LETTER TO THE EDITOR

Pachyonychia congenita with only nail involvement

Dear Editor,

A 10-year-old boy presented with progressive thickening of the nails over the previous 5 years. He did not have any other cutaneous and/or systemic symptoms. He was born to healthy non-consanguineous parents following an uneventful pregnancy. Upon examination, he was found to have symmetrical involvement of the finger and toenails with marked hard thickening of the distal portion of the nail plate resulting in uplifting and tenting (Fig. 1). The palms and soles, oral cavity and scalp were normal. Potassium hydroxide examination of nails did not reveal any fungi. Systemic examination did not reveal any abnormalities.

Pachyonychia congenita is a rare, autosomally-dominant disorder characterized by symmetrical hard thickening of the nails with marked subungual hyperkeratosis, palmoplantar keratoderma with thick callouses, and oral leukokeratosis.1 Various other abnormalities such as hyperkeratotic follicular papules of the extremities, keratosis pilaris, blister formation of feet and palms, hair abnormalities, and hyperhidrosis of palms and soles have also been reported.2 Several clinical variants have been described:

Type I shows palmoplantar hyperhidrosis, extensive follicular hyperkeratosis that also involves the trunk.
Type II is the most common form characterized by scalloped tongue and oral leukokeratosis.
Type III manifests as corneal thickening and cataract.
Type IV is characterized by cysts on the head, neck and chest at puberty, hidradenitis suppurativa.

The age of onset of the various manifestations is variable, but dystrophy of the nails usually presents within the first month of life.3,4 However, many cases have been described with the onset of characteristic nail changes during the second or the third decade of life along with palmoplantar keratoderma, hyperhidrosis and oral leukokeratosis. Such cases could be interpreted as pachyonychia congenita tarda or late-onset pachyonychia congenita. Isolated early-onset nail change is very rare,5-7 and late-onset isolated nail dystrophy is even more unusual.6,8 Our case thus represents a rare presentation of late-onset pachyonychia congenita with only nail involvement.

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REFERENCES

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