Other full case

A 12-year-old boy presenting with unilateral proximal interphalangeal joint swelling

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Summary

Pachydermodactyly is a rare and benign superficial dermal fibromatosis, characterised by asymptomatic soft tissue hypertrophy of the lateral and dorsal aspects of the proximal interphalangeal joints. The majority affected are males, with only a few reported female cases, and usually begins to develop around the age of puberty. Pachydermodactyly affects the second, middle and ring fingers of the hand symmetrically and bilaterally. The majority of reported cases demonstrate bilateral involvement of joints, only a small proportion report unilateral involvement. In this case report the authors review literature and discuss a case of pachydermodactyly presenting a unilateral swelling of the proximal interphalangeal joints of only the left hand in a right-handed young male.

BACKGROUND

This case report highlights the case of unilateral involvement of pachydermodactyly, and the need to consider it as a differential in a child presenting with swelling in the region of the proximal interphalangeal joints. This condition may mimick juvenile idiopathic arthritis. Infact the patient was referred to the paediatric rheumatology clinic to rule out juvenile idiopathic arthritis.

CASE PRESENTATION

We report the case of a 12-year-old Caucasian boy who was referred to the paediatric rheumatology clinic with suspected diagnosis of juvenile idiopathic arthritis. He presented with a 1-year history of swelling of the proximal interphalangeal joints of his left ring and middle fingers. Over the course of the next year, the swelling was noted to increase in size. He had no pain or functional limitation of movement. The main problem associated with the swelling was the friction between the adjacent fingers resulting in blistering. There were no other joint symptoms and no systemic symptoms.

He was not on any regular medication being otherwise fit and healthy with no significant medical history. There was no family history to note including no history of similar swellings. There were no reported behavioural problems. It is important to note behavioural problems, especially in the form of obsessive compulsive behaviour as this has been demonstrated in previous case reports as a contributing aetiological factor.¹ The patient is right-handed and his parents commented that he spends a lot of time playing on games consoles and typing on keyboards with this being a likely aetiological factor due to the risk of skin rubbing during these actions. It is believed by many that repetitive trauma in the form of skin rubbing is involved in the aetiology of both pachydermodactyly and knuckle pads.

On examination there is diffuse thickening of the soft tissues around the proximal interphalangeal joints of the left middle and ring finger, with no other joints involved

(figures 1 and 2). There is a full range of movement of these joints and the other joints of the hand with no other abnormality detected on examination. There were no signs of infection or inflammation of the joints involved. Systemically he looked well.

INVESTIGATIONS

Blood tests were undertaken including full blood count, biochemistry, C reactive protein, bone profile, vitamin D and parathyroid hormone levels all of which were within normal limits. The autoimmune screen, rheumatoid factor and antinuclear antibody factor were negative.

Ultrasound scan of the affected fingers showed that the swellings represented hypertrophy of the soft tissues around the proximal interphalangeal joints of the left middle and ring finger with no abnormality of vascularity within the thickened tissue. There was normal appearance of the flexor and extensor tendons, and no ultrasound evidence of inflammatory synovitis or arthropathy. Plain radiographs of the affected hand showed soft tissue swelling, with no bony abnormality. MRI scan and biopsy were planned but was not done as the diagnosis was obvious on clinical examination and ultrasound findings.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis includes knuckle pads, juvenile idiopathic arthritis, tuberous sclerosis, post-traumatic callosities, granulomas, fibromas, infantile digital fibromatosis and rheumatoid nodules.

TREATMENT

The prognosis of pachydermodactyly is variable. A proportion of cases of pachydermodactyly have been seen to resolve by adult years, although in idiopathic forms of the disease this is not usually the case. The management is usually in the form of counselling the patient and prevention of further repetitive rubbing of the skin. Local corticosteroids have been tried in treatment previously,

Figure 1 Photograph of dorsal aspect of both hands showing diffuse swelling of the lateral aspect of the left third and fourth proximal interphalangeal joints.

but are felt to be ineffective providing no effective topical treatment. There is conflicting evidence for the use of intralesional steroids with some papers reporting reduction in the swelling and cosmetic improvement. Cryotherapy and 50% phenol, as well as surgical excision have been used in treatment; however, reoccurrence of the swelling is common.¹

OUTCOME AND FOLLOW-UP

He was reviewed by the Paediatric Orthopaedic team who felt that as he had no functional limitation or pain, there was no role for surgical intervention.

