

Palmar-Plantar Fibromatosis and Knuckle Pads Associated with Alcohol Consumption

Alkol Tüketimine Bağlı Gelişen Palmar-Plantar Fibromatozis ve Knuckle Pads

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ABSTRACT

Fibromatosis is a benign, locally proliferative disorder of fibroblast. Palmar fibromatosis (Dupuytren disease) is the most common type of the superficial fibromatosis however plantar fibromatosis, or Ledderhose disease is rare. Coexisting conditions include knuckle pads, Peyronie disease, diabetes mellitus, alcoholism, liver disease and epilepsy. Herein, we present a heavy alcohol drinker patient with nodules in his left foot sole and over dorsal aspects of the proximal interphalangeal joints of his hands. He was diagnosed with palmar-plantar fibromatosis and knuckle pads.

Keywords: Fibromatosis, knuckle pads, alcohol

ÖZET

Fibromatozis benign, lokal proliferatif bir fibroblast hastalığıdır. Palmar fibromatozis (Dupuytren hastalığı) en sık görülen süperfisyal fibromatozis tipi iken plantar fibromatozis (Ledderhose) hastalığı nadirdir. Knuckle pads, Peyronie hastalığı, diabetes mellitus, alkolizm, karaciğer hastalığı ve epilepsi durumlarıdır. Burada sol ayak tabanında ve ellerinde proksimal interfalanjial eklemlerinin dorsal yüzlerinde oluşan nodülleri olan alkolik bir erkek hasta sunulmaktadır. Bu hasta palmar-plantar fibromatozis ve knuckle pads tanılarını almıştır.

Anahtar sözcükler: Fibromatozis, knuckle pads, alkol

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Introduction

The fibromatosis are a broad group of fibroblastic proliferations. Palmar fibromatosis (Dupuytren disease) is the most common type of it (1). However plantar fibromatosis (Ledderhose disease) occurs less frequently than the palmar lesion, with an incidence of 0.23% (2). Coexisting conditions include knuckle pads, Peyronie disease, diabetes mellitus, alcoholism, liver disease and epilepsy (3-5). Here we describe a heavy alcohol drinker patient with nodules in his left foot sole and over dorsal aspects of the proximal interphalangeal (PIP) joints of his hands.

Case Report

A 49-year-old male visited us for the complaint of painful nodule in his left foot sole especially by walking or standing for long periods of time. He mentioned that the nodule grew gradually in several months. It was



Figure 1. The hard nodule in medial aspect of the longitudinal plantar arch of the patient's foot.



Figure 2. The fibrotic thickenings of bilateral palmar fascias and the nodules at the distal palmar creases opposite to the fourth fingers.

troubled enough for him to request surgical cure. His family and medical history were unremarkable unless he declared that he had been an alcohol drinker since in his early twenties. His alcohol consumption was 50g/day. He did not participate in any sports or hobbies where he would have to use his feet and hands. He was a small trader.

On examination there was a hard nodule in the medial aspect of the plantar arch of his left foot (Figure 1). It was painful with palpation. He had also a fibrotic thickening of the bilateral palmar fascia of his hands. There were painless nodules at the distal palmar crease opposite to his fourth fingers (Figure 2). Additionally he had normal skin color nodules at the dorsal aspect of proximal interphalangeal (PIP) joints of the bilateral index, middle, ring and small fingers of his hands (Figure 3). They were asymptomatic but cosmetically unappealing. The range of motions of hand and foot joints were normal.

Blood samples were shown no abnormalities except alanine transaminase (ALT), level was high (126 U/L, normal:<50). Radiographies of the hands and feet revealed no abnormalities. On his left foot Magnetic Resonance Imaging (MRI), a well-defined mass in the medial aspect of the plantar aponeurosis was seen. The lesion demonstrates heterogeneous signal intensity that was isointense to hyperintense in comparison to the skeletal muscle on T1- and T2-weighted images and marked heterogeneous enhancement was seen post-intravenous gadolinium (Figure 4A-C).

