

# PACHYDERMODACTYLY : REPORT OF TWO CASES

P. Mansouri., Z. Safaii Naraghi., H. Seirafi and M.R. Mortazavi

Dermatology Research Center, Imam Khomeini Hospital, Faculty of Medicine, Tehran University of Medical Sciences, Tehran, Iran

**Abstract** - Pachydermodactyly is a rare, benign, mostly asymptomatic digital fibromatosis, characterized by swelling on the back and sides of proximal phalanges and (or) proximal interphalangeal joints. It occurs predominantly in young males although a few women including a familial case were recently described. A history of repeated trauma is sometimes available, but the etiology remains unknown. We report two cases one of which had a history of repeated trauma (rubbing) to the involved area.

*Acta Medica Iranica* 37 (4): 232-236 ; 1999

**Key Words:** Pachydermodactyly, knuckle pad, digital fibromatosis, atrophica maculosa varioliformis cutis

## INTRODUCTION

Pachydermodactyly is a rare form of digital fibromatosis, characterized by bulbous, fusiform, asymptomatic soft tissue swelling on the back and sides of proximal phalanges and (or) proximal interphalangeal joints (PIP) of the index, middle and ring fingers. Involvement of little finger is rare, and the thumb is nearly always spared.

## CASE REPORTS

### Case 1

An 18-year-old man presented with a 4-year history of a tic-like habit of rubbing the PIP joints of a hand with the palm of the other during stressful states. There was also a history of clasping and rubbing the web area between left thumb and index finger.

The patient was a student of graphic arts, and had a history of repeated contact with color developer substances of photography. He occasionally played piano as a hobby. The only significant point in his past history was fracture of his left thumb in childhood, which resulted in its thickening and deformity. The family history was negative for similar conditions. He has conflicts with the other members of his family.

Physical examination revealed doughy, mobile, nontender swelling of medial and lateral sides of the PIP joints of the index, middle and ring fingers of the right hand and the ring finger of left hand (Fig. 1). The thumbs, toes, palms and soles were spared. The

overlying skin of several lesions showed mild lichenification but no erythema and scaling. The lichenification of the skin overlying the webspace between the left thumb and index finger was more remarkable. He had full range of motion in all his joints without tenderness, but had hyperlaxity of the PIP joint of the left ring finger. Complete examination showed round and linear facial depressions with various diameters localized on both cheeks. These lesions were preceded by repeated trauma. He had mild facial acne and androgenetic alopecia, too. General clinical examinations were otherwise unremarkable.

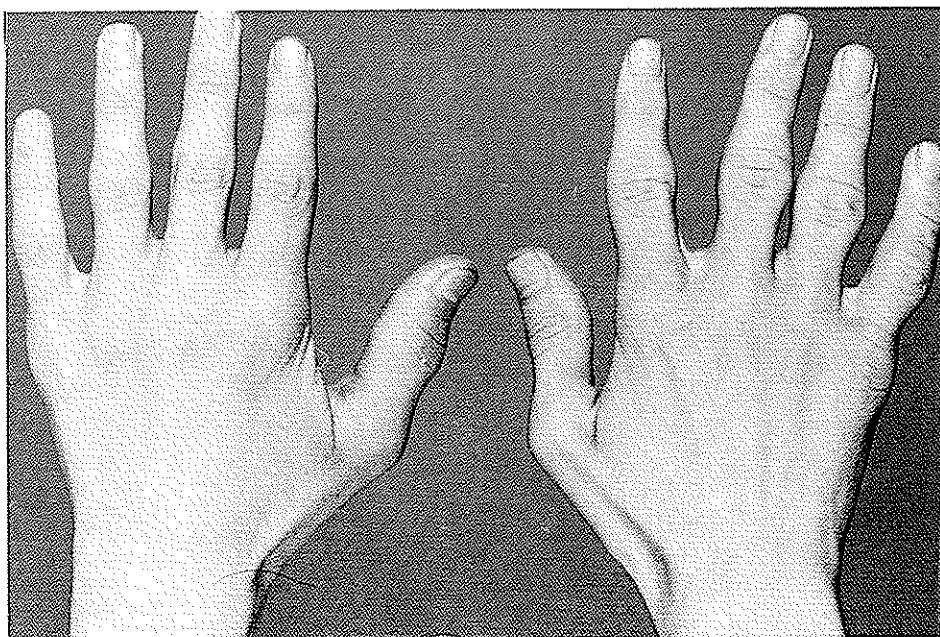
A roentgenogram of his hands (Fig. 2) showed soft tissue swelling with no bony or articular abnormalities. A skin biopsy of pachydermic lesion revealed hyperkeratosis, slight hypergranulosis and epidermal hyperplasia (Fig. 3). The thickened dermis showed deposition of collagen, mucin and proliferation of blood vessels (Fig. 4). There were also increased number of fibroblasts and ribs of thick, hyalinized abnormal collagen bundles encircling the adnexal structures and capillaries. No biopsy could be performed on the face.

