WHAT IS THE DIAGNOSIS

# Generalized keratotic plaques

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## CLINICAL PICTURE

A 13-year-old female patient, with no relevant medical history, consulted the Pediatric Dermatology Department due to multiple asymptomatic 3-year lesions. Initially, they appeared on the back of both feet, progressively spreading to several parts of the body. During the medical consultation, the mother did not report any previous concomitant autoimmune or familial diseases. Physical examination showed multiple wart-like, round and white keratotic plaques at the back of both hands and lateral side of the feet, wrists and knees. The genital area was not affected (Picture 1). Dermoscopy revealed pearly white lesions with inner marked follicular accentuation (Picture 2).

*Foto 1 a,b. Placas blanquecinas hiperqueratósicas en dorso de manos y muñecas.*

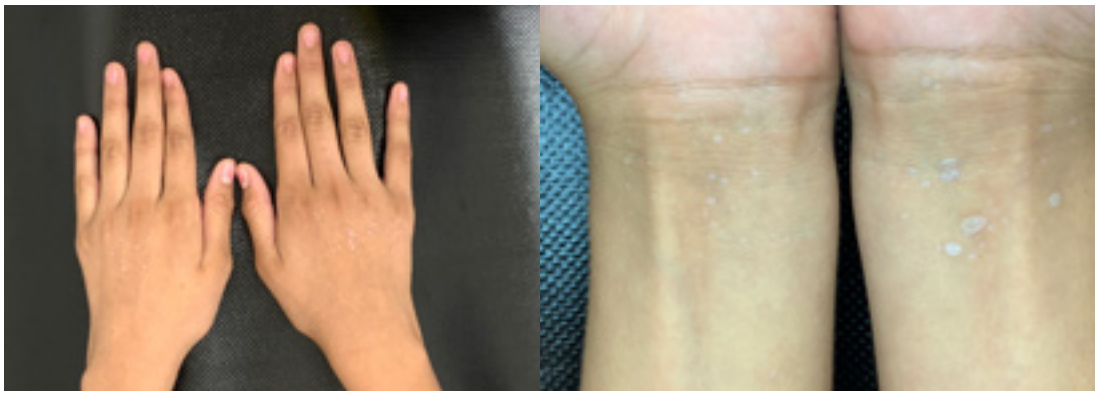
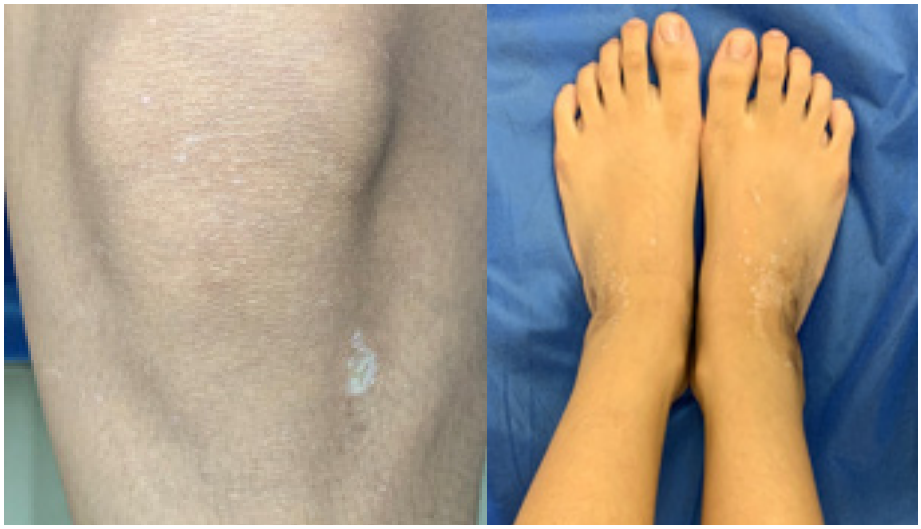
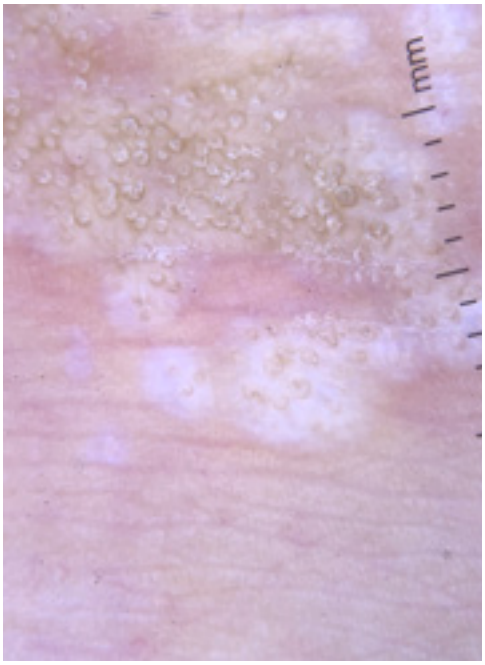


