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A Rare Case of Pachydermodactyly in 25 years Old Male

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Abstract : Pachydermodactyly is a rare digital fibromatosis that can be misdiagnosed with inflammatory rheumatic diseases. So far in the literature only about 150 cases of PDD have been reported. A prompt clinical diagnosis of the disease would prevent inappropriate treatment and unnecessary expensive diagnostic procedures such as biopsy or magnetic resonance imaging. Gold standard therapy for pachydermodactyly is not established yet. A rare case of a 25 years-old male patient with pachydermodactyly is reported. He complained swelling and thhickening around joints in both hands since 7 months ago. He denied having any pain or morning stiffness of the joints and was also free other symptomatic symptomps. Family history was not significant. Dermatological state showed skin colouredhyperkeratotic patches. Laboratory showed no abnormalities. Radiology results shows no bone and joint abnormalities. Dermoscopy examination on lesion shows whitish scaling and cobblestone appearance. Diagnosis of pachydermodactyly is based on anamnesis, physical examination, laboratory findings, and plain radiograph. Early and correct diagnosis is important to prevent unnecessary, expensive, and invasive diagnostic procedure.
Key words: pachydermodactyly, rare case.

Introduction

Pachydermodactyly (PDD) is a rare and benign form of digital soft tissues fibromatosis, which affects the skin of the fingers. The disorder is characterized by asymptomatic, symmetric, progressive soft tissue swelling of the proximal interphalangeal (PIP) joints of fingers II–IV and rarely V. In some cases, the process of the expansion to the metacarpophalangeal joints (MCP) is observed, which is defined as transgredienspachydermodactyly.[1]

In PDD, there are no lesions in the bones and periosteum in an X-ray examination. Also in synovium and joints in ultrasound examination no alterations are observed.The disorder was first described by Basex et al. in 1973, and named by Verbov in 1975: pachy – thick, dermos – skin, dactylos – fingers.[2] So far in the literature

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only about 150 cases of PDD have been reported. Given its rarity, pachydermodactyly is a diagnostic problem for many medical specialities.[3]Some authors assume it to be the result of a habitual behavior, whereas others consider there to be a genetic predisposition.[4]

The etiology of the disease remains unknown, PDD is usually acquired, even though there are some publications that document family cases. It affects adolescent men. It has been suggested that multiple mechanical traumas, such as the habit of interlacing and rubbing the fingers, could lead to skin thickness, what causes secondary, mechanical dermatosis based on compulsive behaviours. The classic type of disease can even simulate polyarticular JIA with involvement of small joints of the hands.[5]

The diagnosis of PDD is usually a clinical one supported by radiological imaging whilst laboratory investigations and autoimmune screening are generally unremarkable.[6] Radiography, ultrasonography and MRI demonstrate soft tissue swelling with no bone abnormality. Whilst some authors state that histology is not necessary to confirm a diagnosis of PDD, the characteristic histological features of PDD include an increase in dermal collagen with varying degrees of hyperkeratosis and acanthosis.[7] Interestingly, collagen analysis is consistent with structure types III and V which differs to the collagen profile of normal skin and electron microscopy showed collagen fibres to be smaller and less uniform.

Treatment of pachydermodactyly consists of rehabilitation, which allows one to stop the progression or to reduce pathological changes. The beneficial effect of subcutaneous injections of glucocorticoids has been reported. Because of the important role of mechanical trauma, stopping any compulsive mechanical skin irritation is recommended. Psychological support and, in some cases, psychiatric therapy are necessary. Differential diagnosis of pachydermodactyly should consider atypical form of the disease, including one-sided occurrence and the possibility of the overlap syndrome with other connective tissue diseases.[8]

Case Report

A 25 years-old woman was referred from Painan Government health Center to dermatology outpatient clinic M Djamilpadang on 11th, February 2019 with suspect Ganglion Cyst.There are swelling on joint both hands since 7 months ago Initially, 7 month ago, there is swelling on joint of the thumbs of left hand, the patient felt no pain nor itchy.The swelling spread to other joint of fingers and both hand.Patient has habit biting fingers and pick fingers with nail clippers.

Patient seek medication to General Practitioner on local clinic and was given Salf and medication that was applied twice a day, but the patient felt no improvement.The patient stop applying medication after a month.Patient never seek other medical help for this symptomps.

Joint paint on both hands other body part was denied. Morning stiffness on both hands and other joint was denied. Movement difficulties was denied.There was no history of applying topical traditional medication. History of trauma before was denied. There was no history of fever

There was no history of losing body weight on the body in a few months or bumps on the body. Patient had no history of swelling on any joint, morning stiffness, fever, and movement difficulties.There was no family history of swelling on joint, morning stiffness, fever, and movement difficulties.

