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Case Report

Swollen digits due to pachydermodactyly resembling inflammatory arthritis

Ayda Acar[1], Fatmagül Dirican[1], Banu Yaman[2], İpek Tamsel[3], Gökhan Keser[4], Işıl Karaarslan[1]

Abstract

Pachydermodactyly is a rare digital fibromatosis, characterized by painless, fusiform swelling in proximal interphalangeal joints. Since it may be misdiagnosed as inflammatory arthritis, it is important to call physicians' attention to this rarely seen condition to avoid unnecessary immunosuppressive treatment. Hereby, we present a case of a pachydermodactyly that had been previously followed-up and treated as an inflammatory arthritis for 4 years. We describe the clinical, radiological, and dermoscopic features of the case.

Keywords: Painless joint swelling, dermoscopy, pachydermodactyly

Introduction

Pachydermodactyly is a digital fibromatosis, characterized by asymptomatic, non-inflammatory fusiform swelling of proximal interphalangeal (PIP) joints of fingers. Pain, tenderness, effusion, and erythema do not accompany joint puffiness (1). It is usually seen in male adolescents. In most of the cases, PIP joints of the second, third, and fourth digits are affected. The skin overlying the affected joint is thickened. A histopathological examination shows epidermal hyperplasia with hyperkeratosis and/or acanthosis, dermal collagen increase, fibroblast proliferation, and mucin deposition (1-3).

Although pachydermodactyly is a rare and benign dermatological pathology, recognizing this condition is very important. Otherwise, it may sometimes be misdiagnosed as inflammatory arthritis in the context of a rheumatologic pathology. Here we present a case of pachydermodactyly that had been misdiagnosed as juvenile rheumatoid arthritis and treated with hydroxychloroquine and acetylsalicylic acid previously.

Case Presentation

A 20-years-old male patient referred to our clinic with the complaint of nodulocystic acne. A medical history revealed that he had been diagnosed with juvenile idiopathic arthritis (JIA) 4 years ago based on the swelling of PIP joints in both hands, and he had been treated with hydroxychloroquine and acetyl salicylic acid since that time. However, the patient did not benefit from the treatment. When the symptoms of the joints were more specifically questioned, it was found out that the patient had an enlargement of the PIP joints since the age of 6, without any symptoms of inflammation such as joint pain, redness, or movement limitation. There was no joint disease in his family history. There was no history of morning stiffness in the hands; besides, there was no sign of systemic inflammation indicator in the previous blood tests and no evidence of rheumatologic joint disease in the previous radiologic examinations. On the whole body examination, fusiform swelling and thickening of the overlying skin of the PIP and metacarpophalangeal (MCP) joints of both hands were seen. The involvement was more prominent in the right hand. Also, yellowish-brown-colored hyperkeratotic plaques on the medial parts of both knees, and lateral and medial malleols (Figure 1-3), were observed. The skin overlying the PIP joints was examined via dermoscopy, and a cobblestone pattern and whitish scale were observed (Figure 4). In the blood tests and liver and kidney function tests, complete blood count, erythrocyte sedimentation rate, and antistreptolizin o (ASO) and C reactive protein values were within normal limits. Rheumatoid factor, antinuclear antibody (ANA), and ANA profile were negative. The case was referred to the rheumatologist, with suspected pachydermodactyly. There was not any bone or joint pathology on the hand X-ray (Figure 5). Sonographic findings such as synovial effusion or hypertrophy were also absent in the PIP joint (Figure 6). Rheumatologic pathology was excluded with the clinical and laboratory findings. The observed hyperkeratotic epidermis, papillomatosis, and dermal fibroblast increase on the histopathology of the overlying skin of the PIP joint confirm the diag-

ORCID IDs of the authors:
A.A. 0000-0001-7507-6999;
F.D. 0000-0002-3992-6738;
B.Y. 0000-0002-2915-0788;
I.T. 0000-0003-3629-2386;
G.K. 0000-0001-7289-1816;
I.K. 0000-0001-5348-027X.

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[1] Department of Dermatology, Ege University School of Medicine, İzmir, Turkey
[2] Department of Pathology, Ege University School of Medicine, İzmir, Turkey
[3] Department of Radiology, Ege University School of Medicine, İzmir, Turkey
[4] Department of Rheumatology, Ege University School of Medicine, İzmir, Turkey

Address for Correspondence:
Ayda Acar, Department of Dermatology, Ege University School of Medicine, İzmir, Turkey

E-mail: aydaerbas@yahoo.com

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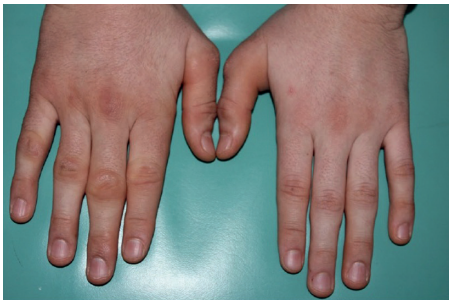

