Pachyonychia Congenita Affecting only Nails

A 9 years old boy presented with painful discolored and thickened nails of all 20 digits since birth. Antenatal, natal and immediate postnatal history was normal. At birth, parents noticed discolored nails of all digits. Over time, they thickened and became deformed, causing occasional pain and difficulty in holding small objects. On examination, all nails were discolored and thickened. There was transverse overcurvature, increasing along the long axis of nail from proximal to distal end. As a result, distal free end was most curved (***Fig*.1**). Palms, soles and mucosae did not reveal any abnormality. Rest of the mucocutaneous examination was noncontributory. Diagnosis of pachyonychia congenita was made. Pachyonychia congenital affecting only the nails is extremely rare.

The presentation is quite distinctive and hardly poses any diagnostic difficulty. Trauma and rarely, onychomycosis may result in pincer nail. However; it is limited to few nails only. Hereditary pincer nail syndrome (having distally directed exostoses on the tibial side of all toe phalanges, an obligatory criterion and may have destructive arthrosis of the terminal joints of digits) and pachyonychia congenita have pincer nail deformities of all nails. Pachyonychia congenita is a rare genodermatosis caused by mutation in keratin gene and is characterized by dystrophic, thickened nails (pachyonychia), symmetric focal palmoplantar keratoderma and oral leukokeratosis. Pachyonychia congenita has been divided mainly into 2 types: PC type 1 (more common, also known as Jadassohn-Lewandowski syndrome) and PC type 2 (also known as Jackson-Lawler syndrome). Palmoplantar keratoderma and oral leukokeratosis are less severe and less frequent in type 2. The distinguishing features of type 2 are natal teeth, steatocystomas, and pili torti. The treatment is mainly surgical with the aid of dermal grafts or partial matricectomy. Conservative treatment with urea 40% or steel brace has met with success; however, treatment has to be continued for a long time.

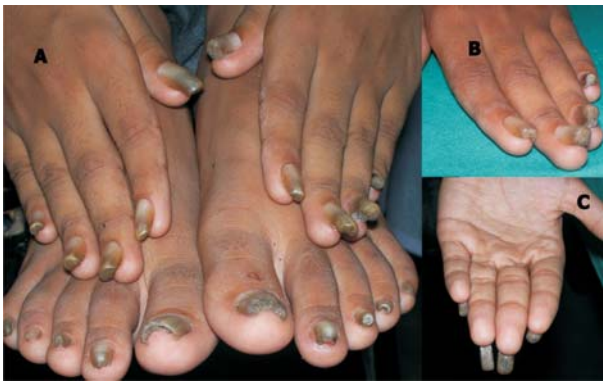

FIG. 1 *Discolored, thickened, deformed nails of all digits (A). Close up view- "Omega" shaped distal end of nail (B). Note absence of palm involvement (C).*

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