprint requests: Bor-Jeng Tsai, Department of Dermatology, Chung Shan Medical University Hospital, No. 110, Sec. 1, Chien-Kuo North Road, Taichung 402, Taiwan, Republic of China.
TEL: 886-4-24739595 ext. 34517  0989-820-693  0926-287-779   FAX: 886-4-24738493   E-mail: tbjcsmu@csmu.edu.tw
The histopathologic examination revealed compact orthokeratosis and irregular epidermal acanthosis, discrete perivascular lymphocyte infiltration and slight proliferation of fibroblasts in the deep dermis. (Fig. 3, 4). Masson trichrome, alcian blue stainings revealed normal collagen and no deposits of mucin in the dermis.

**DISCUSSION**

Pachydermodactyly is a rare, benign form of digital fibromatosis characterized clinically by asymptomatic soft tissue swelling affecting the skin of the lateral aspect of the proximal interphalangeal joints of the fingers. It was first described by Bazax in 1973 and named by Verbov in 1975. It affects predominantly young boys around puberty, and only rarely affects girls. There have been approximately sixty cases reported worldwide. In Taiwan, there had been reported a 5-year-old Chinese boy with tuberous sclerosis and localized pachydermodactyly in 1993. A classification system had been proposed and classified into five types of pachydermodactyly by an Italian group (Table 1). Localized
pachydermodactyly refers to single digit involvement and transgrediens pachydermodactyly refers to examples of cutaneous thickening extending onto the metacarpophalangeal region.

Histopathologic examination of skin specimen demonstrated epidermal hyperplasia with compact orthokeratosis. Thickening of the dermis with an increase in collagen bundles, and a slight proliferation of fibroblast in the reticular dermis. Special stains with Masson trichrome observed increased number of collagen fibers; with Verhoeff-van Gieson, decreased number of elastic fibers that were thinned and elongated in the fibrous proliferation that surrounded adnexal structure. X-ray radiographs of hands shows soft tissue swelling of proximal interphalangeal joints without bony erosions or articular damage. All laboratory tests are unremarkable. Serologies are negative for antinuclear antibody and rheumatoid factor.

The etiology of pachydermodactyly remains unknown. Coexistent conditions reported with pachydermodactyly include tuberous sclerosis, atrophia maculosa varioliformis cutis, Dupuytren’s contractures, carpal tunnel syndrome, and Ehlers-Danlos syndrome.

The isolation of collagen from the involved sites has revealed large amounts of collagen type I, III, and V. This differs from the collagen profile of normal skin, which is composed of type I, III, IV, and VII. Ultrastructural studies revealed decreased diameter of collagen fibrils and increased number of fine-diameter collagen in the reticular dermis.

Pachydermodactyly has a benign evolution. Intralesional administration of triamcinolone hexacetonide or subcutaneous resection of the excess tissues can improve appearance, but topical steroid therapy is ineffective. Psychotherapy for those who have compulsive behavior is a good choice. Continuing observation was suggested.

**REFERENCES**

指(趾)部厚皮症
-病例報告

葉育霖 林鴻欣 蕭玉屏 施宏俊 蔡博正 楊仁宏
中山醫學大學附設醫院皮膚科

指（趾）部厚皮症是一個少見的指纖維瘤病，臨床上特徵是於手指近端指間關節兩側出現無症狀之軟組織腫脹，以發生於青春期前後男孩為主。在此我們報告一位20歲健康男性，表現符合指（趾）部厚皮症的診斷。（中華皮誌：26: 248-251, 2008）