Median Canaliform Dystrophy of Heller

A 5-year-old boy presented with single dystrophic thumb nail since last seven months. Mother gave history of constant biting of thumb nails. On examination, there was median split of right thumb nail with transverse furrows extending form longitudinal split (***Fig.* 1**). All other investigations were non-contributory. He was also diagnosed to have attention deficit hyperactive disorder (ADHD).

Median canaliform dystrophy of Heller (solenonychina, dystrophia unguis mediana canaliformis, nevus striatus unguis) is an uncommon and rare dystrophic condition of nail plate which usually affects thumb nails symmetrically and is characterized by a midline defect of the nail plate leading to longitudinal splitting in the midline with canal formation. It extends from cuticle of nail and shows transverse furrows extending laterally from longitudinal splitting giving characteristic inverted fir tree appearance. It may rarely involve toe nails and other finger nails.

The proposed etiopathogenesis is repeated self inflicted trauma in form of 2habitual nail biting. In the majority of cases the cause is unknown and it may revert back to normal after many months to years. Other proposed that there is also absence of keratinocytes adhesions within nail matrix with dyskeratosis which is responsible for formation of longitudinal grove with splitting of nail plate due to weaker tensile strength. There

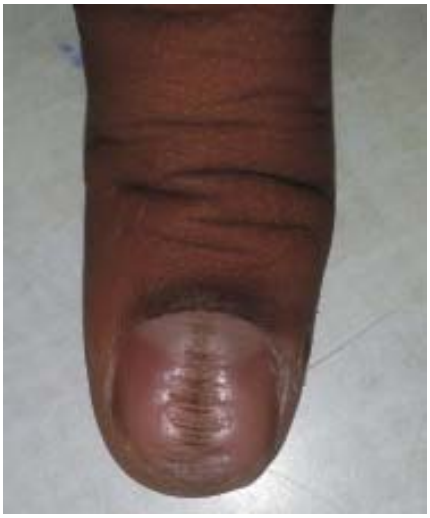

FIG.1 *Dystrophic thumb nail with medlan split.*

are some case reports of median canaliform dystrophy of Heller after using retinoids and with familial occurrence. Subungual tumors like glomus tumor, myxoid tumors, papilloma, squamous cell carcinoma, and melanoma may form longitudinal groove and then splitting the nail plate in the middle portion leading to median canaliform dystrophy.

The treatment includes avoidance of repetitive nail trauma through behavioral counselling, and topical tacrolimus.

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Childhood Bullous Mastocytosis

A 1-year-old girl presented with numerous pruritic bullae all over the body. There were complaints of recurrent episodes of diarrhea and vomiting. Development was normal. She started developing episodes of intense itching at 8 months of age, followed by appearance of erythematous macules, plaques, and tense bullae at the sites of itching as well as other sites (scalp, trunk and upper extremities) (***Fig.* 1 and 2**). Bullae were present on a non-urticated base containing clear fluid. Few of the urticarial plaques showed *peau-d-orange* appearance. Darier's sign was positive. The palms, soles and mucosae were free. Routine investigations and urine analysis were normal. Skin biopsy showed sub-epidermal bulla and an upper dermal inflammatory infiltrate comprising lymphocytes and many mast cells. Toluidine blue staining showed metachromatic granules and a diagnosis of bullous mastocytosis was made. The patient was treated with antihistamines for itching and topical as well as systemic antibiotics for preventing secondary infection. Parents were counseled regarding the prognosis and course of disease and the importance of avoiding certain medications that may provoke mast cell degranulation.

Bullous mastocytosis is a severe variant of mastocytosis, 30% cases manifesting within 6 months of age. The typical childhood disease is linked to Glu-839-