Ledderhose DiseaseAn Unusual Presentation

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ABSTRACT

Plantar fibromatosis, or Ledderhose disease, is a rare hyperproliferative disorder of the plantar aponeurosis. It may occur at any age with the greatest prevalence at middle age and beyond. This disorder is more common in men than woman and it is sometimes associated with other forms of fibromatosis. A 28-year-old Brazilian woman with a six-year history of painless bilateral plantar nodules is described in this article. (*J Clin Aesthet Dermatol.* 2010;3(9):45–47.)

Plantar fibromatosis, or Ledderhose disease (LD), is a rare hyperproliferative disorder of the plantar aponeurosis first described in 1894 by Dr. Georg Ledderhose.¹ Its etiology is unknown. The condition is essentially benign and often associated with Dupuytren's disease. The patient described in this article is a young woman with bilateral disease, no family history, and no predisposing factors associated with fibromatosis.

CASE REPORT

A 28-year-old Brazilian woman presented with a sixvear history of painless bilateral plantar nodules. On examination, she had subcutaneous nodules on the lateral aspect of both feet (Figure 1) and on the medial aspect of the left foot (Figure 2). She had thickening of the plantar fascia with small nodules, which coalesced into an indurated mass (Figure 3). The patient also had flexion contracture of the left hallux. Neither Dupuytren's disease nor knuckle pads were detected. There was no family history of similar nodules. The patient was not taking any medication and was otherwise healthy. Histological examination of a nodule demonstrated fibroblastic proliferation developing into a dense fibrosis. The nuclei of the fibroblasts were of uniform morphology, elongated, and without atypia or mitosis (Figures 4, 5, and 6). The cells were CD34 negative.

DISCUSSION

Superficial fibromatosis includes plantar fibromatosis (Ledderhose disease), palmar fibromatosis (Dupuytren's disease), penile fibromatosis (Peyronie's disease), and knuckle pads. Although its incidence is well described on the hands, it is less commonly seen on the feet.

LD is a relatively rare foot disorder of unknown etiology. Although it is listed as a "rare disease" by the Office of Rare Diseases (ORD) of the National Institutes of Health (NIH), which means that it affects less than 200,000 people in the United State's population, there are authors who report Ledderhose disease as a relatively common condition, with plantar contracture developing in approximately 25 percent of middle-aged or elderly individuals (1 of every 4 with Dupuytren's contracture).

It is characterized by local proliferation of abnormal fibrosis tissue in the plantar fascia. The nodules are typically slow growing and most often found in the central and medial portions of the plantar fascia. This tissue is locally aggressive and progressively replaces the normal plantar aponeurosis.⁴

LD may occur at any age with the greatest prevalence at middle age and beyond. This disorder is more common in men than woman and it is sometimes associated with other forms of fibromatosis, such as Dupuytren's disease in the hand, Peyronie's disease, or knuckle pads. Only 25

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Figure 1. Subcutaneous nodules on the lateral aspect of the right foot and medial aspect of left foot



Figure 2. Closer view of the medial aspect of the left foot showing the subcutaneous nodules and retraction



Figure 3. Thickening of the plantar fascia with small nodules, which coalesce into an indurated mass

percent of patients have bilateral involvement.⁴

In LD, as in Dupuytren's contracture, repeated trauma, diabetes, and epilepsy have been reported in association with the development of the lesions. $^{4-6}$

Children may also be affected by this process. Fetsch et al⁷ described 56 cases of palmar-plantar fibromatosis in children and preadolescents. A family history was available for 25 patients. Only one patient had concurrent disease involving both feet. Jacob et al⁸ proposed the term *benign anteromedial plantar nodules of childhood* to describe the distinct pediatric form of plantar fibromatosis.

Although in Dupuytren's disease an autosomal dominant inheritance has been demonstrated, familial cases of plantar fibromatosis are rare. Dartoy et al¹⁰ examined the relatives of seven patients with plantar fibromatosis and did not find any cases. Sammarco et

al¹¹ reported one familial case in a series of 18 patients, suggesting that inherited plantar fibromatosis is very rare. Graells Estrada et al⁹ reported an additional case of familial plantar fibromatosis.

For reasons unique to the anatomy of the plantar fascia, contracture of the toes is rare. ^{12,13} A differential diagnosis for the heel pain along the medial arch could be a benign thickening of the plantar fascia associated with plantar fibromatosis. ¹⁴

Nonsurgical treatments of LD include intralesional cortisone injections, ^{15,16} stretching, orthotics, and nonsteroidal antirheumatic drugs and physiotherapy. ¹⁶ Nonsurgical treatments should be performed when clinical symptoms occur. Surgical treatment is indicated in cases of persistent pain. The standard procedure includes a partial fasciectomy of the plantar aponeurosis. ¹⁶ After partial resection, there is a high recurrence rate with an increased risk of complications and more aggressive ingrowth into anatomical structures. ^{16,17} Some authors recommend a complete fasciectomy as the primary procedure of choice. ^{16–18} Postoperative radiotherapy can be used to diminish the chance of recurrence. ¹⁹

Any dermatologist who discovers Garrod's nodes or knuckle pads should search for early fibromatosis of the palmar and plantar fascia, or at least warn the otherwise unaffected patient that he or she may be susceptible to the development of palmar and plantar fibromatosis and Peyronie's disease.²⁰

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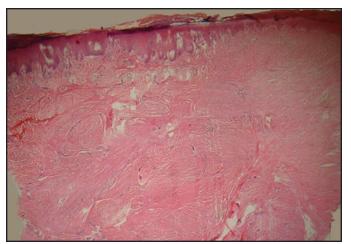


Figure 4. Low power view showing dermis replaced by fibroblastic proliferation with short fascicles or nodular disposition of proliferating cells (H&E, 4X)

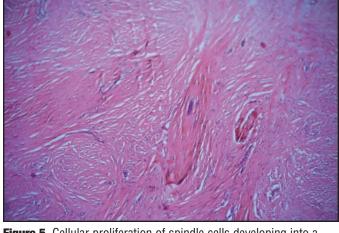


Figure 5. Cellular proliferation of spindle cells developing into a dense fibrosis associated with moderate amounts of collagen without atypia or mitosis (H&E, 10X)

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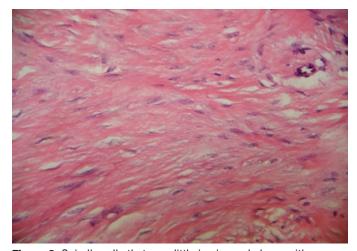


Figure 6. Spindle cells that vary little in size and shape with normochromatic nuclei, which are elongated with uniform morphology (H&E, 40X)

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