He was consulted with Plastic and Reconstructive Surgery and nodules in his hands and left foot sole were explored. On macroscopic examination the tissue was



Figure 3. The nodules at dorsal aspect of the proximal interphalangeal joints of bilateral index, middle, ring and small fingers.

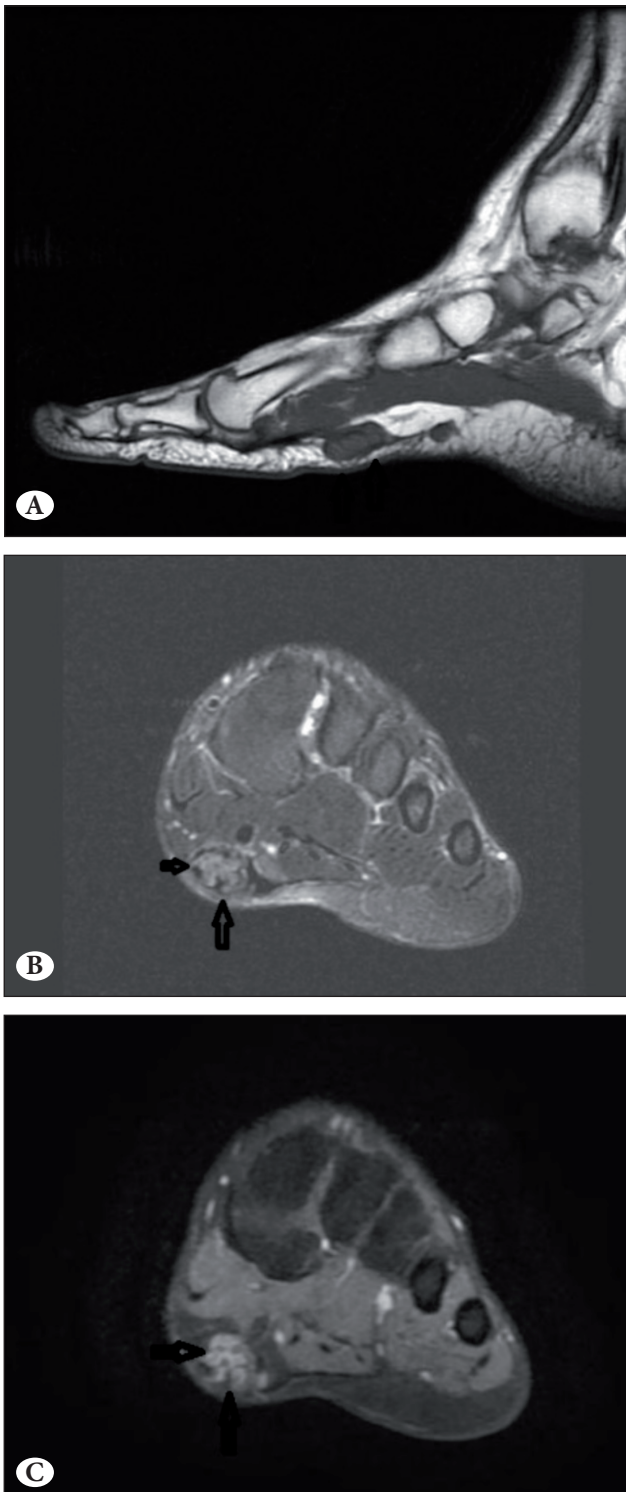


Figure 4. A) Sagittal T1- weighted image B) Coronal T2-weighted fat-suppressed image C) Coronal T1-weighted fat-suppressed contrast- enhanced image show heterogeneously enhancing lesion in the plantar aponeurosis.