Dermatologic state found found skin coloured hyperkeratotic patches on all fingers on both hands. Dermoscopy examination shows whitish scale and cobblestone appearance. Radiology examination show no bone and jointabnormalities

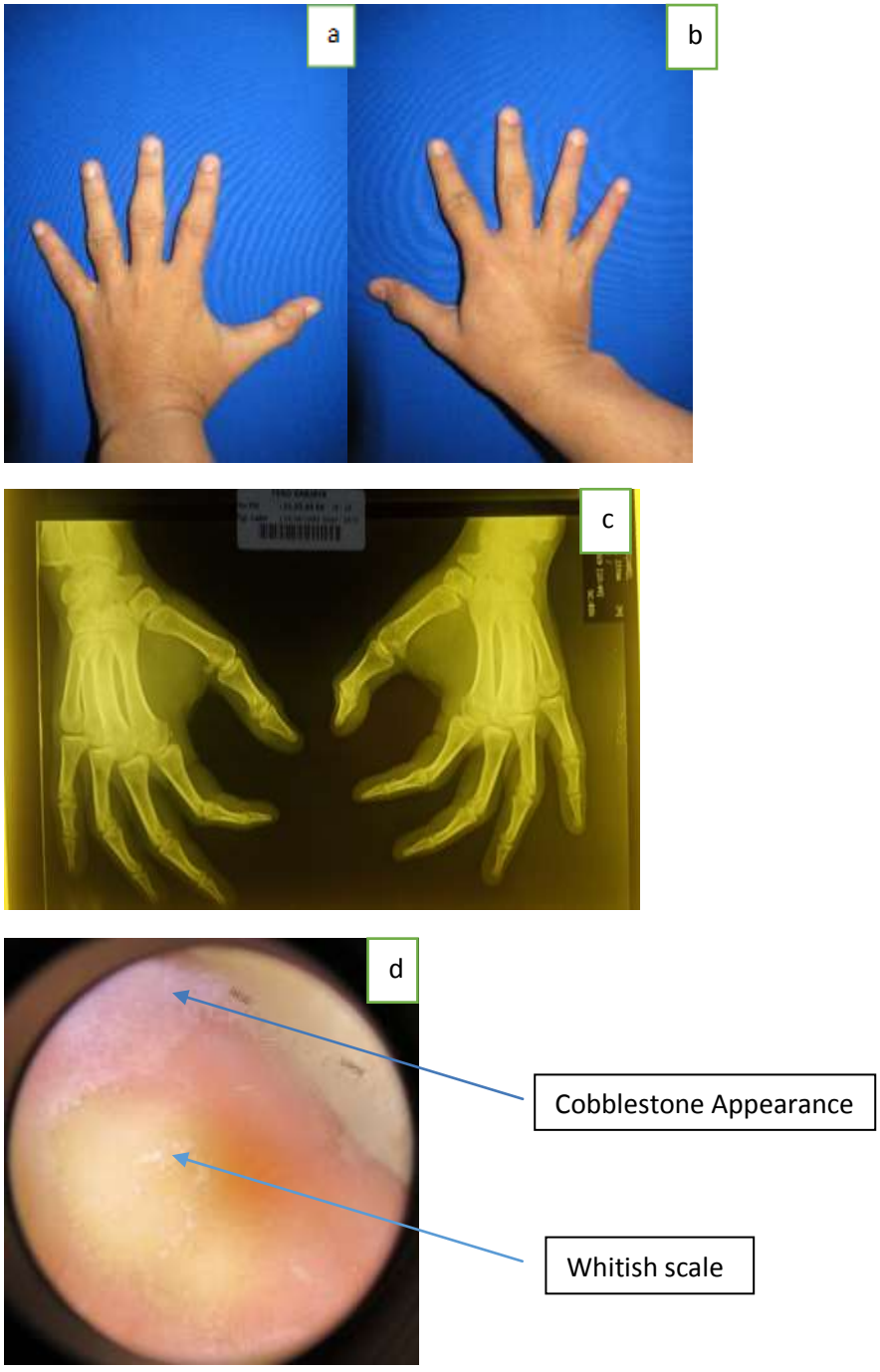

Figure 1. (a)-(b) lesion on lateral aspect of both hands (c) Plain radiography shows no joint and bone abnormality (d)dermoscopy shows whitish scale and cobblestone appeareance

Discussion

We reported a case of Pachydermodactyly in 25 years old man. The diagnosis was made based on anamnesis, physical examination, radiograph findings and laboratory findings. In this patient, there were swelling on joint on both hands with no joint paint nor itchy since 7 month ago.

Pachydermodactyly is a rare, benign disorder of the skin characterized by a fusiform swelling of the soft tissue at the proximal interphalangeal joints in the hands of young males.[1] In literature, only 150 cases is reported worldwide. There is no pain or restriction of motion. Hyperkeratosis of the affected areas is a common feature of pachydermodactyly. [2]

When the disease extends to the palms or other extremities are involved, it is called pachydermodactyly

transgrediens. In our case, the patient presented with bilateral involvement of the proximal and distal interphalangeal joints of all fingers on both hands. Since the initial description in 1973, the etiology of pachydermodactyly remains unknown.[3] Although a familial inheritance has been suggested, most often it is considered to result from repeated mechanical trauma, reaction to emotional stress,or as a habitual behavior with over-rubbing of the hands and fingers. Based on this behavior, misdiagnosis is common and the patient is often referred to a psychiatrist for consultation and treatment.[4] In our case, this patient has no familial history with pachydermodactyly. The patient has habit of biting fingers and picking fingers with toenail.

