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Pachydermodactyly: A Case Report of a Little-Known and Benign Form of Digital Fibromatosis

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Male, 16-year-old
Final Diagnosis: Pachydermodactyly
Symptoms: —
Medication: —
Clinical Procedure: —
Specialty: Orthopedics and Traumatology • Radiology • Rheumatology

Objective: Rare disease**Background:** Pachydermodactyly, a benign and self-limiting cause of cutaneous fibromatosis, is an under-recognized condition. This little-known condition is important, as it mimics inflammatory arthropathy.**Case Report:** A 16-year-old male presented with a 2-year history of progressive, asymptomatic soft tissue swelling of the proximal interphalangeal joints (PIPJs) affecting the second to fourth fingers bilaterally. He had participated in kayaking for the last 1–2 years. Physical examination revealed only painless, saccular, subcutaneous swelling around the PIPJs. Plain radiograph and MRI showed cutaneous thickening around the PIPJs, with no evidence of arthropathy. Blood investigations including inflammatory markers and autoimmune panel were normal. Skin biopsy showed thickened collagen in the dermis, acanthosis, and hyperkeratosis. The patient and family opted for observation and cessation of kayaking. Follow-up at 2 years showed stable disease. A diagnosis of pachydermodactyly was made.**Conclusions:** Progressive, asymptomatic swelling of proximal interphalangeal joints, especially in young males, should prompt the consideration of pachydermodactyly as a potential differential diagnosis. Because of its benign course, correct recognition can help to prevent undue patient anxiety and avoid inappropriate investigation and treatment.**MeSH Keywords:** Asymptomatic Diseases • Fingers • Musculoskeletal Abnormalities**Full-text PDF:** <https://www.amjcaserep.com/abstract/index/idArt/923344>

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Background

Soft tissue swelling of the digits have a wide range of differential diagnosis, but inflammatory arthropathy is often the main concern. Pachydermodactyly, which is a rare but benign and self-limiting cause of cutaneous fibromatosis, usually occurs in young males, is an under-recognized condition [1–3]. A diagnosis of inflammatory polyarthropathy, especially polyarticular juvenile idiopathic arthritis, is often considered in the diagnosis of patients with pachydermodactyly. Being unaware of this benign entity can lead to undue anxiety and unnecessary treatment [4–6]. Here, we present a case of this little-known entity, which was recognized promptly and managed appropriately.

Case Report

A 16-year-old otherwise healthy adolescent male presented with a 2-year history of progressive, asymptomatic soft tissue swelling of the proximal interphalangeal joints (PIPJs) in multiple fingers bilaterally. He did not have morning stiffness, fever, rash, mouth ulcer, or uveitis. He remained active in sports with no loss of function reported. He participated in kayaking for the past 1–2 years. He reported no participation in contact sports, rock-climbing, or cycling. He engaged in routine use of a computer, but there was no history of excessive computer-gaming. He was not on any medication. There was no history of psychological problems or family history of polyarthrititis.

Physical examination revealed soft, saccular, subcutaneous swelling of the radial and ulnar aspects of the PIPJs of the ring, middle, and index fingers bilaterally, right more than left (Figure 1). No warmth, tenderness, joint laxity, or loss of range of motion were elicited, and there were no nail changes.

A plain radiograph revealed soft tissue swelling without loss of joint space, periarticular osteoporosis, periostosis, erosion, or osteophytes (Figure 2). An MRI showed T1 hypointense, mildly T2 hyperintense, and minimally enhancing cutaneous thickening, with no joint effusion, synovitis or tendinitis (Figure 3). Blood test results, including inflammatory markers, thyroid function test, insulin-like growth factor-1 (IGF-1), and autoimmune panel, were all normal.

