

## Dermoscopic Features of Pagetoid Dyskeratosis of the Palm<sup>☆</sup>



### Dermatoscopia de la disqueratosis pagetoide palmar

Pagetoid dyskeratosis is a frequent incidental finding in various skin lesions.<sup>1</sup>

The patient was a 27-year-old woman who consulted for an asymptomatic hyperpigmented lesion that had arisen on the left palm 18 months earlier. Examination revealed a dark brown, hyperpigmented macule measuring 40 × 3 mm, with well-defined borders and a linear appearance (Fig. 1). Dermoscopy showed a parallel pattern of the cutaneous ridges (Fig. 2). Incisional biopsy of the lesion revealed dyskeratotic cells in a pagetoid pattern (Fig. 3). Based on these findings we made a diagnosis of pagetoid dyskeratosis of the palm. Cryotherapy led to complete resolution of the lesion. After 2 years the patient remains practically asymptomatic. A very faint yellow macule persists at the same site, with no significant alterations on dermoscopy.

Pagetoid dyskeratosis can present clinically as a hyperpigmented macule. A literature search revealed only 3 cases in which this condition affected the palm.<sup>2-4</sup> To date, the dermoscopic pattern of a lesion located on the hand has only been described in 1 case.<sup>2</sup> That patient presented a dark brown macule over the distal phalanx of the fifth finger; the dermoscopic features included a parallel pattern of the cutaneous ridges and a fibrillary pattern that led to the need to exclude palmoplantar melanoma.

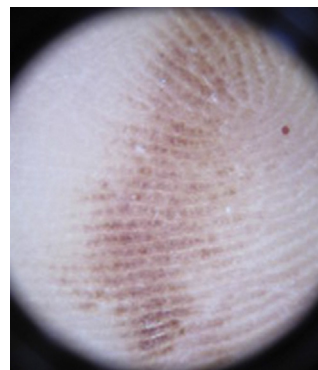
The etiology and pathogenesis of this condition are unknown, although it appears to be related to recurrent friction over the area. The prognosis is good.

On histopathology, dyskeratotic cells are observed together with larger-than-usual, round keratinocytes with pale cytoplasm and a pyknotic nucleus surrounded by a pale halo, in a pagetoid distribution.<sup>1</sup>

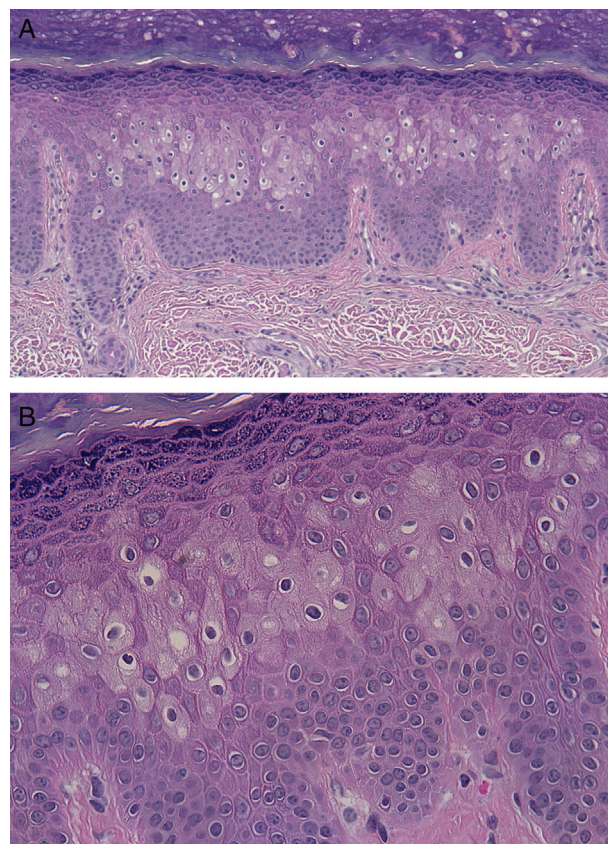


**Figure 1** Hyperpigmented macule with a linear pattern and well-defined borders, located on the palm.

The dermoscopy findings in this patient should be noted, as this pattern was suggestive of malignant melanoma, though it was also compatible with a diagnosis of subcorneal hematoma. Pagetoid dyskeratosis must be included in the differential diagnosis of pigmented palmar lesions that show a parallel cutaneous ridge pattern on dermoscopy.



**Figure 2** Dermoscopy of the lesion showing a parallel pattern of the cutaneous ridges.



**Figure 3** Histopathology: dyskeratotic cells in a pagetoid distribution. A, Hematoxylin and eosin, original magnification × 200. B, Hematoxylin and eosin, original magnification × 400.

