

Ledderhose disease: A case report with palmar fibromatosis, keloid and partial response to oral retinod

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ABSTRACT

Ledderhose disease is a rare hyperproliferative disorder of fibrous tissue. We present the case of a 40-year-old man who presented with bilateral plantar fibromatosis, dupuytren contracture and keloid, with partial response to oral acitretin.

Key words: Ledderhose disease; plantar fibromatosis; dupuytren contracture

INTRODUCTION

Plantar fibromatosis or ledderhose disease is a rare hyperproliferative disorder of fibrous tissue [1], occurring over the medial sole of the foot with tendency for local recurrence but do not metastasize [2]. Patient described in this report is a man with bilateral plantar fibromatosis, dupuytren contracture and keloid, with partial response to oral Acitretin.

CASE REPORT

A 40-years-old hospital employer man referred to our clinic with multiple painful nodules in plantar surface of his feet (Figs 1A-D), in spring 2013.

20 years ago (1994) he noticed stiffness and flexion deformity in the third finger of his right hand. The patient underwent two surgical operations with the diagnosis of dupuytren contracture. Six years later (2002) slow growing nodule appeared in plantar surface of his left foot, and it was gradually progressive with a dull ache type of pain. Initial management consisted of a skin biopsy of this foot lesion which led to the confirmation of scar tissue, then a surgery performed and report of pathology, confirmed the previous diagnose. A few months later lesion recurred again at the site of operation on left sole and additionally the same nodules occurred in right

sole (Figs 1A-D). The lesions were painful, somewhat preventing him from working. On examination, there were multiple skin colour to violaceous firm nodules with sharp margin and scale-crust on some of them in plantar, medial and lateral aspect of his feet (Figs 1A-D). The size of biggest nodule on right sole was 2.5*2.9 cm and 3.5*3.0 cm on left sole. No flexion contracture of toes or neurovascular deficits was noted. Motion of the feet was within the normal range. Also he had a scar of dupuytren contracture surgery in right hand and two keloides in his chest and shoulder (Fig. 2). The patient reported no history of any diseases such as diabetes, seizure, trauma and he did not consume alcohol. Also there was no family history of similar lesions.

Laboratory data such as CBC/ESR/FBS/renal and liver function tests were normal. Pathology slides reviewed again and in microscopy the epidermis revealed hyperkeratosis, hypergranulosis and irregular acanthosis. In the dermis, there was proliferation of bland looking spindle shaped fibroblasts arranged vertically, within dense collagenous stroma suggestive of fibromatosis (Figs 3A-C).

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

How to cite this article: Sadr Eshkevari Sh, Nickhah N. Ledderhose disease: A case report with palmar fibromatosis, keloid and partial response to oral retinod. Our Dermatol Online. 2015;6(1):52-55.

Submission: 05.09.2014; **Acceptance:** 04.11.2014

DOI: 10.7241/ourd.20151.13

MRI of lesions revealed small masses without invasion to muscle or tendon sheets (Fig. 4).

To management, we recommended him to change his job and suggested him not to stand for long time any more. Then prescribed the patient Acitretin 20 mg/day.

He consumed Acitretin for two months and stopped it for a month. However, the patient's symptoms, including pain reduced a lot and some of the nodules became flat and smaller in size (The dimensions of biggest nodule on right sole were 2.5*2.7 cm and 3.5*2.6 cm on left sole). In addition scales and crusts disappeared significantly (Figs 5A,B and 6A,B).

The patient's informed consent was obtained.

DISCUSSION

Fibromatoses are a family of fibroblastic proliferation sharing similar histology, an infiltrative pattern of growth and a tendency to recur after excision [2]. In general the superficial types are less aggressive than deep ones (such as desmoid) [3]. Plantar fibromatosis (ledderhose disease), palmar fibromatosis (Dupuytren contracture), penile fibromatosis (Peyronie's disease) and knuckle pads are benign superficial neoplastic proliferation of fibroblasts and/or myofibroblasts. Patients with palmar fibromatosis have additional areas of plantar fibromatosis in 5% to 20% of cases and penile fibromatosis in 2% to 4% of cases [4]. Recently, Trybus et al. reported an association rate of 14.85% of Ledderhose in 101 cases of Dupuytren contracture [5]. An increased incidence of knuckle pads has been observed [6,7]. Plantar fibromatosis occurs less frequently than palmar lesions with incidence of 0.23% [8].

