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Pachydermodactyly: A Classical Case of This Rare Disease

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Sir,

Pachydermodactyly is an acquired digital fibromatosis of unknown etiology involving dorsolateral and proximal part of fingers and prsents as non- inflammatory soft tissue swelling. Only 62 cases have been reported in literature so far. It is found most commonly in young males, male to female ratio is estimated to be 5:1 [1]. Histopathology features include increased dermal collagen with little or no inflammatory cells. Here we describe typical presentation of this rare disease.

A 23 years old male student presented with asymptomatic persistent swelling localized over dorsolateral part of fingers II-IV bilaterally (right > left) for last 3 years (**Figures 1, 2**).

The swelling was more pronounced over PIP and MCP joint area sparing palmar part of fingers.

It was progressing in size initially, now it seems to be non-progressive for last 6 months. It was non tender and was having no signs of inflammation viz erythema, pain etc. overlying skin was hyperpigmented, grossly thickened and having mildly increased skin markings. Skin over toes was normal. There was no history of similar disease in family nor was the history of trauma or exaggerated manual work. Biopsy showed hyperkeratosis, hypergranulosis over a thickened dermis consisting of increased collagen. There were no inflammatory cells (**Figures 3, 4**).

Pachydermodactyly was first described by *Bazex* et al in 1973 as pachidermie digitale des primières phalanges [2] but later in 1975 was recognised as a variant of knuckle pad by *Verbov*. It is an acquired disorder affecting young males almost exclusively, though familial and female case have been reported in literature. Clinically it is characterized by soft tissue swelling over dorsal and lateral part of proximal phalanges bilaterally. Distal involvement is rare. Involvement of only 1 finger is extremely rare- only 4 cases have been reported [3]. The etiology is largely unknown but repeated minor trauma was postulated as etiology [3]. Baldazzi and cols has described five types [4] - classical form , localized form (monopachydermodactyly), transgressing form, familial and associated with tuberous sclerosis. Biopsy from the lesion shows hyperkeratosis, hypergranulosis overlying thickened dermis-consisting of increased col-

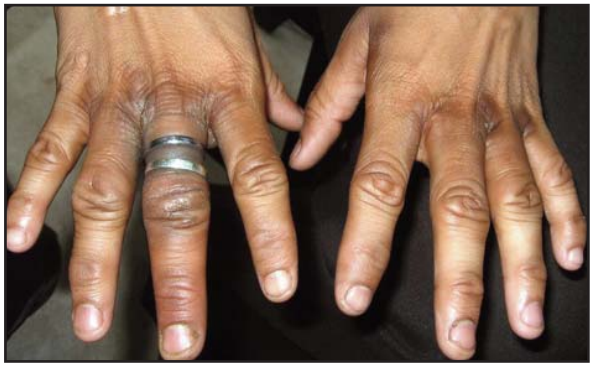

Figure 1. Bilateral diffuse ill-defined swelling over PIP and MCP joints

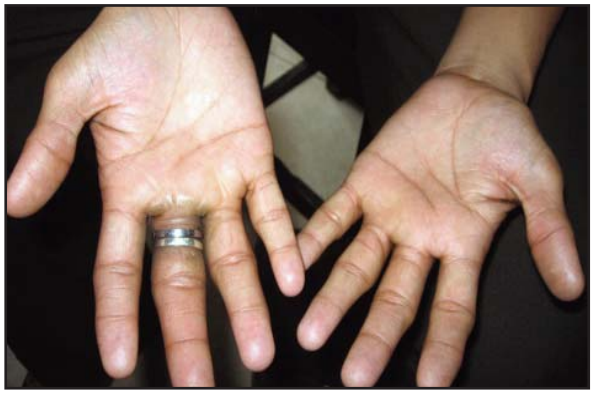

Figure 2. Sparing of palmar surface

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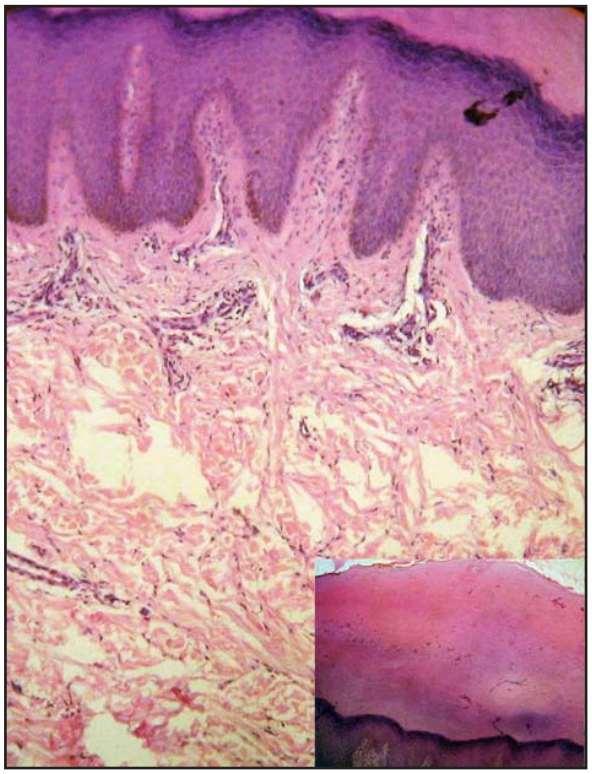

Figure 3. Photomicrograph (H&E stain x 100) showing acanthosis with grossly thickened dermis with absence of inflammatory cells. Insert picture showing hyperkeratosis

lagen sometimes arranged haphazardly arranged, slight proliferation of blood vessels with little or no inflammatory cells. Draluck et al using special stains showed increased deposition of collagen and mucin in dermis [**5**]. Further studies revealed increased collagen is of type 3 and 5, deviation from normal collagen profile [**5**]. Moreover diameter of collagen fibrils were found to be low by electron microscopy. Differential diagnosis include knuckle pad, post traumatic callosity, foreign body granuloma, fibroma, rheumatoid nodules, arthritis, thyroid acropachy. There is no successful therapy available. Topical therapy

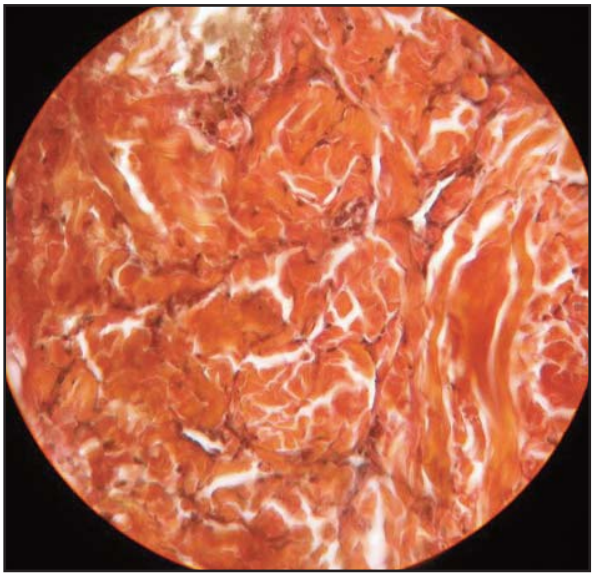

Figure 4. photomicrograph (VVG stain X 400) showing increased collagen

with steroid is ineffective. Intralesional steroid has been used with variable results [**3**]. Surgical excision is an effective option in selected cases.

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