Figure 1. Yellowish-brown-colored hyperkeratotic plaques on the medial parts of both knees, and lateral and medial malleols

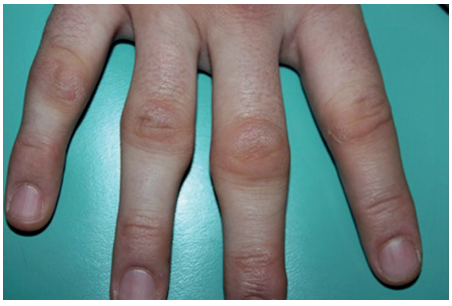

Figure 2. Yellowish-brown-colored hyperkeratotic plaques on the medial parts of both knees, and lateral and medial malleols

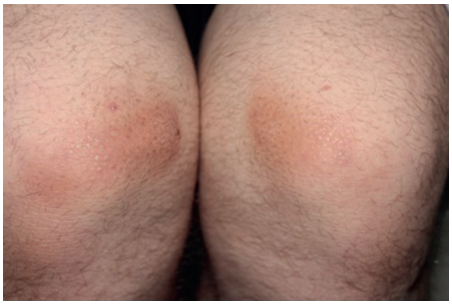

Figure 3. Yellowish-brown-colored hyperkeratotic plaques on the medial parts of both knees, and lateral and medial malleols

nosis of pachydermodactyly (Figure 7, 8). The cessation of the hydroxychloroquine and acetyl salicylic acid treatment and re-examination of the case 3 months later was suggested by the rheumatologist. The case was re-examined both by the dermatologist and rheumatologist after 3 months. The patient had no complaints of pain and increased swelling after the drugs were discontinued. The initial complaint of the patient was the acneiform lesions on his face; this condition was managed separately, and appropriate treatment was recommended. Written informed consent was obtained from the patient who agreed to participate in this study.

Discussion

Pachydermodactyly was first described in 1973 by Bazex as fibromatosis and hyperplastic dermis of digits II-IV (4). Verbov named this clinical condition pachydermodactyly in 1975 (5).

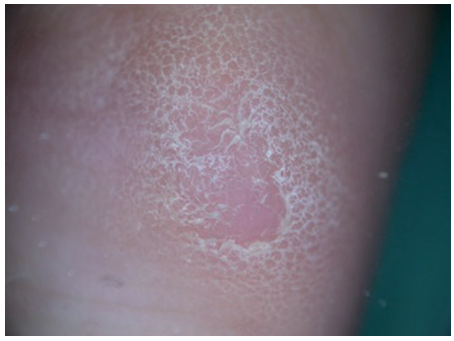

Figure 4. The skin overlying the PIP joints has examined via dermoscopy, and a cobblestone pattern and whitish scale

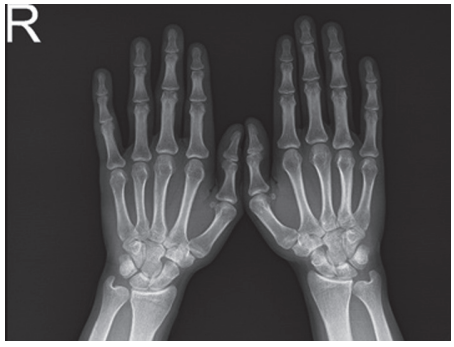

Figure 5. There was not any bone or joint pathology on the hand X-ray

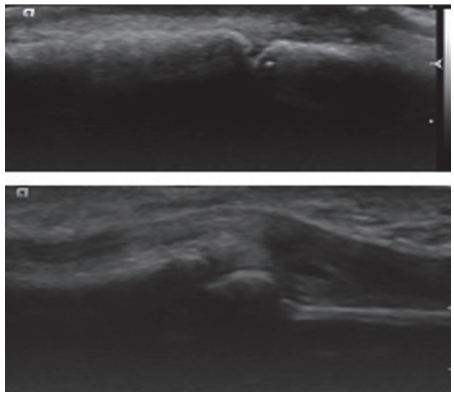

Figure 6. Sonographic findings such as synovial effusion or hypertrophy were also absent in the PIP joint

Since those first descriptions, nearly 160 cases have been reported in the literature.

In the classical form, the involvement of PIP joints of the hand is common. In addition to this classical form, the cases with distal interphalangeal or MCP involvement, as well as the cases with the toe, knee, or planter region involvement have also been reported (2, 3, 6, 7). Although our case is consistent with the classical form, yellowish-brown-colored hyperkeratotic plaques on the knees were also accepted as the extension of the disease.

