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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 callosities, 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.

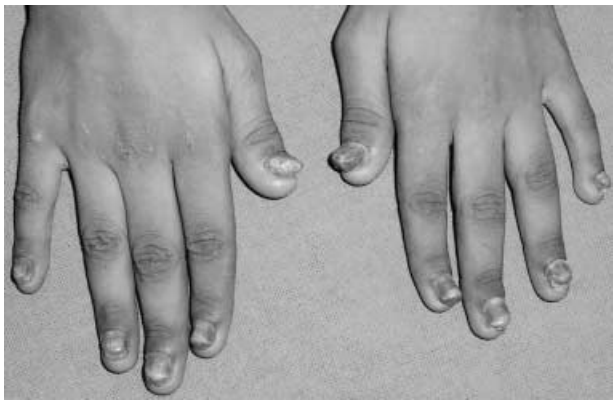

Figure 1. The typical nail changes of pachyonychia congenita.

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REFERENCES

1 Johnson Jr BL, Honig P. Congenital diseases (Genodermatosis). In: Elder DE, Elinitsar R, Johnson Jr BL, Murphy GF, (eds). *Levers Histopathology of the Skin*. 9th edn. Lippincott Williams and Wilkins, Philadelphia, 2005; 144–145.
2 Su DPW, Chun SL, Hammond DE, Gordon H. Pachyonychia congenia: A clinical study of 12 cases and review of literature. *Pediatr Dermatol* 1990; **7**: 33–38.
3 Iraci S, Bianchi L, Gatti S, Carrozzo AM, Bettini D, Nini G. Pachyonychia congenita with late onset of nail dystrophy – A new clinical entity? *Clin Exp Dermatol* 1993; **18**: 478–80.
4 Lucker GPH, Steijlen PM. Pachyonychia congenita tarda. *Clin Exp Dermatol* 1995; **20**: 226–229.

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5 Chang A, Lucker GPH, Kerkhof PCM, Steijlen PM. Pachyonychia congenita in the absence of other abnormalities. *J Am Acad Dermatol* 1994; **30**: 1017–1018.
6 Pryce DW, Verbov JL. A family with pachyonychia congenita affecting the nails only. *Clin Exp Dermatol* 1994; **19**: 521–522.
7 Dogra S, Handa S, Jain R. Pachyonychia congenita affecting the nails only. *Pediatr Dermatol* 2002; **19**: 91–92.
8 Nanda S, Grover C, Chaturvedi KU, Garg VK, Reddy BSN. Pachyonychia congenita tarda with nail manifestation only. *Pediatr Dermatol* 2005; **22**: 283–285.