### Case 2

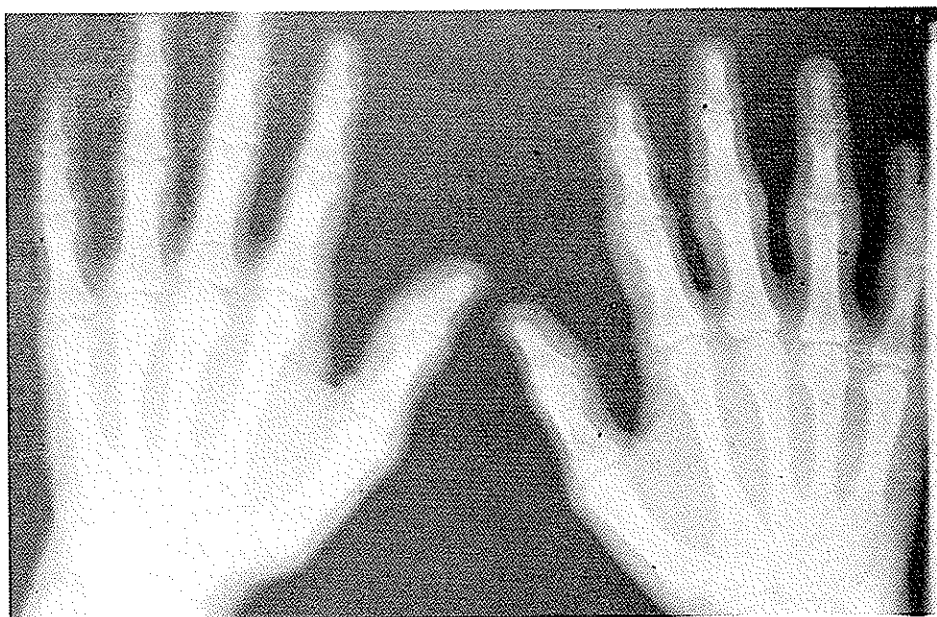
A 13-year-old female, high-school student had a 2-year history of asymptomatic swelling on the proximal phalanges and knuckles of the right hand. There was no history of tic or joint pain. She took part in gymnastic activities and drawing as a hobby. Past medical and family histories were not significant.

Physical examination revealed fusiform, nontender cutaneous thickening of proximal phalanges of the right hand. Involvement of the right middle and ring fingers was more prominent. Fingers of the left hand, the palms, soles and toes were spared. The skin on the back of the right hand, adjacent to the involved area was hyperkeratotic and slightly pigmented. The fingernails of both hands showed fine irregular pitting (Fig. 5). The PIP joints were not tender or limited in movement. The findings on general physical examinations were unremarkable. Roentgenograms of both hands revealed nothing more than "soft tissue swelling".

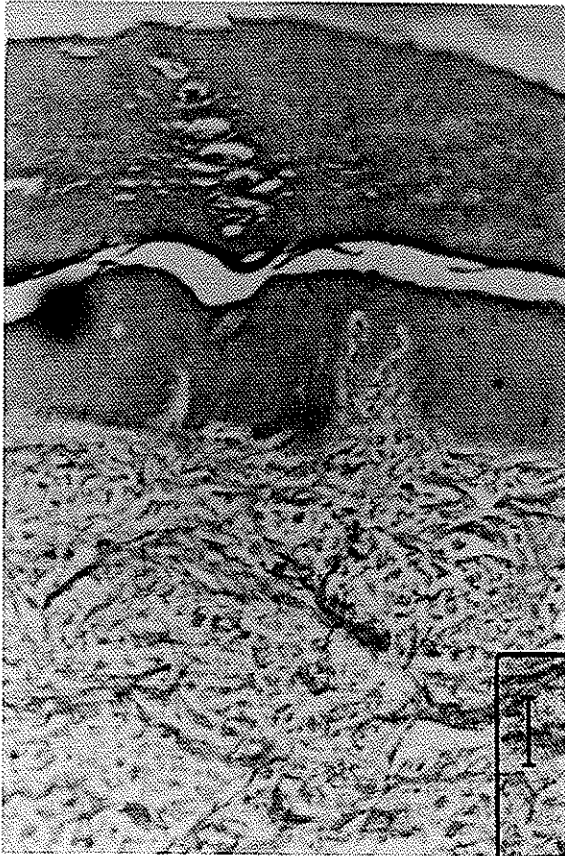
Skin biopsy of the pachydermic area showed hyperkeratosis, hypergranulosis and irregular acanthosis in the epidermis. The dermis showed fibroplasia and disorganization of connective tissue, increased number of fibroblasts and collagen bundles extending in different directions.



