Yellow Nail Syndrome

A 16-year-old boy presented to us with recurrent chest infections, swelling of both lower limbs and discoloration of nails. Nails showed yellowish discoloration along with thickening and excessive curving of the finger nails (***Fig.* 1**), which had not responded to antifungals. A chest *X*-ray and computed tomography scan of the chest showed bilateral moderate pleural effusions. Thoracocentesis revealed an exudative pleural fluid with lymphocytic predominance.

Yellow nail syndrome is a clinical triad consisting of yellowed/thickened nails, lymphedema and recurring respiratory symptoms. Structural or functional lymphatic abnormalities and increased vascular permeability to albumin are proposed pathologic mechanisms. The syndrome is linked to a variety of underlying diseases, including rheumatoid arthritis, malignancy, immunodeficiency and endocrine disorders. The syndrome is characterized by remissions and relapses. Management is primarily supportive and palliative.

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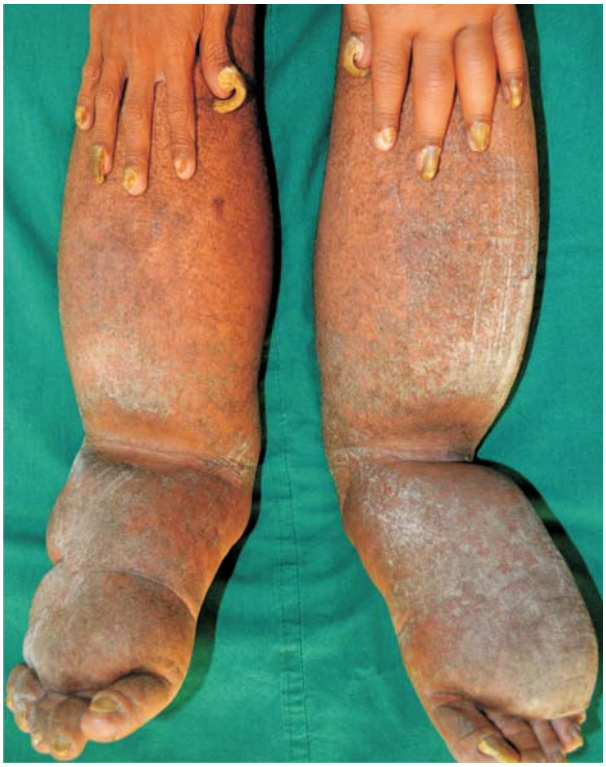

Fig. 1 *Yellow, thickened and curved nails with pitting edema in both lower legs.*

Mongolian Spots in GM1 Gangliosidosis

A 6-month-old girl born out of second degree consanguinity presented to us with multiple greyish-blue, well- to ill-defined macules and patches of varying sizes, involving chest, abdomen, back and buttocks (***Fig.* 1**). Child had global developmental delay with developmental age of 2-3 months. Physical examination revealed coarse facial features, hepatomegaly and pansystolic murmur. Further investigations revealed normal fundus, hypertrophic cardiomyopathy and tiny persistent ductus arteriosus. α-Galactosidase activity in peripheral blood leukocytes was significantly reduced, indicative of type I (infantile) GM1 gangliosidosis.

Typical and limited mongolian spots are benign blue or slate gray macular lesions of varying sizes, most commonly located on lumbosacral region. Mongolian spots usually fade during the first few years of life, but they occasionally persist. Those associated with inborn errors of metabolism are extensive and show no sign of resolution. Extensive

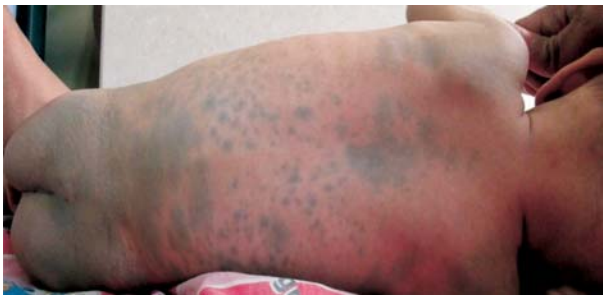

Fig. 1 *Multiple greyish-blue macules and patches of varying sizes involving whole back and buttocks.*

Mongolian spots are most frequently associated with GM1 gangliosidosis, Hurler-Scheie syndrome, Niemann-Pick disease, Hunter syndrome and α-mannosidosis. The findings of generalized mongolian spots in an infant may represent underlying storage disorders thereby allowing identification of families at risk.

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