Misdiagnosis as arthritis and knuckle pad is also common; however, the typical thickening of the skin in addition to the absence of pain and morning stiffness, synovitis on clinical examination, and typical serologic findings should allow differentiation of inflammatory arthropathy from pachydermodactyly.[5] In this case, patient has no morning stiffness, no pain, no movement difficulties. Despite the rarity of the disease, Bardazzi et al. proposed a classification of pachydermodactyly into different forms including the classical pachydermodactyly, localized or monopachydermodactyly, pachydermodactyly transgrediens (extension to the palms or proximal fingers), familial pachydermodactyly, and pachydermodactyly associated with tuberous sclerosis.[6] This patients had classical form of pachydermodactyly.

We consult to internal medicine department for suspect rheumatoid artrhritis, and the patient was tested for Rheumatoid Factor, which came out negative result. The patient got metil prednisolone, Na diclofenac and lansoprazole. The patient felt no improvement after took medication for just few days, and refused to continue treatment.

The diagnosis of pachydermodactyly is usually clinical. Routine laboratory investigations are normal, as the disease is noninflammatory. Imaging shows soft tissue swelling of the involved interphalangeal joints without any bone or joint involvement.[6] Routine laboratory on this patients shows no abnormalities and x ray examination shows no bone and joint abnormalities.

Biopsy and histological examination can further support the diagnosis, although this is not necessarily required in all cases.Typical histological findings of pachydermodactyly are dermal hyperkeratosis, acanthosis, an absence of inflammatory cells, and in some cases mucin deposits. Collagen type III and IV fibers are present in the involved sites and differ in fiber diameter in comparison with normal sites. According to Rancy,et al. and Ulusoy et al, routine biopsy is not indicated unless more sinister diagnoses are under consideration.[7]

Zabotti et al, in 2017 studied the role of dermoscopy in pachydermodactyly diagnosis. They found Whitish Scaling and Cobblestone appeareance in dermoscopy examination on pachydermodactyly patient.[4]

The disease is usually benign and nonprogressive; therefore, symptomatic treatment alone is all that is usually required.[8]. In patients with progressive pachydermodactyly, with severe deformity and functional impairment of the hands, surgical treatment may be performed, focusing on the excision of the subcutaneous hypertrophic soft tissue of the joints. Alternatively, infiltration with triamcinolone acetonide may lead to regression of symptomatic lesions.[9]

In our case, the patient was planned to be treated with triamsinolon injection, and was encouraged to stop biting fingers and picking fingers with nail clippers. Psychological support and, in some cases, psychiatric therapy are necessary.[10] In this patient, DLQI Score is 14, meaning this disease has large effect on patients life, despite benign nature of pachydermodactyly. This patient need pscychiatric support, unfortunately the patient shows reluctance to follow therapy and consultation procedure.

Conclusion

- We reported a case A 25 years-old man with Pachydermodactyly,
- Diagnosis was made by anamnesis, physical examination, radiology examination, and dermoscopy.
- Tha patient was encourage to minimize hand trauma and planned to get Intralesional triamsnilonon injection

References

1. Kavecan I, et al. Pachydermodactyly: A rare Type of Macrodactyly as a Dermatological Sign of Compulsive Behaviour And Repetitive Minor Trauma, Iran Red Crescent Med J. 2018 : 20 (9) e 79992.
2. Chu H et al. Pachydermodactyly : A Benign Cutaneus Condition That May be Misdiagnosed as a Joint Disorder. J Rheumatol 2016.43 1615-1616.
3. Abdurrahman W,:Acase of Pachydermodactyly in seventeen years old associated with Repetitive Minor Trauma. . Dermatology Online Journal22 (8) :15.
4. Zabotti A et al. :Pachydermodactyly: The Role of Ultrasonography and dermoscopy for diagnosis. Oxford Journals.2017 :56 : 703.
5. Eduardo et al, A Novel Treatment for Knuckle Pads with intralesional florouracil. Arch Dermatol/vol 143 2017.
6. Chen C et al: Pachydermodactyly : Three new cases in Taiwan. Journal of Microbiology Immnuology and Infection (2012).
7. Tolis K et al: Pachydermodactyly : An Unknown Entity .J Hand Surgery am. 2016**38**(2):91-95, 1999.
8. Rency R et al: Pachydermodactyly : A case Report Including Histopathology. J Hand Surgery Am 2016.
9. Zuher Z et al: Pachydermodactyly A report of Two Cases. *Rheumatologia,2016*:54.3 :136.140.
10. Mendis,et al. Pachydermodactyly- A Rare Presentation of Digital Fibromatosis . Sri lanka Journal of Dermatology, 2017:19:42-43.

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