Pachydermodactyly – a rare presentation of digital fibromatosis

K V Mallawaarachchi, C N Gunasekara, W S J Mendis

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Abstract

Pachydermodactyly is a rare acquired form of digital fibromatosis in which patients present with fusiform soft tissue swelling mainly around proximal interphalangeal joints. Etiology of the condition is unclear, but in most of the cases there is an element of repetitive mechanical injury to the skin. Because of its' clinical features, these patients could present to clinicians other than dermatologists such as rheumatologists and orthopedic surgeons. It is essential that the clinician identify this rather benign condition to avoid subjecting their patients to unnecessary and costly investigations. Herein, we report a 17 years old male who presented with soft tissue swelling of proximal interphalangeal joints of both hands over 5 months. He was diagnosed as having pachy-dermodactyly and treated successfully with intralesional triamcinolone acetonide injections.

Introduction

Pachydermodactyly, a term derived from the Greek words pachy (thick), dermo (skin) and dactylos (finger), is a form of digital fibromatosis characterized by soft tissue swelling mainly around the proximal interphalangeal (PIP) joints of fingers. It predominantly affects PIP joints of fingers II-IV mostly symmetrically[1]. Most of the cases are reported in otherwise healthy young men. Mean age of onset is 21.2 years and male to female ratio is around 3:2[1,2]. However, some clinicians believe that male to female ratio is as high as 5:1[3]. Familial cases are not common. This is a rare but distinct clinical entity, which is seen in healthy individuals and also in those with compulsive habits of hand manipulation.

Case report

A 17 years old student presented to the dermatology clinic with progressive swelling around PIP joint of both hands for 5 months' duration. He denied having any pain or morning stiffness of the joints and was also free of other systemic symptoms. Family history was not significant. This has been treated with potent topical steroids for some time without much improvement.

He was otherwise healthy physically and psychologically. However, after detailed questioning, he admitted having a habit of rubbing and interlacing fingers frequently.

Examination revealed a diffuse fusiform swelling of the lateral sides of PIP joints bilaterally and symmetrically. The swellings were more prominent on fingers II to IV while the little fingers were mildly affected (Figure 1). There were some areas of hypopigmentation on dorsal aspects of the fingers probably secondary to long term application of potent corticosteroids. The skin overlying metacarpophalangeal (MCP) joints was rough and loose. Nail changes were absent and palmer skin was normal. Joints were normal clinically and the range of motion was within normal limits.

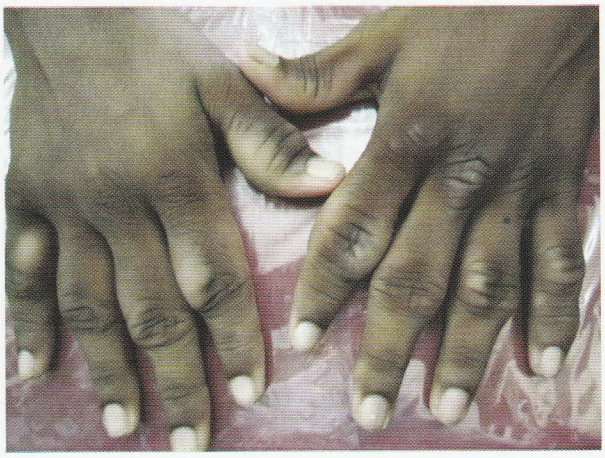

Figure 1.

Skin biopsy revealed marked hyperkeratosis, acanthosis and papillomatosis in the epidermis. Dermis showed irregular and thickened collagen bundles extending into subcutaneous tissue with minimal inflammation.

Hand x ray was reported as having bilateral para articular soft tissue thickening in the PIP joints. There were no bony or articular abnormalities radiologically. Complete blood count, liver and renal

[1]*Senior Registrar in Dermatology, Colombo North Teaching Hospital, Ragama, Presently, Acting Consultant Dermatologist, Base Hospital, Kahawatta,* [2]*Consultant Dermatologist, Colombo North Teaching Hospital, Ragama, Presently, Consultant Dermatologist, National Hospital, Sri Lanka* [3]*Consultant Dermatologist, Colombo North Teaching Hospital, Ragama, Sri Lanka.*

functions, CRP, rheumatoid factor and thyroid function tests were all within normal range.

The diagnosis of pachydermodactyly was made and patient was advised to avoid compulsive habit of manipulating the fingers. Intralesional triamcinolone acetonide (20 mg/ml) was administered twice one month apart. This led to a successful outcome and helped to improve the unsightly appearance of his fingers.

Discussion

Pachydermodactyly is diagnosed clinically depending on the clinical appearance of the fingers along with negative investigations for underlying systemic diseases. It is important to inquire about any joint symptoms and to perform a thorough examination of the joints to exclude underlying rheumatological conditions. Basic blood investigations and x ray examinations are worth doing as negative results will rule out systemic disease. However, extensive blood investigations and imaging are not necessary in otherwise healthy individuals.

Pathogenesis of pachydermodactyly is unclear but a detailed history about repetitive hand manipulation and minor injury to the skin is important in establishing the diagnosis. As noted in our patient, pachydermodactyly is reported in individuals who have the habit of repetitive finger lacing, rubbing and gripping of fingers[1,4]. These actions expose index, middle and ring fingers to exogenous friction which results in a reactive process of fibromatosis. This has also been seen in individuals with certain psychiatric disorders such as obsessive compulsive disease, anxiety and tic disorders. In addition, the condition has been reported in those who engaged in occupations and sports such as poultry processing workers and gymnasts which results in repetitive mechanical injury to the skin[2,5].

Histology usually reveals features of thick fibrotic dermis and variable degree of overlying hyperkeratosis and acanthosis. Mucin deposition in the dermis, entrapment of eccrine glands and vascular and fibroblast proliferation are also reported[6]. Increased collagen in the dermis consists of collagen types III and V which differs from the collagen profile of the normal dermis[1].

Pachydermodactyly is classified in to five different clinical types 1. Classical, 2. Localized, 3. Transgrediens - extending to metacarpophalangeal joints, 4. Familial and 5. Associated with tuberous sclerosis[7].

The differential diagnosis includes true and pseudo knuckle pads. The differences between them and pachydermodactyly are the locations and the clinical appearance. Knuckle pads are well circumscribed keratotic fibrous growths on the dorsal aspects of the inter phalangeal areas whereas pachydermodactyly is a diffuse less circumscribed swelling which is located laterally around PIP joints. In addition, true knuckle pads are known to co-exist with fibrosis of palmer, planter and penile aponeurosis fascia, a feature not seen in pachydermodactyly. Other differential diagnosis includes thyroid acropachy, juvenile chronic and rheumatoid arthritis, degenerative collagenous plaques of the hands, juvenile hyaline fibromatosis, pachydermoperiostosis and progressive nodular fibrosis of the skin.

Most clinicians opt not to treat pachydermodactyly actively as changing behavior and avoiding triggering factors help to improve the appearance of the fingers. When it comes to treatment there are only a few options available. Surgical resection of excessive tissue has been done successfully in some patients. Intra lesional steroid injection is also a proven treatment method which we used in our patient with good outcome. Higuchi et al has used Tranilast, a drug that inhibits collagen, to treat a young girl with pachydermodactyly successfully[8].

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