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Ahmad Almigdad
Department of Orthopedic, Royal Medical Services, Amman, Jordan

Ayman Mustafa
Department of Orthopedic, Royal Medical Services, Amman, Jordan

Ahmad Alzoubi
Department of Orthopedic, Royal Medical Services, Amman, Jordan

Yasmin AlSaidat
Department of Pathology, Royal Medical Services, Amman, Jordan

Kamal Hurani
Department of Orthopedic, Royal Medical Services, Amman, Jordan

Alaeddin Al-Wakhyan
Department of Orthopedic, Royal Medical Services, Amman, Jordan

Corresponding Author:
Ahmad Almigdad
Department of Orthopedic, Royal Medical Services, Amman, Jordan

A typical presentation of palmar fibromatosis in adult female: A case report

Ahmad Almigdad, Ayman Mustafa, Ahmad Alzoubi, Yasmin AlSaidat, Kamal Hurani and Alaeddin Al-Wakhyan

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Abstract
Palmar fibromatosis is a benign proliferative disorder of the palmar fascia which leads to the formation of cords and nodules and progressive contracture of the fingers and thumb and consequently affects hand function.
In this review, we report an atypical presentation of palmar fibromatosis in a thirty-one-year lady who underwent surgical excision for a mass in the second interphalangeal web space extending to the middle finger's radial side. The histopathological study of excised mass was compatible with the diagnosis of palmar fibromatosis.

Keywords: Dupuytren's disease, palmar fibromatosis, tumor

Introduction
Palmar fibromatosis (also known as Dupuytren's Disease) is a benign proliferative disorder of the palmar fascia which leads to the formation of cords and nodules and progressive contracture of the fingers and thumb and consequently affects hand function [1-4]. Genetic, immunological and environmental factors interact to promote the development of palmar fibromatosis [5-7].
Males are affected twice compared to females, and Caucasian males of northern European descent are mostly affected [8]. The ring finger is the most common site, followed by the little finger [3]. Dupuytren disease has ectopic manifestations such as Lederhosen disease, where the plantar fascia is involved [9]. Mild disease is treated by physiotherapy and Clostridium histolyticum collagenase injection, while surgical fasciotomy is needed in metacarpophalangeal joint flexion contractures exceeding thirty degrees and interphalangeal flexion contractures [10-12].
In this review, we report an atypical presentation of palmar fibromatosis in a thirty-one-year lady who underwent surgical excision for a mass in the second interphalangeal web space extending to the middle finger's radial side. The histopathological study of excised mass was compatible with the diagnosis of palmar fibromatosis.

Case report
A thirty-one-year lady presented with a symptomatic mass on the right hand in the radial side of the base of the middle finger with hardening on the volar aspect of the second web space for a three-year duration. The mass doubled in size over the last three months, and the patient developed new-onset pain localized to the mass and horseshoe paresthesia at the radial side of the middle and ulnar side of the index finger. The patient did not recoil a history of trauma to the finger, and the hand function was not impaired.
Radiograph demonstrated soft tissue swelling around the proximal phalanx of the middle finger with well-defined soft tissue calcification on the volar radial side of the proximal phalanx and erosion in the middle of the proximal phalanx as well as the third metacarpal head, Figure 1.

