HEREDITARY PALMOPLANTAR KERATODERMA ASSOCIATED WITH PRIMARY (CONGENITAL) LYMPHEDEMA

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ABSTRACT

The palmoplantar keratodermas are a heterogenous group of hereditary disorders of keratinization. They are characterized by epidermal thickening and a yellow waxy appearance of the palms and soles. Genetic studies have linked various forms of palmoplantar keratoderma to markers on chromosomes one, twelve, and seventeen, and several genes have been identified. Primary lymphedema is occasionally present at birth (congenital lymphedema or Milroy’s disease), but more commonly develops at puberty (lymphedema praecox). Genetic studies have linked various autosomal dominant forms of primary lymphedema (Milroy’s disease and lymphedema distichiasis), to genes on chromosomes five and sixteen respectively. We report a case of palmoplantar keratoderma in a child with congenital lymphedema. To our knowledge, this has not been previously described and may represent a new phenotype for future genetic study.