PACHYDERMODACTRYLY - ROLE OF LOCAL CORTICOTHERAPY

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PACHYDERMODACTYLY- ROLE OF LOCAL CORTICOTHERAPY (Abstract): Pachydermodactyly is a rare skin disease, defined as a benign form of digital fibromatosis. It is clinically characterized by painless hypertrophy of the skin around the proximal interphalangeal joints of the fingers, more common in males. This rare condition can affect both hands and is often associated with mechanical injury of the skin. Although there is no specific therapy, cessation of mechanical trauma associated with topical corticosteroid therapy can lead to satisfactory results. We present 3 sporadic cases of classical pachydermodactyly, anatomopathologically confirmed by the presence hyperkeratosis and acanthosis, which responded favorably to topically applied corticosteroids under occlusive dressings. Keywords: FIBROMATOSIS, PACHYDERMODACTYLY, STEROID THERAPY

Included in the category of superficial fibromatoses, pachydermodactyly defines a skin condition characterized by painless swelling of the tissues around the proximal interphalangeal joints of the fingers, as shown by its name (gr. pachy = thick, derma-atos = skin, dactylos = finger).

Pachydermodactyly is more common in males (male/female ratio 3/2). Clinically it is characterized by hypertrophy of the dorsolateral aspects of the fingers (especially the index, middle, and ring fingers), most often bilateral and symmetrical. The hypertrophic changes are progressive and limited to the skin around the proximal interphalangeal joint, the affected skin segment presenting no other changes (1).

Pachydermodactyly was first described by Bazex in 1973, and until 2014 only 150 cases have been reported worldwide, but the total number is certainly higher, many of them not being yet reported (2).

The medical literature describes cases of pachydermodactyly associated with carpal tunnel syndrome, Dupuytren's contracture, gynecomastia, varioliform macular atrophy, thyroid dysfunctions, Ehlers-Danlos syndrome, but the most common association is with local microtrauma. Based on these findings, Bardazzi et al. (6) described five clinical forms of pachydermodactyly:

- classic - associated to mechanical trauma and affecting more fingers;
- localized or monopachydermo-dactyly;
- familial;
- extended to the metacarpophalangeal joints and dorsal aspect of the hand;
- symptomatic - associated to tuberous sclerosis;
CASE REPORT
We present 3 sporadic cases of classic pachydermodactyly in male children aged 8, 11 and 13 years, two of them living in rural and one in urban area. The disease was diagnosed one year earlier, and the patients were monitored at the Dermatology Clinic of the Iasi "Sf. Spiridon" University Emergency Hospital (fig. 1).

Fig. 1. Skin changes around the proximal interphalangeal joints

The children presented to the clinic with asymptomatic, diffuse thickening of the skin around the proximal interphalangeal joints of fingers II-IV, bilateral in two cases and unilateral in one case. Physical examination revealed the absence of joint pain and local warmth.

Case history showed that the changes occurred many years ago, being caused by repetitive micro trauma related to farm work in which rural children take active part and also to the habit of rubbing the hands together associated especially with emotional states, behavior noticed by their parents before the occurrence of symptoms.

Laboratory tests ruled out an autoimmune-mediated inflammatory disease, the biological, biochemical and immunological tests being within normal ranges. Also, X-rays of the hands showed no joint abnormality and secondary nuclei normal for age (fig. 2).

Fig. 2. X-ray image
Comparative ultrasound assessment of the hands denied the presence of any abnormal collections within the radiocarpal, metacarpophalangeal, and proximal interphalangeal joints, and appearance of normal extensor tendons, without tenosynovitis.

The clinically evident deformities around the proximal interphalangeal joints were solely due to skin thickening. At this level, skin thickness was 3.4 mm, compared to 0.7 mm in the unaffected areas on the dorsal aspect of the wrist (5-fold higher). The thickness of the scalp was also measured, and as it was normal a diagnosis of pachydermoperiostosis (Touraine-Solente-Gale syndrome) was ruled out.

Together, the clinical and imaging findings made the diagnosis of pachydermodactyly, subsequently confirmed by histopathological examination, which revealed irregular thickening of the skin with slightly elongated dermal papillae, acanthosis and discrete spongiosis (fig. 3); the corneous layer exceeded the thickness of the epidermis and the granular layer was prominent (fig. 4); there was extensive collagenization of the dermis, and fat glands and lobules were divided by thick connective tissue septa (fig. 5, 6).

Fig. 3. Irregular epidermis with prominent granular layer, HE, x 4

Fig. 4. Keratin crust thicker than epidermis, HE, x 4.

Fig. 5. Marked collagenization of dermis, VG, x 4

Fig. 6. Fat lobules divided by connective tissue septa, PAS, x 4
In all three cases, the performed investigations, particularly pathological examination, allowed us to differentiate the lesions from lesions with similar symptomatology, such as pachydermoperiostosis, juvenile chronic arthritis, rheumatoid arthritis, gout (gouty tophi), lichen myxedematosus, psoriatic acropachy-dermodactyly, acromegaly, fibrosarcoma, arthritis of the fingers (Heberden nodes), and joint involvements associated with endocrinopathy.

Caught in an early stage of disease, the patients had a positive response to topical corticosteroids, other invasive therapeutic procedures not being necessary. The described changes were classified as having the same degree of soft tissue injury even if in children with bilateral involvement they were present in a greater number of fingers. Potent topical corticosteroids under occlusive dressings (8-10 hours/day for 3 weeks, then a decreased frequency of applications) were used.