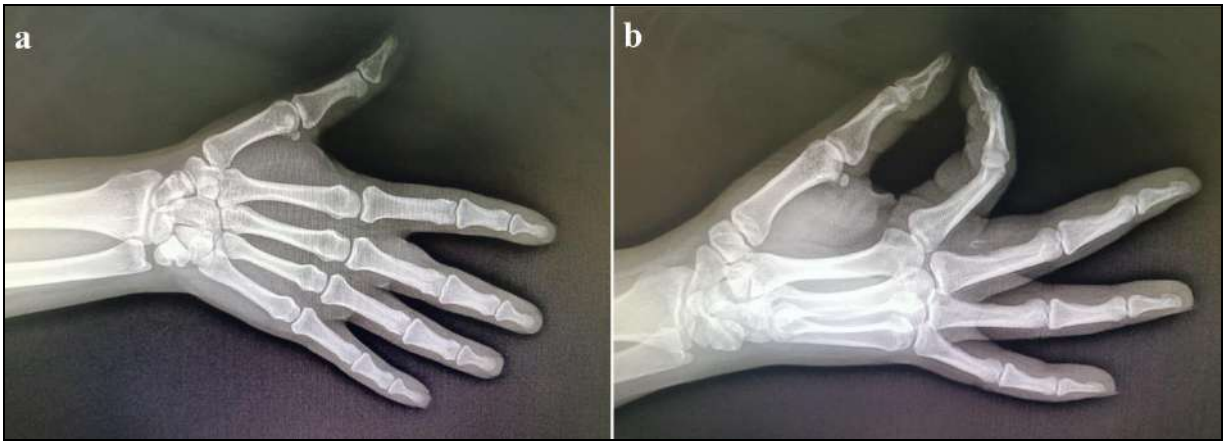

Fig 1: Hand X-ray. a) Anterior-posterior view revealed soft tissue swelling around the middle finger proximal phalanx. b) Oblique view showed well-defined soft tissue calcification on the volar radial side of the proximal phalanx and cortical erosion in the proximal phalanx and the third metacarpal head

MRI revealed a 6 * 13 * 21 mm T1 hypointense area in the volar radial aspect of the middle finger proximal phalanx. The post-contrast study suggests an inflammatory process with fibrotic changes and a 3.5 mm subchondral cyst in the radial side of the third metacarpal head and a 3 mm cyst on the proximal phalanx, most likely an erosive lesion, Figure 2.

