Introduction

Pachydermotactylly (PDD) is a benign digital fibromatosis, usually involving the proximal interphalangeal (PIP) joints of the hands. A 14.5-year-old male patient was assessed for deformity on his fingers progressing in the last five years and tic disorder in the last eight years. Physical examination revealed that his bilateral 2nd, 3rd, and 4th PIP joints were painless, swelling on lateral sides. On magnetic resonance imaging, the thickening predominantly involved the tissue around the same PIP joints. Other laboratory investigations were normal. Because of the involvement of small joints of the hands, the symptoms can be confused with inflammatory rheumatic disease, especially with juvenile idiopathic arthritis and rheumatoid arthritis. In this paper, we address pachydermotactylly to assist rheumatologists in making the diagnosis of this disease correctly.

Key words: Pachydermotactylly, juvenile, idiopathic arthritis

Abstract

Pachydermotactylly (PDD) is a benign digital fibromatosis, usually involving the proximal interphalangeal (PIP) joints of the hands. A 14.5-year-old male patient was assessed for deformity on his fingers progressing in the last five years and tic disorder in the last eight years. Physical examination revealed that his bilateral 2nd, 3rd, and 4th PIP joints were painless, swelling on lateral sides. On magnetic resonance imaging, the thickening predominantly involved the tissue around the same PIP joints. Other laboratory investigations were normal. Because of the involvement of small joints of the hands, the symptoms can be confused with inflammatory rheumatic disease, especially with juvenile idiopathic arthritis and rheumatoid arthritis. In this paper, we address pachydermotactylly to assist rheumatologists in making the diagnosis of this disease correctly.

Key words: Pachydermotactylly, juvenile, idiopathic arthritis

Introduction

Pachydermatocytly (PDD) is a benign digital fibromatosis, usually involving the proximal interphalangeal (PIP) joints of the hands [1]. It is clinically characterized by asymptomatic, symmetrical, and fusiform swelling of the second to fourth PIP joints of both hands [1]. Because of the involvement of small joints of the hands, it can be confused with inflammatory rheumatic disease, especially with juvenile idiopathic arthritis and rheumatoid arthritis. Awareness of pachydermotactylly would assist rheumatologists in making diagnosis of this disease correctly.
Case Report

A 14.5-year-old male patient was referred to our rheumatology clinic for assessment of deformity on his fingers progressing in the last five years. There was no morning stiffness, fever, rashes, mouth ulcer, uveitis, cardiovascular, or gastrointestinal symptoms but tic disorder for the last eight years. When he was angry or anxious, he would chew the second fingers. He had been on epilepsy treatment for two years with levasitaretam. Family history was normal. Physical examination revealed that his bilateral 2nd, 3rd, and 4th PIP joints were painless and swelling on lateral sides (Figure 1). His hemogram, erythrocyte sedimentation rate, C-reactive protein, rheumatoid factor, antinuclear antibodies, thyroid functions, blood lipids and uric acid levels were normal. X-rays exams only revealed augmentation of soft parts in the PIP without joint narrowing or bone erosions (Figure 2). On magnetic resonance imaging, the thickening was seen predominantly on the tissue around the 2nd, 3rd, and 4th PIP joints (Figure 2). The patient was given local hand care and referred to pediatric psychiatry.

Discussion

PDD was described by Bazex et al. in 1973. Two years later, Verbov coined the word pachydermodactyly [1], a non-inflammatory disease characterized by fusiform soft-tissue swelling of the PIP joints, especially on the lateral sides of the joints. The etiology of PDD is unknown, mechanical injury, often caused by a psychiatric or neurological disorder, can be inferred in 35-65% of the published cases [2,3]. Almost all of the patients have asymptomatic bilateral involvement of the second to fourth PIP joints of the hands. The progress of the disease may include increased dermal accumulation of collagen. PDD may also be seen with occupational exposure, Asperger syndrome or obsessive compulsive disorders [4]. Our patient had a history of behavioral and neurological problems.

Bardazzi et al. [5] suggested classifying PDD into five main types: 1) classic pachydermodactyly, both idiopathic and associated with trauma, and primarily described in male patients with several affected fingers; 2) mono or localized pachydermodactyly, which may also be idiopathic or induced.
trauma; 3) transgradient pachydermodactyly, in which cutaneous changes may extend to the palms, particularly to the metacarpophalangeal joints; 4) familial pachydermodactyly; 5) pachydermodactyly which is associated with tuberous sclerosis and may sometimes be painful [5]. Our patient may be classified as type I based on his clinical presentation.

Chen et al. [6] reported that PDD diagnosis criteria are as follows: 1) the patient has no symptoms; 2) morning stiffness is absent; 3) pain on motion and tenderness to palpitation is absent; 4) finger swelling is radial or ulnar in location, rather than circumferential; 5) laboratory test results are unremarkable; 6) plain radiographs show only soft tissue swelling. Pachydermodactyly must be considered in the differential diagnosis of juvenile idiopathic arthritis, because it could be mistaken for the polyarticular form. Differential diagnoses, most of which could be clinically differentiable, include Thiemann’s disease, juvenile fibromatosis, Garrod’s pads, tophi, xanthomatous deposits, pachydermoperiostosis and paraneoplastic acropachydermodactyly.

Thiemann’s disease (hereditary necrosis of epiphysis) has an initially similar clinical appearance but shows progression with painful deformity of the fingers and limitations in function, and X-rays are sometimes helpful for differential diagnosis [2]. Another diagnosis to rule out would be knuckle pads in which lateral sides of finger are usually not affected. The lesions are usually skin-colored and freely movable [7]. Pseudo-knuckle pads may be considered as a form of collosity that appears after repeated trauma. This type of knuckle pads has been described in children with obsessive behavior as chewing pads and in adults as occupational disorder. The lesions disappeared after rubbing was stopped [7]. Although our patient has chewed mostly the second fingers, the lateral sides of other fingers have been affected too.

There is no effective medical treatment for PPD. Skin care education, psychotherapy for those who have obsessive–compulsive behavior are provided [6]. Intraläsional corticosteroids may be administered and localized subcutaneous resection in selected cases may be an effective cosmetic option [8]. Long-term prognosis of PDD is not known.

An awareness of PDD would assist the rheumatologist in making the diagnosis of this confusing disease. A prompt diagnosis would avoid an unnecessary search, relieving patient’s and relatives’ anxiety and sparing the patient from inappropriate treatment with steroids or cytotoxic agents.

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**References**