The patient was managed conservatively and after 2 year regular follow-up he remains stable without any complications or functional limitation or the need for any significant intervention.

DISCUSSION

Pachydermodactyly was first reported in 1973 by Bazex *et al*² when they noted hypertrophy of the lateral aspects of the proximal phalanges which they could not classify into the previously known syndromes, consequently leading them to suggest the possibility of a new condition. In 1975, Verbov³ described pachydermodactyly as a variant of true knuckle pads after studying a 19-year-old male with asymptomatic diffuse swellings over the dorsal and lateral aspects of the proximal interphalangeal joints of some fingers. Literature review reveals approximately 90 previously reported cases of pachydermodactyly predominantly in adults, with only a small number reporting unilateral involvement, and in children.¹

Pachydermodactyly is an acquired disease of unknown aetiology in the majority of cases. Aetiology is thought to be related to repeated trauma to the skin, and previously

the suggestion of a background of behavioural problems including obsessive compulsive behaviour has been raised as a contributing factor. In our reported case, the patient is right-handed and playing video games and typing requiring both the hands, could not easily result in swelling of the third and fourth left proximal interphalangeal joints. He had no behavioural problems. We suspect that causes other than repetitive trauma might trigger a local swelling of the tissues around the finger joints. There have been previous case reports describing association with Dupuytren's disease, carpal tunnel syndrome, gynaecomastia, juvenile chronic arthritis, tuberous sclerosis, thyroid dysfunction, Ehlers-Danlos syndrome and atrophica maculosa varioliformis.^{1 4 5}

The differential diagnosis of pachydermodactyly includes that of knuckle pads. Knuckle pads also known as Garrods' Pads, are benign, asymptomatic, well-circumscribed digital fibrosis of the dorsal aspect of the metacarpophalangeal and proximal interphalangeal joints. They are felt to have similar aetiology and are often related to a history of repetitive trauma. Knuckle pads are noted in equal prevalence between genders and more commonly seen in adults over the age of 40 years. In comparison pachydermodactyly affects adolescents, and the majority affected are male.^{1 5}

Knuckle pads are thought to be an epidermal response where as pachydermodactyly is a dermal response and they can be differentiated histologically. In knuckle pads there is hyperkeratosis, orthokeratosis and acanthosis and a dermal inflammatory infiltrate. Pachydermodactyly does not demonstrate orthokeratosis; instead there is hypergranulosis and thickening of the dermal layer. There is an increase in fibroblasts and collagen types I, III and V extending into the subcutaneous tissue, varying from that of the normal skin profile.^{4 6}

Figure 2 Photograph of the left hand showing diffuse swelling of the lateral aspect of the third and fourth proximal interphalangeal joints.

Learning points

- ▶ Pachydermodactyly is an acquired disease of unknown aetiology presenting as swelling over the dorsal and lateral aspect of the interphalangeal joints.
- ▶ This condition may mimic juvenile idiopathic arthritis affecting the interphalangeal joints.
- ▶ In managing a child with pachydermodactyly it is important to address the contributing factors such as repetitive friction of the skin, and underlying behavioural issues.

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Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Beltraminelli H**, Itin P. Pachydermodactyly—just a sign of emotional distress. *Eur J Dermatol* 2009;**19**:5–13.
2. **Bazex A**, Dupré A, Teillard J. Pachydermie digitale des premières phalanges par hyperplasie conjonctive dermique et aplasie hypodermique. *Bull Soc Fr Dermatol Syphiligr* 1973;**80**:455–8.
3. **Verbav J**. Letter: Pachydermodactyly: a variant of the true knuckle pad. *Arch Dermatol* 1975;**111**:524.
4. **Balassiano V**, Sodre CT, Ramos-e-Silva M. Pachydermodactyly: report of a case. *J Eur Acad Dermatol Venereol* 1996;**6**:260–3.
5. **Mansouri P**, Safaai Naraghi Z, Seirafi H, *et al*. Pachydermodactyly: report of two cases. *Acta Medica Iranica* 1999;**37**:232–6.
6. **Morginson WJ**. Discrete keratodermas over the knuckle and finger articulations. *AMA Arch Derm* 1955;**71**:349–53.

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