Ledderhose disease, described in 1897, characterized by one or more slow growing nodules, most often on the medial half of the mid foot. Most lesions are asymptomatic and if they invade the adjacent structures they become painful. The pain increased by prolonged standing or walking and rarely produces contracture [9]. Nodules may be multiple in 33% of cases [10]. The condition affect any age but more often in younger age group, most often age 30 [11,12] and men are affected twice as often as females. Bilateral involvement is seen in 20-50% of cases [13,14].



Figure 1: (A and D) The patient presents with multiple firm nodules on sole of both feet



Figure 2: Scar of dupuytren contracture surgery and keloid in his chest

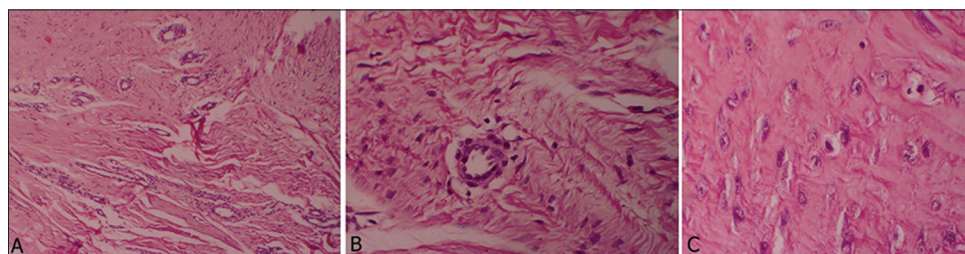


Figure 3: (A-C) Histopathology. Proliferation of bland looking spindle shaped fibroblasts arranged vertically, within dense collagenous stroma



Figure 4: Magnetic Resonance Imaging. Small masses without invasion to muscle or tendon sheets



Figure 5: (A) Sole lesions before treatment (left side) and after treatment (right side)



Figure 6: Lateral lesions before treatment (left side) and after treatment (right side)

The etiology of ledderhose disease remains unknown but genetic predisposition as well as repeated trauma

is thought to play a role in its pathogenesis [9]. Some association with diabetes, alcoholic liver disease and epilepsy have been described [2]. Our case had none of these associations or predisposing factors, also he didn't indicate any family history of fibromatosis. Radiographs are frequently normal in Ledderhose disease, since the lesion is not encapsulated. Evaluation is most commonly performed with ultrasound and MRI. MRI is an excellent, non invasive method for delineating the extent of the lesion and planning surgical treatment [15], as it showed no invasion to muscle and tendon sheet in our patient.

In asymptomatic patients conservative managements such as footwear modification may be helpful [13]. For those suffering from pain of large lesions which cause disability, excision of the lesion with total fasciectomy is desirable with the lowest incidence of recurrence [16]. Kan and Hovius reported no recurrence after graft repair in two brothers, with a long term follow up, ranging from 14-25 years [17].

Adjuvant radiotherapy decreases the rate of recurrences but should be used selectively because of its side effects [18]. High-energy focused extracorporeal shockwave therapy reduces pain in plantar fibromatosis [19].

There are no specific medical treatment for plantar fibromatosis, other than symptomatic relief and supportive measures. In the other hand, Viera et al reported that retinoic acid and other vitamin A derivatives produce a marked reduction in human fibroblast proliferation by interfering with DNA synthesis in vitro. Retinoids also exhibit an inhibitory effect on TGF- β 1-induced type I collagen gene expression in human fibroblasts [20]. Furthermore the proliferation and activity of cultured fibroblast cells and type III collagen synthesis are inhibited as the concentration of retinoids are increased in the medium [21]. Etretinate, free acid of etretinate and 13-cis retinoic acid(RA), reduce type IV collagen synthesis in vitro, the largest decrease being found with free acid of etretinate and 13-cis-RA [22].

So with regard to the results of a two monthes course of oral Acitretin in our case and above mentioned data, it may be a worth trying medical opportunity in cases, who deny surgery, with multiple surgical failure, or are not good candidate for surgery. However, these results require confirmation in future studies.

CONSENT

The examination of the patients was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the patient for publication of this article and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Source of Support: Nil, **Conflict of Interest:** None declared.