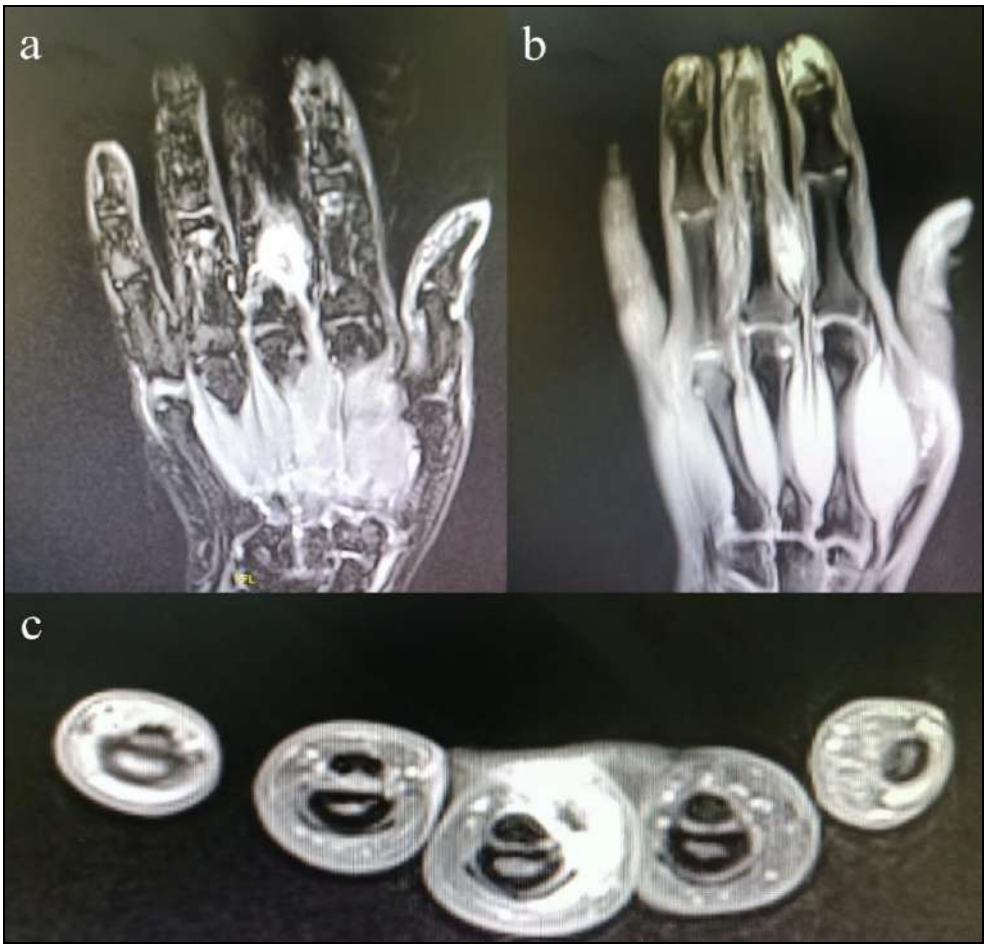

Fig 2: a) STIR Coronal MRI. b, c) Post-contrast T1 coronal and axial MRI cut demonstrated hyperintense lesion on the volar radial aspect of the proximal phalanx of the middle finger with central hypo intensity in the post contrast study, features suggestive of an inflammatory process

The patient underwent surgical excision under Wide Awake Local Anesthesia No Tourniquet (WALANT). Z incision was planned on the palm over the second web space and extended directly on the radial side of the middle finger, Figure 3. Surgical dissection revealed extensive fibrous tissue on the radial side of the middle finger and a grayish soft tissue mass on the first web space measuring 2*2*0.5 cm. The radial digital nerve of the middle finger was injured with dissection of the tough fibrous tissue tethered to the radial side of the middle finger proximal phalanx. The wound was closed using interrupted non-absorbable 3/0 Nylon, the dressing was applied, and the patient was encouraged regarding occupational therapy.

Fig 3: a) Z-incision was planned over mass in the volar aspect of the palm and extended to the radial side of the middle finger. b) Firm grayish soft tissue mass measuring 2*2*0.5 cm on the volar second web space. c) Post mass, and fibrous tissue excision revealed intact flexor tendon and A2 pulley.

Excised specimens were sent for histopathological study. Microscopic examination showed fragments of hypocellular lesion composed of bland spindle cells with pale eosinophilic cytoplasm and dense collagen bundles in the background with no evidence of necrosis or increased mitotic activity. Epithelioid sarcoma was excluded as a differential diagnosis by the lack of mitotic activity, atypia, and complementary studies, which revealed that some cells showed nuclear reactivity for b-catenin but not for cytokeratin immunostain. Therefore, the microscopic features and ancillary studies are compatible with the palmar fibromatosis diagnosis, Figure 4.