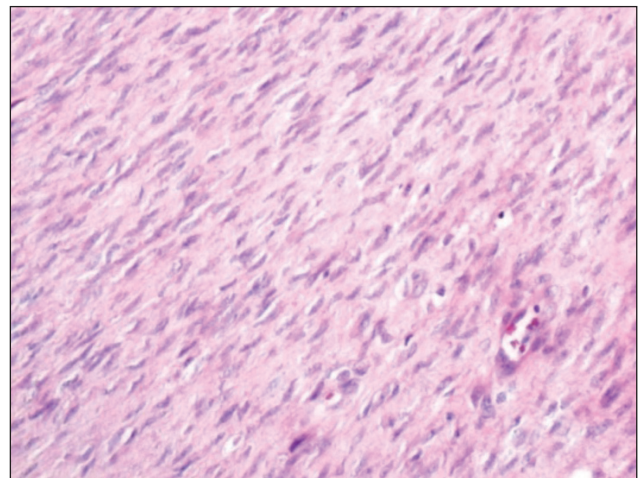


Figure 5. A well defined lesion consists of fibroblasts with minimal increase in cellularity and collagen deposition. Fibroblasts were uniform in size and shape with normochromatic nuclei and small pinpoint nucleoli. Mitotic activity was rare with no atypical mitotic figure.

firm and on cut sections showed gray-white surface. Histological examination revealed a well defined lesion consists of fibroblasts with minimal increase in cellularity and collagen deposition. Fibroblasts were uniform in size and shape with normochromatic nuclei and small pinpoint nucleoli. Mitotic activity was rare with no atypical mitotic figure (Figure 5). The nodules were pathologically diagnosed as fibromatosis and knuckle pads.

Discussion

Fibromatosis represents a wide group of benign, locally proliferative disorder of fibroblast with a similar histologic appearance containing spindle-shaped myofibroblastic cells, dense deposits of intercellular collagen fibers, variable amounts of extracellular myxoid matrix, and compressed and elongated vessels (6).

Palmar fibromatosis (Dupuytren disease) is the most common type of the superficial fibromatosis (1). Clinically, patients present with painless, subcutaneous nodules involving the palmar aspects of the fingers, usually the fourth and fifth digits (7). The nodules may progress to fibrous cords and cause traction on the underlying flexor tendons of the fingers and this results in the flexion contractures. The process is bilateral in 40–60% of patients (1,8,9). Radiographs may be normal or demonstrate flexion contractures of the metacarpophalangeal (MCP) and PIP joints (1).

Coexisting morbidities include plantar fibromatosis, knuckle pads, penile fibromatosis (Peyronie's disease),

diabetes mellitus, epilepsy, keloids, alcoholism, liver disease, manual labor with vibration exposure, smoking, hyperlipidemia and complex regional pain syndrome (1-5,10). Several studies have shown a high prevalence of Dupuytren's disease in alcoholic patients but the evidence from controlled studies is conflicting. Su et al. found the prevalence of Dupuytren's disease was higher among heavy drinkers in only 51- to 60-year age band (11). Burge et al. established that Dupuytren's contracture was associated with an Alcohol Use Disorders Test (AUDIT) score greater than 7. In this study mean weekly alcohol consumption was 7.3 units for cases and 5.4 units for controls ($p = 0.016$) (12). Liver disease in itself has also been suggested as an etiological factor in Dupuytren contracture. Results of morphological investigation of hepatic tissues of persons suffering from Dupuytren contracture chronic hepatitis and hepatic fibrosis were adduced (13). Attali et al. found a similar prevalence between alcoholic patients with and without liver disease, leading to the conclusion that alcohol rather than liver disease was associated with Dupuytren contracture. However, it was noted that in those with chronic cirrhotic liver disease, the presence of Dupuytren contracture strongly suggested an alcoholic cause, with a 90% positive predictive value (14). Valencia-Martin et al. reported the threshold between average moderate drinking and average heavy drinking was $\geq 40g/day$ (15). Our patient's alcohol consumption was 50g/day. It seems that alcohol has a role in the aetiology of fibromatosis in our patient. He had been a heavy alcohol drinker since in his early twenties and elevated ALT level was the only concomitant pathologic laboratory finding. The mechanism whereby alcohol intake is associated with concomitant is unclear. Suggestions include effects on local circulation in palm, damage to fatty tissue provoking a fibrotic response, and changes in prostaglandin production, but none of these mechanisms has been clearly established (10).