**Fig. 1.** Doughy, mobile, nontender swelling of medial and lateral sides of the PIP joints of the index, middle and ring fingers of right hand and the ring finger of left hand.



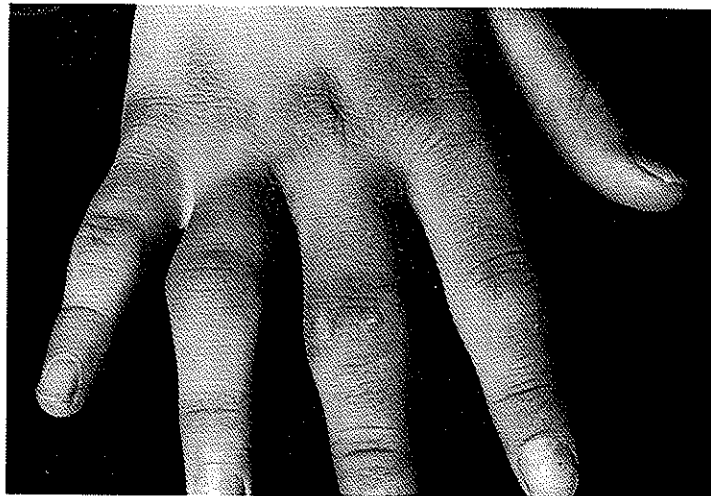
**Fig. 2.** Soft tissue swelling with no bony or articular abnormalities.



**Fig. 3.** Histopathology of the lesion showed hyperkeratosis and hyperplasia of epidermis, deposition of collagen and mucin and proliferation of blood vessels.



**Fig. 4.** Closer view of the dermis, showing increased collagen bundles and fibroblasts.



**Fig. 5.** Skin biopsy of pachydermic lesion revealed hyperkeratosis, slight hypergranulosis and epidermal hyperplasia

## DISCUSSION

Pachydermodactyly (PD) was first described by Bazex (1) in 1973 and later on by Verbov (2), who considered it as a form of atypical knuckle pad. More than 20 similar cases have been reported since then. All cases presented as soft tissue thickening on the back and sides of the proximal phalanges and (or) PIP joints of the hands. Nearly all of the reported cases, except four, were male. Two of the female patients presented sporadically (3,4) and two other cases, a mother and her daughter, were the first reported patients of familial pachydermodactyly (5). PD is mainly known as an acquired and sporadic disease, but a congenital variant has been reported as part of tuberous sclerosis complex (6). PD is usually asymptomatic, but, a painful variant has been reported, (4,7). As a rule, there is bilateral involvement of fingers in PD. Only a single case of localized PD, limited to one digit is mentioned in the literature (4). The index, middle and/or ring fingers are predominantly involved. Involvement of little finger is uncommon, and the thumb is nearly always spared. The toes, palms and soles are spared too. Tanguas et al (8) reported a case of PD transgrediens in which fibromatous thickening of the skin affected not only the finger joints, but also the dorsum of hands. One of the cases of familial PD reported by Russo and co-workers (5), showed extension of pachydermic lesions up to the sides of the palms.

The skin may show lichenification (3). There are no inflammatory signs, although mild erythema and scaling has been reported (9). No underlying bony or articular abnormalities are detectable in these patients. Histopathologic examination shows marked thickening of dermis by deposition of haphazardly arranged collagen fibers; sparse, elongated elastic bundles; occasional increased number of fibroblasts; and mucin deposition (3,4). Reichert and co-workers (10) showed an increase in collagen types III and V. Ultrastructural studies revealed increased number of fine-diameter collagen fibers in reticular dermis.

The differential diagnosis includes true knuckle pad, Garrod's pad, infantile digital fibromatosis, pachydermoperiostosis and psoriatic acropachydermodactyly. True knuckle pads are sporadic or autosomal dominant. Hard, circumscribed lesions on PIP or DIP joints (knuckles) with distinct histopathological features, differentiate them from PD (11). Gardos pads are mechanical swellings of the distal interphalangeal area especially in professional violinists (12). Differential diagnosis is made by distinct clinical or histological features.

PD has been reported in association with gynecomastia (7), carpal tunnel syndrome and Dupuytren's contracture (10). Callot and co-workers claimed the association of PD with "atrophia maculosa varioliformis cutis" (AMVC), without performing a facial biopsy of their reported case. AMVC (14, 15) is a rare idiopathic type of dermal atrophy, characterized by spontaneous linear and varioliform depressions on the cheeks, and nearly normal histologic findings of epidermis and dermis; i.e. a shallow depression of epidermis, a relatively normal dermis with slightly decreased elastic fibers and absence of fibrosis or inflammation. It seems obvious that a histopathologic examination is mandatory to differentiate AMVC from more common simple scars with dermal fibrosis (14), our case 1 also had linear and round facial depressions on both cheeks with a questionable history of trauma. No biopsy could be performed on the face.