*Foto 2 a,b. Placas blanquecinas hiperqueratósicas en rodillas y cara lateral de ambos pies.*





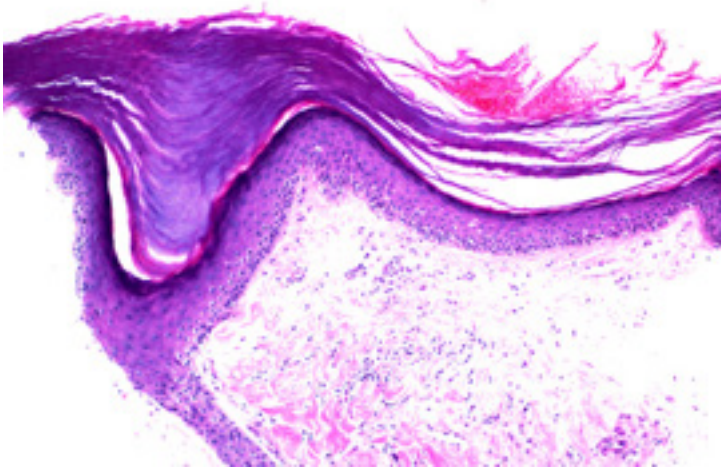


*Foto 3 (IZQ). A la dermatoscopía se visualizan placas blanquecinas con tapones córneos.*
*Foto 4 (SUP). Histología con HE donde se observa características histopatológicas de la enfermedad*

## WHAT IS THE DIAGNOSIS?

Histopathology revealed an epidermal hyperkeratosis with follicular plugging, atrophy of the stratum Malpighi and flattening of the rete ridges, in conjunction with homogenized collagen bands, dilated telangiectatic vessels and an inflammatory mononuclear perivascular infiltrate affecting the superficial dermis (Picture 3). Based on the clinicopathological findings, Lichen sclerosus was diagnosed. In consideration with the main clinical aspect of the lesions, a keratotic variant was contemplated. Treatment initiated with topical corticosteroids and UVA phototherapy, which resulted in partial clinical improvement and regression of the keratotic component.

## DISCUSSION

Lichen sclerosus (LS) is a chronic inflammatory disease of the skin which rarely occurs in infants. It mainly affects anogenital and perineal skin. However, concurrent or isolated extragenital lesions can be developed with minor occurrence.[1,2,3] Nowadays, there is strong evidence that the disease may have an autoimmune etiopathogeny in genetically susceptible patients.[1,2] Among pediatric patients, prepubescent girls are the most commonly affected. Typical lesions are white polygonal papules that coalesce into plaques and progressively acquire a smooth, atrophic and porcelain-white surface.[1,2,3] A rare clinical and atypical variant known as keratotic lichen sclerosus has been described to have appeared in extragenital areas, characterized by prominent follicular plugs and wart-like lesions.[4,5] Its diagnosis is established based on a clinical-pathologic correlation. Characteristic microscopic findings in early lesions correspond to orthohyperkeratosis, atrophy, follicular occlusion by cornified plugs, a lichenoid mononuclear infiltrate and a homogenized and edematous papillary dermis. In older lesions, inflammatory infiltrates become scarce and patchy, while the superficial dermis turns sclerotic and hyalinized.[1-5] The main differential diagnoses of LS are vitiligo, postinflammatory hypopigmentation, flat warts, lichen planus, lichen simplex chronicus and morphea.[1,2] Topical ultrapotent corticosteroids are the first-line treatment.[1,2,6] Treatment duration corresponds to three months.

Afterwards, they are applied when necessary. Approximately, 75% of patients become asymptomatic and 25% show subjective improvement.[6] Complementary phototherapy treatment is a favorable option in extragenital cases that potentiate the spread of the disease.[5,6]

## REFERENCES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