Jones proposed the term knuckle pads in 1923 as circumscribed fibromatous thickenings overlying finger joints (16). They are mostly noted on dorsal side of PIP joints, but are occasionally seen on MCP and distal interphalangeal (DIP) joints (17). The clinical characteristic in our case was that all knuckle pads were formed on the PIP, but not the MCP and DIP joints. The onset age varies, but onset in 40s is most frequent. They grow to be some 10–15 mm in diameter in the course of a few weeks or months, then persist permanently (18). Generally, they are considered benign, occasionally associated with pain when they first appear, sometimes cosmetically embarrassing and sometimes spontaneously disappearing. It may be inherited or acquired. Trauma is not a significant factor, but some reports have described external factors as the most common cause of knuckle pads in adults (19). Our patient's nodules were

asymptomatic but cosmetically unappealing. He had not used his hands for any specific task in his job, and had no past medical history associated with external factors, suggesting that the alcohol intake might be role of disease occurrence. Histopathological characteristics of knuckle pads are similar to palmar fibromatosis (20). Occupational callosities, Herberden's nodes of osteoarthritis, rheumatoid nodules, pachydermodactyly, granuloma annulare and erythema elevatum diutinum, are generally considered for differentiation from knuckle pads (17). Our case was clinically and pathologically distinguished from these diseases.

Plantar fibromatosis, or Ledderhose disease is a hyperproliferative disorder of plantar aponeurosis first described in 1894 by Dr. Georg Ledderhose (21). It is listed as a "rare disease" by the Office of Rare Diseases of the National Institutes of Health, which means that it affects less than 200,000 people in the United State's population (22). The etiology of plantar fibromatosis remains controversial, with prior trauma considered likely. Chromosomal variations have been seen in some lesions (23). It may occur at any age with the greatest prevalence at middle age and beyond. This disorder is more common in men than woman (24). Patients present with one or more subcutaneous nodules, which most frequently arise in the medial aspect of the plantar arch (78%) and can extend to the skin or deep structures of the foot (25).

Most lesions are asymptomatic, only becoming symptomatic when the lesion invades adjacent structures such as neurovascular bundles, muscles, or tendons. Alternatively, some patients complain of aching pain after walking or standing for long periods of time (1). Our patient's foot pain was the only complaint of him to visit us and troubled enough for him to request surgical cure.

Radiographs are frequently normal in patients with plantar fibromatosis. MRI is the best modality to determine infiltration of the lesion into the surrounding tissues and therefore it is most helpful for preoperative planning. MRI imaging may demonstrate well or ill-defined superficial lesions along the deep plantar aponeurosis, which typically blend with the adjacent plantar musculature. Lesions typically show heterogeneous signal (92%), which is isointense to hypointense to skeletal muscle on T1W (100%) and T2W (78%) sequences (1).

The treatment of plantar fibromatosis is often conservative and consists of footwear modifications aimed at relieving symptoms. Radiotherapy, extracorporeal shockwave therapy and surgical treatment also have been applied (1). Surgical resection was needed for our patient's lesion. Because it caused significant disability and was refractory to nonoperative

methods of management.

We think that clinicians should be aware of the possibility of coexistence palmar fibromatosis and knuckle pads with Ledderhose disease which is the rare form of fibromatosis. We consider that patients who present with fibromatosis should be informed of the association with alcohol consumption and advice to reduce excessive alcohol consumption is important.