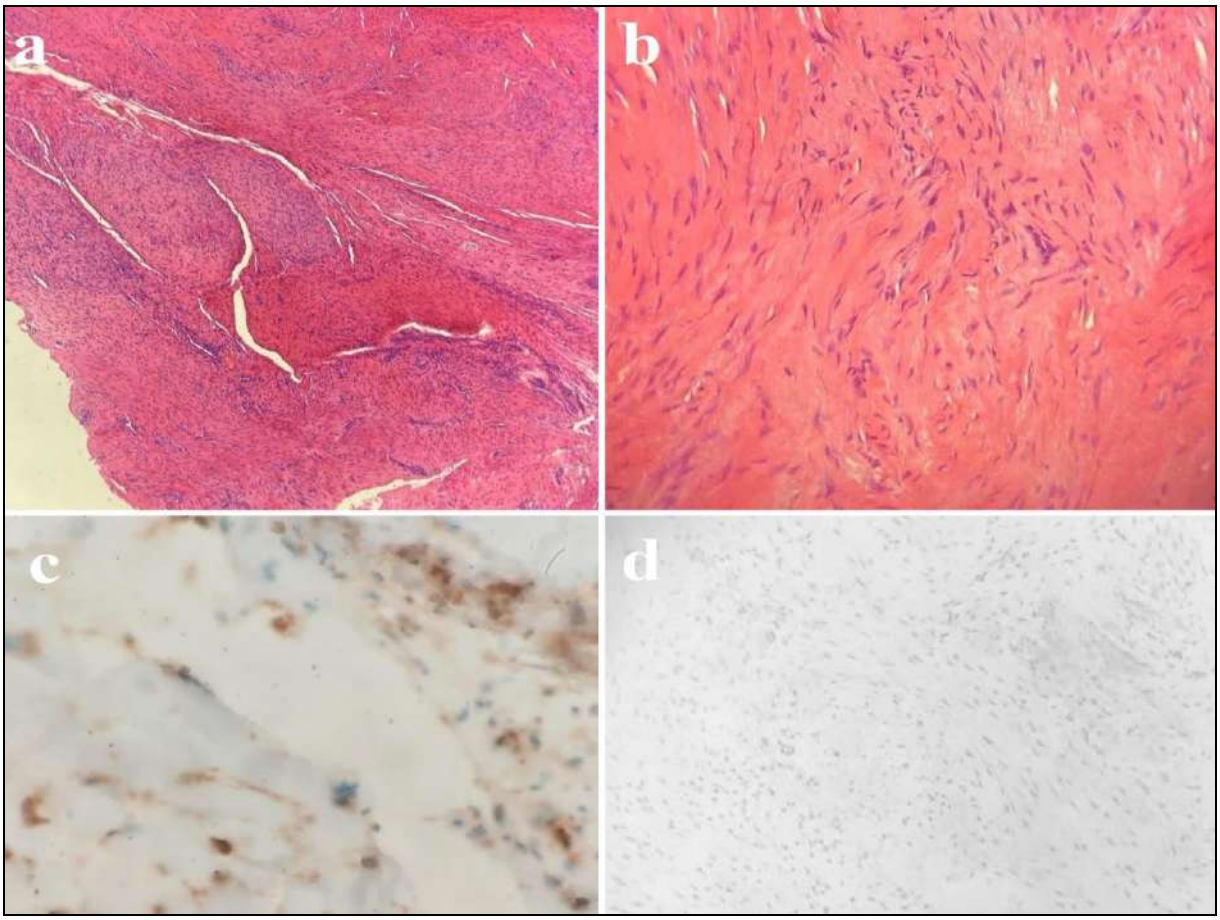

Fig 3: a, b) Haematoxylin and Eosin section reveals bland oval to spindle cells arranged in fascicles with some collagen bundles in between; no evidence of necrosis, pleomorphism, or increased mitotic activity. c, d) Ancillary studies revealed that some cells showed nuclear reactivity for b-catenin (c) but not for cytokeratin immunostain (d).

Discussion

Tumors and tumor-like lesions are frequent in hand, and the orthopedic surgeon should be familiar with the clinical characteristic of each tumor [13]. However, some pathologies are atypically presented and require a scientific approach for correct diagnosis and management. The palmar fibromatosis (Dupuytren's Disease) appeared as firm nodules that expand into fibrous collagenous cords and extend into the digits. With the progression of the disease, the cords mature, thicken and contract, resulting in permanent flexion deformities [14]. About 20 to 40% of patients develop a flexion deformity that affects hand function [15].

Dupuytren's disease is diagnosed by the presence of subcutaneous nodules and cords that progress to flexion

contractures of the digits. However, imaging studies are just needed in certain patients whose presentation is not classical. Ultrasound and MRI are helpful modalities for the atypical presentation of the disease to differentiate it from other soft tissue pathologies [16, 17]. Nevertheless, histology is the confirmatory diagnosis in such a scenario [3].

In this review, we reported a thirty-one-year lady who developed an asymptomatic mass on the radial side of the proximal phalanx of the middle finger for three years duration. The mass doubled in size and extended to the volar second interphalangeal web space in the last three months, and the patient developed new-onset paresthesia at the radial side of the middle and ulnar side of the index fingers due to compression of the tumor on the digital nerve. The imaging workup suspected an inflammatory process and the possibility of a foreign body. However, histological studies were compatible with palmar fibromatosis. Therefore, we report this case because the clinical presentation regarding the location, features, and progression, and the MRI finding of the mass, were not the classical scenarios.

Conclusion

Palmar fibromatosis typically presents as nodules and cords. However, palmar fibromatosis may present as a deep mass in hand with atypical features. Therefore, palmar fibromatosis should be considered in a differential diagnosis for hand tumors.

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