The exact etiology of pachydermodactyly is unclear, but repetitive mechanical skin stimulation

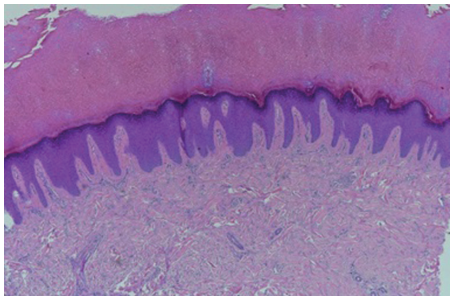

Figure 7. Hyperkeratotic epidermis, papillomatosis, and dermal fibroblast increase on the histopathology of the overlying skin of the PIP joint confirm the diagnosis of pachydermodactyly

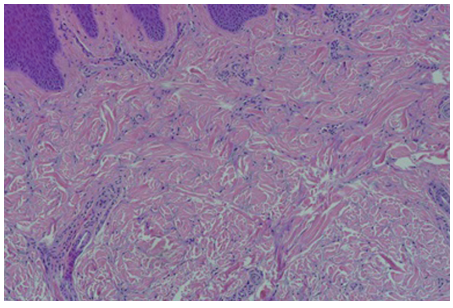

Figure 8. Hyperkeratotic epidermis, papillomatosis, and dermal fibroblast increase on the histopathology of the overlying skin of the PIP joint confirm the diagnosis of pachydermodactyly

has been suggested as a triggering factor (3). In our case, no history of repetitive trauma was found.

Some familial cases have also been described. In some other reports, associations such as tuberous sclerosis, Ehler Danlos syndrome, and atropia maculosa varioliformis have been reported (8, 9). There was no such an association in the present case.

To the best of our knowledge, there is only a single report in the literature describing dermoscopic features of pachydermodactyly as a cobblestone pattern and whitish scale corresponding to papillomatosis and hyperkeratosis (10). Dermoscopic features in our case were concordant with those reported findings.

Pachydermodactyly is diagnosed by the presence of typical clinical findings and the absence of bone and joint involvement on the radiological imaging and inflammation clues in the blood tests. In the present case, with the clinical, radiologic, and laboratory findings, the diagnosis of pachydermodactyly and ruling out of a rheumatologic condition was possible.

There have been few reports in the literature about the treatment of pachydermodactyly including intralesional steroid injection, surgical resection (6), and tranilast (2). Since our patient was pleased to learn that he had no rheumotologic disease, he did not ask for any treatment.

Conclusion

In conclusion, pachydermodactyly should be considered in the differential diagnosis of inflammatory arthritis. The correct diagnosis will spare the patient unnecessary systemic immunosuppressive treatments.

Informend Consent: Written informed consent was obtained from the patient who participate in this study.

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References

1. Kutzner HH, Kamino H, Reddy VB, Pui J. Fibrous and fibrohistiocytic proliferations of the skin and tendons. Bolognia JL, Shaffer JV, Cerroni L, editors. Dermatology. 4th ed. China; Elsevier; 2018.p.2068-85.
2. Higuchi C, Tomita T, Yoshikawa H. Pachydermodactyly treated with tranilast in a young girl. Case Rep Orthop 2014; 2014: 132854. [CrossRef]
3. Dallos T, Oppl B, Kovács L, Zwerina J. Pachydermodactyly: a review. Curr Rheumatol Rep 2014; 16: 442. [CrossRef]
4. Bazex A, Dupre A, Teillard J. Pachydermie digitale des premières phalanges par hyperplasie conjonctive dermique et aplasie hypodermique. Bull Soc Fr Dermatol Syphiligr 1973; 80: 455-8.
5. Verbov J. Letter: Pachydermodactyly: a variant of the true knuckle pad. Arch Dermatol 1975; 111: 524. [CrossRef]
6. Dias JM, Costa MM, Romeu JC, Soares-Almeida L, Filipe P, Pereira da Silva JA. Pachydermodactyly in a 16-year-old adolescent boy. J Clin Rheumatol 2012; 18: 246-8. [CrossRef]
7. Vale LR, Coeli FR, Michalany N, Hassun KM, Porro AM. Transgrediens pachydermodactyly: report of a case. An Bras Dermatol 2009; 84: 190-3. [CrossRef]
8. Bardazzi F, Neri I, Fanti PA, Patrizi A. Pachydermodactyly in two young girls. Pediatr Dermatol 1996; 13: 288-91. [CrossRef]
9. Callot V, Wechsler J, Hovnanian A, Revuz J. Pachydermodactyly and atrophia maculosa varioliformis cutis. Dermatology 1995; 190: 56-8. [CrossRef]
10. Zabotti A, Errichetti E, Cereser L, Pegolo E, Quartuccio L, Stinco G, et al. Pachydermodactyly: the role of ultrasonography and dermoscopy for diagnosis. Rheumatology (Oxford) 2017; 56: 703.