Rheumatology Quiz

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A 57-year-old woman with spondyloarthritis presented with a single subcutaneous nodular lesion (Figure 1A, arrow) on the plantar aspect of her left foot for a week. Mild pain at the lesion was precipitated only during walking. Ultrasound examination in longitudinal view of the lesion revealed a fusiform hypoechoic nodule (#) of the plantar fascia (PF). No internal flow was detected on the power Doppler image. (Not shown)

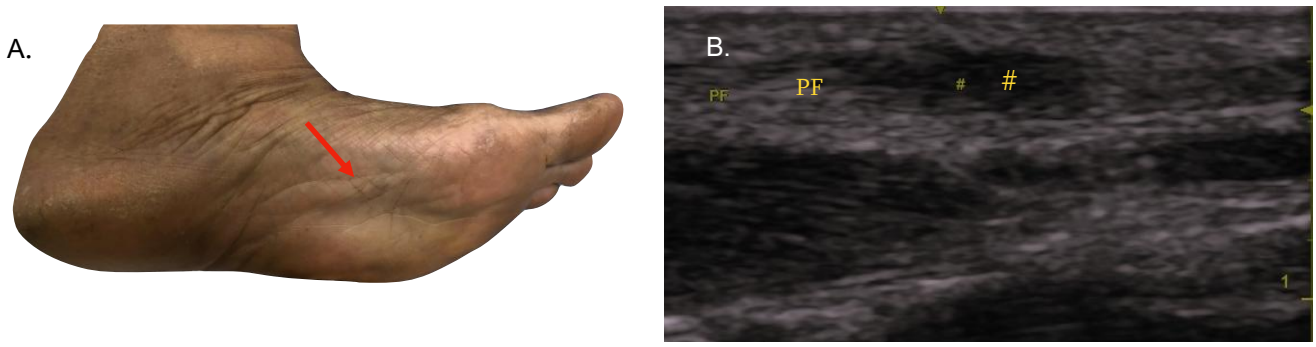

Figure 1

Questions

1. What is her diagnosis?
2. What are the conditions associated with the disease?

Answers

1. Ledderhose's disease (plantar fibromatosis).
2. Dupuytren's contracture, Peyronie's disease, knuckle pads, epilepsy, diabetes mellitus, frozen shoulder, chronic liver disease, alcoholism, smoking, repeated trauma, plantar fasciitis, and phenobarbital use.

Short review

Ledderhose's disease, or plantar fibromatosis, is a benign fibroblast proliferative disorder of the plantar aponeurosis characterized by the formation of fibrous nodules on the sole of the foot. While the condition is most famously associated with the German physician Dr. George Ledderhose, who presented 50 cases in 1897, earlier accounts may exist in the medical literature, with potential descriptions by Madelung in 1875.[1] It is a rare disease with a higher prevalence in males and typically presents in Caucasians over the age of 60.[2] Lesions are usually asymptomatic but may become painful. Bilateral involvement is seen in 25% of cases. It belongs to the superfamily of superficial fibromatoses, sharing a histopathological basis with Dupuytren's contracture (palmar) and Peyronie's disease (penile).[1,2] Ledderhose's disease usually does not cause flexion contracture of the structures, unlike Dupuytren's disease.[2] Patients present with slow-growing, firm nodules predominantly affecting the medial and central bands of plantar aponeurosis.[3,4] Nodules range from 0.5 to 3.0 cm.[4] The Sammarco and Mangone classification grades severity from focal lesions (Grade 1) to extension into skin and muscle (Grade 4).[2] The disease process involves benign fibroblast proliferation and excessive collagen deposition.[2] Unlike Dupuytren's contracture, Ledderhose's nodules often appear more hypercellular.[2]

Ledderhose's disease is frequently associated with various conditions, including systemic diseases such as diabetes mellitus, liver disease, epilepsy, and frozen shoulder.[2,4] Additionally, it is linked to fibromatosis disease, specifically Dupuytren's, Peyronie's disease or knuckle pads.[2,3,4] Furthermore, certain medications, such as phenobarbital, anti-TNF therapy, and Vemurafenib, have been associated with disease progression.[2,4] Lastly, repeated trauma, chronic alcohol consumption and smoking are considered lifestyle factors that may contribute to the development of Ledderhose's disease.[3,4]

Ultrasound and magnetic resonance imaging (MRI) are the images of choice, though ultrasound is the most accessible and less costly imaging test for diagnostic confirmation. On ultrasound, findings include hypoechoic, fusiform nodules with a lack of intrinsic vascularity on Doppler.[3,4,5] Alternating linear bands of hypoechogenicity and isoechogenicity, known as "Comb Sign," can be seen in 51% of cases.[4] MRI demonstrates nodules that typically show low signal intensity on both T1- and T2-weighted sequences due to its fibrous nature. Hyperintensity on T2 suggests high cellularity and a more aggressive lesion.[2,3,5] MRI is a valuable tool that aids in assessing the severity and determining the extent of the disease.[6] Consequently, it plays a crucial role in surgical planning.

Differentiation from other foot pathologies is crucial. In plantar fasciitis, pain is proximal, at the calcaneal insertion, in contrast to Ledderhose nodules, which are typically located in the mid-substance.[2,5] In chronic plantar fascial ruptures, there is usually a history of trauma or corticosteroid injection, and the tear can be diagnosed through a clinical examination or ultrasound imaging.[5] When a foreign body is suspected, ultrasound or MRI can aid in the diagnosis. Sarcoma must be excluded in cases of rapid growth or atypical imaging.[2]

Treatment aims to alleviate pain and restore function. Conservative management is the standard first-line approach due to high surgical recurrence rates. In asymptomatic patients, they may be kept under observation. If they start to have pain when walking, nonsteroidal anti-inflammatory drugs (NSAIDs) and physical therapy can be initiated. The rehabilitation program aims to achieve pain relief, manage contractures, strengthen muscles, improve functionality and mobility, and correct gait.[7] They can start wearing custom-made orthotics or pads that help diminish the stress on the fascia.[2,8] Intralesional corticosteroid may soften nodules and reduce pain by inhibiting fibroblast proliferation. However, it risks fat pad atrophy and fascia rupture.[8] Other treatment options in Ledderhose's disease include radiotherapy, topical or intralesional verapamil, and collagenase clostridium histolyticum.[2,8]

Surgery is reserved for refractory pain or functional disability. Recurrence is the primary limiting factor. There are three primary surgical interventions, including local excision, wide excision, and complete fasciectomy.[4,8] Recurrence rates following excision are approximately 60%.[2,8]

Diagnosis of Ledderhose's disease is based on physical examination and complementary imaging tests consistent with the nature of this disease. In rare cases, histological confirmation is required. For rheumatologists, recognizing Ledderhose's disease is essential not only for differential diagnosis against inflammatory enthesopathies but also due to its frequent association with systemic conditions.

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