AUTOIMMUNE CONNECTIVE TISSUE DISEASES

LAUGIER-HUNZIKER SYNDROME ASSOCIATED WITH SCLERODERMA: A DIAGNOSTIC CHALLENGE

Agung Rheza [(1)] - Sondang Sirait [(1)] - Roro Krisanti [(1)] - Michael Angriawan [(1)] - Benny Nelson [(1)]

Faculty Of Medicine Universitas Indonesia/dr. Cipto Mangunkusumo National Public Hospital, Dermatology & Venereology, Jakarta, Indonesia [(1)]

Background: Scleroderma is an autoimmune connective tissue disease of unknown etiology that affects the skin, blood vessels and internal organs. Limited scleroderma is characterized by fibrotic skin changes that are limited to the fingers, hands and face and includes the CREST syndrome. In diffuse scleroderma, generalized fibrotic skin changes are seen, usually start in the fingers and hands but spread to involve the forearms, arms, trunk, face and lower extremities. Laugier-Hunziker syndrome is a rare, acquired mucocutaneous hyperpigmentation often associated with longitudinal melanonychia. In most cases, the clinical diagnosis should be confirmed by further diagnostic methods.

Observation: A 25-year-old Indonesian woman was referred to our hospital with a chief complaint of stiffness and thickening in both arms and legs for 6 months. We found sclerosis lesions, sclerodactyly, Raynaud phenomenon, and melanonychia in some fingernails. Histopathological examination from the skin showed hyperkeratosis with shortening rete ridges, thickening of subepidermis with fibrous connective tissue that contain collagen and consist of the core fibrocytes and fibroblasts, with the conclusion: scleroderma. Result from dermoscopic examination of the periungual were avascular areas and bushy capillaries. Homogenous regular band-like pigmentations with indistinct borders were found on fingernails. We also found brownish pigmentations in her oral cavity and labia mayora which suggests Laugier-Hunziker syndrome.

Key Message: Clinicians should be more familiar with typical features of these conditions and aware of important differential diagnosis. The evaluation involves a detailed history and physical examination, including visual inspection of all the affected sites. Dermoscopy and biopsy of any suspicious lesion should be performed for dermoscopic and histopathological evaluation.