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## References

1. Kanitakis J, Lora V. Pagetoid dyskeratosis: A frequent pitfall in dermatopathology. *Eur J Dermatol.* 2010;20:123–4.
2. Toyonaga E, Inokuma D, Abe Y, Abe R, Shimizu H. Pagetoid dyskeratosis with parallel ridge pattern under dermoscopy. *JAMA Dermatol.* 2013;149:109–11.
3. Frenk E, Delacr ez J. Hydropic degeneration of epidermal keratinocytes. An alteration leading to patchy hyperpigmentation. *Dermatologica.* 1974;148:135–42.

4. Wang LC, Medenica MM, Shea CR, Busbey S. Pagetoid dyskeratosis of the hand. *J Am Acad Dermatol.* 2004;50:483–4.

L. Loidi,<sup>a,\*</sup> J. Mitzelena,<sup>a</sup> A. C rdoba,<sup>b</sup> I. Yanguas<sup>a</sup>

<sup>a</sup> *Servicios de Dermatolog a, Complejo Hospitalario de Navarra, Pamplona, Spain*

<sup>b</sup> *Servicio de Anatom a Patol gica, Complejo Hospitalario de Navarra, Pamplona, Spain*

\*Corresponding author.

E-mail address: [leyre.loidi.pascual@navarra.es](mailto:leyre.loidi.pascual@navarra.es) (L. Loidi).

## Congenital Fascial Dystrophy or Stiff Skin Syndrome: A Case Report<sup>☆</sup>



### Distrofia fascial cong nita o s ndrome de la piel r gida: presentaci n de un caso

Congenital fascial dystrophy (CFD) or stiff skin syndrome is a rare skin disease that was described in 1971 by Esterly and McKusick.<sup>1</sup> The condition is characterized by noninflammatory fibrosis of the subcutaneous cellular tissue and of the muscle fascia, leading to hardening of the skin and interference with movement of the underlying joints. It can be hereditary and show a very variable degree of severity, sometimes causing minimal symptoms, as in the case we describe below, in which the diagnosis was made in adulthood.

The patient was a 46-year-old man with no past medical history of interest. He had been referred from another hospital with a clinical suspicion of morphea profunda that had not responded to treatment with oral corticosteroids. The patient stated that since childhood he had had difficulty performing certain movements, such as flexing the trunk, and that it had been impossible to administer intramuscular injections into the right buttock. He also said that his daughter had similar symptoms. Examination revealed difficulty raising a skin fold in the lumbar and right gluteal regions (Figs. 1 and 2), where the skin was hard to the touch, and a limitation of movement of the right hip, particularly flexion. Investigations performed at the previous hospital included an autoimmunity study that was normal, magnetic resonance imaging, which excluded bone or muscle involvement, and skin biopsy, which was reported to be consistent with morphea profunda. The biopsy was reviewed: thickened collagen was observed in a horizontally oriented lattice pattern in the deepest layers of the dermis, and there was no inflammatory infiltrate (Fig. 3). We made a diagnosis

of CFD based on the clinical and pathological findings, and physiotherapy was recommended.

CFD is a very rare disorder and descriptions are scarce in the literature.<sup>2</sup> It affects men and women equally, and a family history is found in 30% of patients. In the case described, the targeted history enabled us to identify a daughter with similar symptoms, though, at the time of writing, no additional tests had been performed to confirm the diagnosis.

The pathogenesis of this disease was unknown until recently.<sup>3</sup> However, Loeys et al.<sup>4</sup> discovered that all affected individuals had a mutation in the *fibrillin-1* gene. This protein, which is also mutated in patients with Marfan syndrome, is mainly involved in the formation of microfibrils (formed of fibrillin polymers), which, together with elastin, form the elastic fibers. This mutation leads to a disorganized accumulation of microfibrils in the dermis; this can be observed on confocal or electron microscopy. These accumulations produce abnormal activation of another molecule, transforming growth factor (TGF- $\beta$ ), which has the ability to promote collagen deposition in the dermis. The increased TGF- $\beta$  levels in patients with CFD leads to greater collagen deposition in the deeper regions of the dermis, the subcutaneous cellular tissue, and the muscle fascia.

Clinical manifestations are usually present at birth, but they can also appear during the first 6 years of life.<sup>2</sup> Patients with CFD have clearly circumscribed areas of hardened skin with no visible changes to the skin surface; these areas arise particularly around the pelvic or shoulder girdles and on the proximal areas of the thighs.<sup>5</sup> Less common findings include hypertrichosis, hyperpigmentation in the affected areas,<sup>6</sup> and the presence of subcutaneous nodules on the distal phalanges of the fingers.<sup>4</sup>

The main problem caused by this condition is limitation of joint movement. In the majority of patients this limitation is mild and does not interfere excessively with daily life; however, changes can sometimes be very widespread and can even limit lung capacity.<sup>2</sup> If the symptoms are mild, the diagnosis may not be made until adult life.

Not only the clinical manifestations but also the microscopic findings are very important. Microscopy reveals a proliferation of collagen tissue, particularly in the muscle fascia and in the subcutaneous cellular tissue, although there are reports of cases in which only the reticular dermis

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