References

1. Walker EA, Petscavage JM, Brian PL, Logie CI, Montini KM, Murphey MD. Imaging features of superficial and deep fibromatoses in the adult population. *Sarcoma*, vol.2012, Article ID 215810, 17 pages, 2012. doi:10.1155/2012/215810.
2. Yost J, Winters T, Fett HC. Dupuytren's contracture: a statistical study. *The American Journal of Surgery* 1955; 90: 568-571.
3. Couto-Gonzalez I, Brea-Garcia B, Taboada-Suárez A, González-Álvarez E. Aggressive Dupuytren's diathesis in a young woman. *BMJ Case Rep* 2010 Sep 20; 2010.pii: bcr1220092592. doi: 10.1136/bcr.12.2009.2592.
4. Caroli A, Zanasi S, Marcuzzi A, Guerra D, Cristiani G, Ronchetti IP. Epidemiological and structural findings supporting the fibromatous origin of dorsal knuckle pads. *J Hand Surg Br* 1991; 16: 258-262.
5. Geoghegan JM, Forbes J, Clark DI, Smith C, Hubbard R. Dupuytren's disease risk factors. *J Hand Surg Br* 2004; 29: 423-426.
6. Weiss SW, Goldblum JR, Enzinger FM. Fibromatoses. In: Weiss SW, Goldblum JR, ed Enzinger and Weiss' Soft Tissue Tumors. Philadelphia, USA: Mosby Elsevier, 2008: 227-228.
7. Ellis H. Baron Guillaume Dupuytren: Dupuytren's contracture. *J Perioper Pract* 2013; 23: 119-120.
8. Gonzalez SM, Gonzalez RI. Dupuytren's disease. *West J Med* 1990; 152: 430-433.
9. Trojian TH, Chu SM. Dupuytren's disease: diagnosis and treatment. *Am Fam Physician* 2007 Jul 1; 76: 86-89.
10. Hart MG, Hooper G. Clinical associations of Dupuytren's disease. *Postgrad Med J* 2005; 81: 425-428.
11. Su CK, Patek AJ Jr. Dupuytren's contracture. Its association with alcoholism and cirrhosis. *Arch Intern Med* 1970; 126: 278-281.
12. Burge P, Hoy G, Regan P, Milne R. Smoking, alcohol and the risk of Dupuytren's contracture. *J Bone Joint Surg Br* 1997; 79: 206-210.
13. Iskra NI, Shatrova KM, Hur'iev SO, Kuz'min Vlu, Roshchin HH. Morphological changes in hepatic tissues in patients, suffering Dupuytren's contracture and chronic hepatitis. *Klin Khir* 2013; 2: 15-19.
14. Attali P, Ink O, Pelletier G, Vernier C, Jean F, Moulton L, et al. Dupuytren's contracture, alcohol consumption, and chronic liver disease. *P. Arch Intern Med* 1987; 147: 1065-1067.
15. Valencia-Martín JL, Galán I, Guallar-Castillón P, Rodríguez-Artalejo F. Alcohol drinking patterns and health-related quality of life reported in the Spanish adult population. *Prev Med* 2013; 57: 703-707.
16. Jones HW. Two cases of "knuckle pads". *Br Med J.* 1923; 1: 759.
17. Koba S, Misago N, Narisawa Y. Knuckle pads associated with clubbed fingers. *J Dermatol* 2007; 34: 838-840.
18. Addison A. Knuckle pads causing extensor tendon tethering. *J Bone Joint Surg Br* 1984; 66: 128-130.
19. Mackey SL, Cobb MW. Knuckle pads. *Cutis* 1994; 54: 159-160.
20. Lagier R, Meinecke R. Pathology of "knuckle pads". Study of four cases. *Virchows Arch A Pathol Anat Histol* 1975; 365: 185-191.
21. Ledderhose G. Über zerreisungen der plantarfascie. *Arch Klin Chir* 1894; 48: 853-856.
22. Fausto de Souza D, Micaelo L, Cuzzi T, Ramos-E-Silva M J. Ledderhose disease: an unusual presentation *Clin Aesthet Dermatol.* 2010; 3: 45-47.
23. Murphey MD, Ruble CM, Tyszko SM, Zbojniec AM, Potter BK, Miettinen M. From the archives of the AFIP: musculoskeletal fibromatoses: radiologic-pathologic correlation. *Radiographics* 2009; 29: 2143-2173.
24. Fausto de Souza D, Micaelo L, Cuzzi T, Ramos-E-Silva M. Ledderhose disease: an unusual presentation. *J Clin Aesthet Dermatol* 2010; 3: 45-47.
25. Morrison WB, Schweitzer ME, Wapner KL, Lackman RD. Plantar fibromatosis: a benign aggressive neoplasm with