A skin biopsy showed thickened collagen bundles in the dermis, acanthosis with thickening of the stratum spinosum, and hyperkeratosis with thickening of the stratum corneum. No significant deposition of mucin was detected. The patient and family opted for observation and cessation of kayaking. Follow-up at 2 years showed stable disease with stable painless skin thickening, blood test results were normal, with no evidence of arthropathy on repeat imaging.



Figure 1. Photograph of both hands shows saccular, soft tissue swelling of the radial and ulnar aspects of the proximal interphalangeal joints of the ring, middle, and index fingers bilaterally, right more than left.

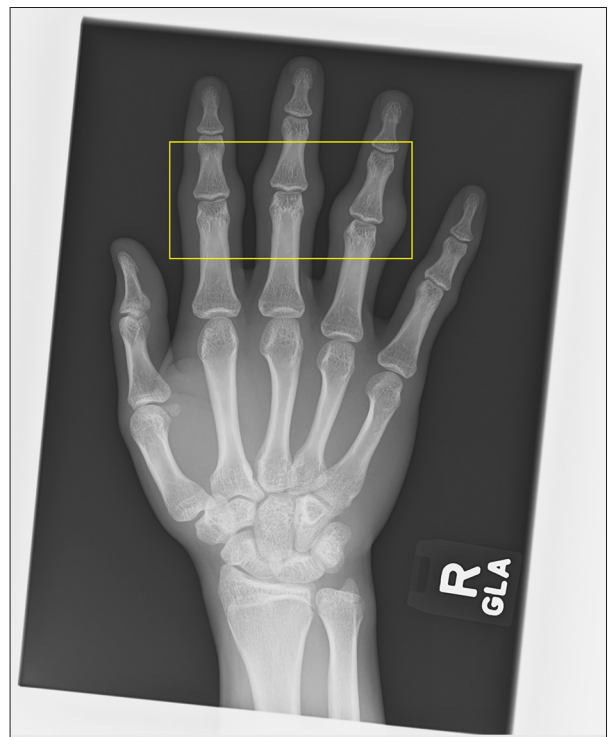


Figure 2. Radiograph of the right hand shows soft tissue swelling without loss of joint space, periarticular osteoporosis, periostosis, erosion, or osteophytes (yellow box).

Discussion

Pachydermodactyly (PDD), a term coined by Verbov in 1975 [7], is characterized by asymptomatic soft tissue thickening and cutaneous fibromatosis of the radial and ulnar aspect of the proximal interphalangeal joints (PIPJs). It typically affects the