Psychological counseling (both child and parents), focused on the discontinuation of the habit of rubbing the hands together in situations of mental or emotional distress and teaching new relaxation techniques aimed at correcting this behavior, was also recommended.

The course was favorable, improvements being obtained after the first weeks of local corticosteroid therapy (fig. 7).

**DISCUSSION**

Pachydermodactyly is regarded as a localized form of superficial fibromatosis that mostly affects young adult with a mean age of 21.2 years according to Sagransky. Most commonly, the changes occur on the dorsal and lateral aspects of proximal phalanges of the index and middle fingers, with symmetrical involvement of both hands (3, 4, 5).

The mechanisms involved in the pathogenesis of this disease are not fully understood, some authors claiming that it is induced by repetitive minor local trauma. In this respect, Sagransky et al. (4) reported 2 cases of pachydermodactyly in poultry processing plant workers handling a significant amount of chicken per days at a fast pace. Other authors suggest as a possible mechanism the habit of rubbing the hands together, which in time leads to diffuse skin thickening and hyperpigmentation along the metacarpophalangeal joints.

The clinical features and especially a history of repetitive trauma are enough to make a diagnosis of pachydermodactyly, but histopathologic findings are useful in excluding other diseases with similar manifestations.
Male gender, and childhood or adolescence, are major contributing factors in the clinical expression of pachydermodactyly.

Thus, histopathologic examination is not necessary for making the diagnosis since the diagnosis of pachydermodactyly is clinical; however it can support the diagnosis by revealing an increase in dermal collagen with variable degrees of hyperkeratosis and acanthosis. The dermis is thickened, with coarse collagen bundles, slight fibroblast proliferation, increased fibroblastic activity, and sometimes collagen deposition around sweat glands. The association of acanthosis with dermal thickening is not specific but characteristic of pachydermodactyly and is a useful criterion to distinguish pachydermodactyly from other similar diseases such as keratoderma, where dermal thickening is not present, or fibromatosis, where epidermal changes are usually absent (7).

Caught in the early stages, the patients responded favorably to local corticosteroid therapy, the dermatocorticosteroids proving their efficacy through their anti-inflammatory effects and ability to inhibit cell proliferation and collagen synthesis (8).

The anti-inflammatory effect of corticosteroids is due to the inhibition of the release of phospholipase A2, enzyme responsible for the synthesis of prostaglandins, leukotrienes and other arachidonic acid derivatives; corticosteroids also inhibit transcription factors such as activator protein-1 and nuclear factor-κB, involved in the activation of proinflammatory genes (lipocortin and p11CBP (calpactin binding proteins). Their immunosuppressive properties consist in the suppression of the synthesis and effects of hormonal factors involved in the inflammatory response, thus inhibiting the migration of leukocytes to the site of inflammation and interfering with the functions of endothelial cells, granulocytes, mast cells, and fibroblasts. The antiproliferative role is exerted through inhibition of DNA synthesis and mitoses, partially explaining the therapeutic action of these agents in dermatological diseases (9).

The efficacy of topical corticosteroids depends on their potency and degree of skin penetration, their activity being closely related to the ability to bind to a specific receptor in the cytosol, and also to the vehicle into which it is incorporated.

In addition to local action, important is the systemic absorption of topical corticosteroids. Systemic absorption depends on the pharmacokinetic properties of the molecule, as well as on the integrity of stratum corneum and the presence/absence of inflammation; thus, topical agents applied on the skin or mucous membranes have increased absorption if the skin surface is injured or covered by an occlusive dressing (10).

Skin absorption occurs via diffusion of the dermatocorticoid in the stratum corneum. Hydrophilic steroids have a higher penetration rate through the stratum corneum than the hydrophobic ones, while keratinocytes are penetrated more rapidly with lipophilic agents, which explain the multitude of clinical effects of dermatocorticosteroids. After diffusion in keratinocytes, corticosteroids bind to the glucocorticoid receptor in the cytoplasm, inducing the cascade of reactions from which the mediators of messenger RNA synthesis result. The binding capacity of the glucocorticoid receptor in the cytoplasm is greater the lipid solubility of the dermatocorticosteroid is higher. In the human body there are many cells that contain receptors for glucocorticoids, thus explaining the receptor-mediated effects of
glucocorticoids (11).

In the use of local corticosteroids, a rather important problem is the phenomenon of tolerance, which could require a discontinuous drug administration schedule, using a double dose every two days. Another problem is withdrawal, which can be avoided, as in the case of systemic corticosteroids, by progressively spaced applications, or maintaining everyday applications but using a lower potency topical corticosteroid. Also to keep in mind are the complications, occurring especially when potent local corticosteroids (classes 3 and 4) are used; these include skin atrophy, stretch marks, purpura, delayed wound healing, rosacea, pigmentation disorders, and the most frequent systemic complications are hypothalamic-pituitary-adrenal axis suppression and Cushing’s syndrome.

CONCLUSIONS

This case report presents the management of three cases of pachydermodactyly, a rare entity, with an favorable outcome at local corticotherapy. Its purpose is to attract attention to the importance of clinical findings and minimal non-invasive investigations that can make the diagnosis of pachydermodactyly, and could avoid unnecessary costs incurred by sophisticated imaging investigations or inappropriate therapies.

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