The role of mechanical traumatism in the etiology of PD is controversial. Iraci and co-workers (16) and Lautenschlager and co-workers (17) reported cases of PD, who had sustained repeated trauma because of gymnastic activities and "tic-like" habits, respectively. Itin and Lautenschlager (18) argued that PD was a psychocutaneous disorder, and history of repeated mechanical traumatization was a pre-requisite for diagnosis. The kinds of repeated micro trauma suggested by these authors as the cause of PD are very common and could hardly explain all of the cases of such an uncommon disease (13). There are several reported cases of PD that had not had exogenous influences (18), and the cause of PD in them remains unknown.

There is no effective topical treatment for PD. Intralesional corticosteroids may result in reduction of swelling and help to improve the cosmetic appearance of lesions (7,18). Subcutaneous resection over the involved PIP joints is another alternative (19) but long-term follow-up data are not yet available. In the patient with positive history of tic-like habits, this obsessive-compulsive behavior has to be stopped. Discontinuation of this habit will lead to a marked amelioration in finger swelling (17,18). Psychotherapy may be more useful than surgery or injection of intralesional steroids in these patients (20). Other cases of pachydermodactyly had gymnastic activity and habitual tics. The role of drawing and graphic arts in both cases, in the development of pachydermodactyly remains obscure. It appears to be a distinct clinical entity of multifactorial etiology. It comprises of at least two varieties: one is trauma-associated and the other is "idiopathic". The diversity of associated conditions, the higher occurrence in males, and the reported familial cases all are problems that the "psychocutaneous hypothesis" can not explain.

## REFERENCES

1. Bazex A, Dupreo A, Teillard J. Pachydermic digital ... Bull Soc Fr Dermatol Syphiliger. 1973; 80: 455-8.
2. Verbor J. Pachydermodactyly, a variant of tru knuck up Arch. Dermatol 1975; 111: 524.
3. Draluck JC, Kopf AW, Hodak E. Pachydermodactyly, first report in a woman. J Am Acad Dermatol 1992; 27: 303-5.
4. Bardazzi F, Fanti PA et al: Localized pachydermodactyly in a woman. Acta Derm Venereol (Stockh) 1994; 74: 152-3.
5. Russo F, Rodriguez - Pichardo A. Familial pachydermodactyly. Acta. Derm. Venereol (Stockh). 1994; 74: 386-7.
6. Lo WL, WONG CK. Localized pachydermodactyly in tuberous sclerosis. Clin. Exp. Dermatol. 1993; 18: 146-7.
7. Curley RK, Hudson PM, Marsden RA. Pachydermodactyly: a rare from of digital fibromatosis - Clin Exp Dermatol 1991; 16: 121-.
8. Yanguas I, Goday JJ, Soloeta R. Pachydermodactyly transgrediens Acta Dermatol Venereol (Stockh) 1994; 74(3): 217-8.
9. Sola A, Vasquez Doval J, et al. Pachydermodactyly transgrediens. Int J Dermatol 1992; 31: 796-7.
10. Reichert CM, Costa J, Barsky SH, et al. Pachydermodactyly. Clin Orthop Relat Res 1985; 194: 252-7.
11. Meunier L, Pailler C, et al. Pachydermodactyly or acquired digital fibromatosis. Br J Dermatol 1994; 31(5): 744-6.
12. Rimmer S, Spielvogel RL. Dermatologic problems of musicians. J Am Acad Dermatol. 1990; 22: 657-63.
13. Callot V, Wechsler J, et al. Pachydermodactyly and atrophia maculosa varioformis cutis. Dermatology 1995; 190: 56-8.
14. Kolenik SA, Perez MI. Atrophia maculosa varioliformis cutis. J Am Acad Dermatol. 1994; 30: 837-40.
15. Marks VJ, Miller OF. Atrophia maculosa varioliformis cutis. Br J Dermatol 1986: 115: 105-9.
16. Iraci S, Luea Bianchi, Tomassoli M. Pachydermodactyly a case of an unusual type of reactive digital fibromatosis, Arch Dermatol 1993; 129: 247-8.
17. Lautenschlager S, Itin PH, et al. Pachydermodactyly reflecting obsessive compulsive behavior Arch Dermatol 1994; 130: 387.
18. Itin PH, Lautenschlager S. Pachydermodactyly a psychocutaneous disorder. Dermatology 1995; 190: 1-3.
19. Ffleter TB, Mrie C, Adams JP. Pachydermodactyly: a case report and discussion of pathologic entity. J Hand Surg 1984; 9A: 764-6.
20. Blanc F: Pachydermodactylye. Un trouble de la personalite. Abstract Dermatol Hebdo 1995; 255: 6-8.