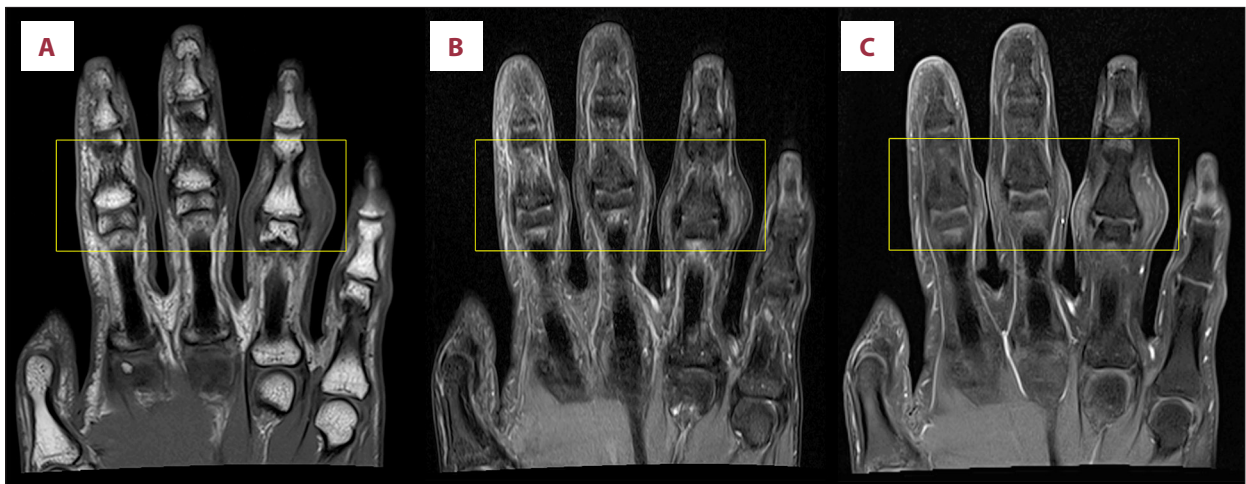


Figure 3. Coronal T1-weighted (A), coronal T2-weighted inversion recovery (B), and coronal T1-weighted post-contrast (C) MRI images of the right hand show saccular, T1 hypointense, mildly T2 hyperintense, and minimally enhancing cutaneous thickening around the proximal interphalangeal joints (yellow boxes). There is no evidence of arthropathy, with no joint effusion, synovitis, or bony involvement.

second through fourth digits bilaterally with occasional involvement of the fifth digit [1,2,8]. Findings are asymmetrical in one-third of patients. It usually affects otherwise healthy young male subjects, with a male-to-female ratio of 3.9 to 1 and a median age of 18 years old [1,6].

The etiology remains unclear, but repetitive and excessive mechanical irritation of the periarticular skin is thought to be the main contributing factor [3,9], and this was observed in our patient, who had a history of kayaking. Genetic predisposition and association with tuberous sclerosis or Ehlers-Danlos syndrome have also been described. However, PDD is usually an isolated sporadic occurrence [8].

The salient feature of PDD is absence of pain with no loss of function or involvement of the bone and joint, even in long-standing disease or progression of swelling [1,4]. Out of about 100 cases reported in the literature, there was only 1 case of deforming PDD with non-erosive subluxation of the interphalangeal joint reported, and this was deemed atypical [10]. A painless course with preserved joint function and normal blood test practically exclude inflammatory arthropathy such as juvenile inflammatory arthropathy [1,3,6].

Another close mimic is pachydermoperiostosis [11]. Pachydermoperiostosis classically presents as digital clubbing and skin thickening. The secondary form, caused by chronic systemic disease, is better known as hypertrophic osteoarthropathy [1,12]. Nonetheless, it is usually symptomatic with pain, involves the tubular bone, with prominent periostosis on radiograph, and shows increased uptake on bone scintigraphy [11,12]. Differentiation should trigger a clinical search for an underlying cause.

Other skin callosities of the hand and fingers that may resemble PDD are knuckle pads (also known as Garrod's pads in violinists) and pseudo-knuckle pads (also known as chewing pads in children). They tend to affect the extensor and dorsal surfaces of the fingers and hands rather than the radial or ulnar aspect in PDD, and they tend to be focal. However, some authors consider them as variants of PDD in the same spectrum of disease [1]. However, differentiation is not important, as they all run a benign course.

There is no universally accepted treatment. Most authors suggest that cessation of triggering activities may be sufficient. Administration of topical steroid or intralesional triamcinolone injection have been reported, with variable effects [1,2,8,13]. Surgical excision has been considered in selected cases for cosmetic purposes [14].

Our case demonstrates the typical clinical feature and demographics of PDD. Absence of abnormal blood test results and the classic appearance of saccular soft tissue swelling of the PIPJs without structural involvement of the joints on MRI further support the diagnosis. Prompt recognition enabled the clinician to provide appropriate reassurance to the patient and his parents, without subjecting him to inappropriate treatment. Avoiding misdiagnosis and recognizing this little-known but benign entity is the key teaching point of this case report.

Conclusions

Progressive, asymptomatic swelling of proximal interphalangeal joints, especially in young males, should prompt the consideration of pachydermodactyly as a potential differential

diagnosis. Because of its benign course, correct recognition can help to prevent undue patient anxiety and avoid inappropriate investigation and treatment.

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Conflict of interest

None.