Sclerodermatomyositis

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The classification of rheumatic diseases is still challenging due to several reasons. First, those diseases have several differential clinical features, which giving overlap symptoms. Second, the etiopathogenesis of those diseases remains elusive. Diagnosis of overlap syndrome is made when there are more than one well-defined connective tissue diseases in one patient, which may develop simultaneously or sequentially.[1,2] The prevalence of overlap syndrome among autoimmune diseases is 25%.[2]

The term sclerodermatomyositis or scleromyositisis is used to describe an overlap syndrome in patients with scleroderma and dermatomyositis/polymyositis (DM/PM).[2,3,4] Sclerodermatomyositis usually affects adults, and it is rarely found in children.[4] The clinical features of this syndrome are myalgia or myositis, arthralgia, scleroderma-like skin changes, Raynaud's phenomenon (RP),[2,3] interstitial lung disease, calcinosis,[3] mask-like facies, dysphagia or esophageal dysmotility,[4] as well as the presence of specific antibody Pm/Scl.[2] Skin manifestations as the part of dermatomyositis include periorbital erythema and Gottron's papules.[3]

We report this case due to its very rare occurrence. According to medical records in the Department of Dermatology as well as Rheumatology at Hasan Sadikin Hospital, Bandung, this is the first case recorded in the last 10 years.

CASE REPORT

An 18-year-old, single, Sundanese female, came to our institution with chief complaint of rashes on her arms, hands, legs, and abdomen.

Since 2 years prior to this visit, she has had complaint of joint pain on both elbows, with rashes on the surface of her knees and elbows, as well as photosensitivity and oral ulcers. At that time, his physician considered systemic lupus erythematosus (SLE) as the cause of her symptoms. Laboratory test results were normal, except for leucopenia (white blood cell count of $4 \times 10^3/mm^3$). At that time, the patient also had negative antinuclear antibody (ANA) and anti–double-stranded DNA (anti-dsDNA) tests. She was given nonsteroidal anti-inflammatory drugs (NSAIDs) and subsequently showed improvement.

One year prior to admission, she started to have difficulty swallowing foods and liquids, also accompanied with rough skin on her face, rashes on her eyelids, face, chest and back, as well as photosensitivity. Afterwards, she developed erythematous rashes on her arms, abdomen and both legs, accompanied with skin tightening. She also had alopecia and weight loss.

Physical examination on admission at our institution revealed normal vital signs. There were heliotrope rashes on her palpebras, and Gottron's papules as well as Gottron's sign over the surface of her metacarpophalangeal joints and elbows. She had rashes on her neck (shawl sign, V-sign), sclerodactyly, and muscular atrophy of her upper extremities. Rodnan skin score was 19. On palpation, the skin on her abdomen, arms, hands, legs, and feet felt tight and taut. She had difficulty raising her arms and step on ladders or stand up from sitting position.

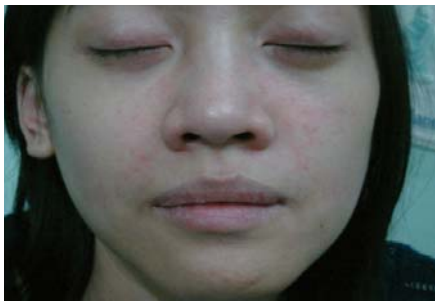

Figure 1 Heliotrophe on patient's eyes

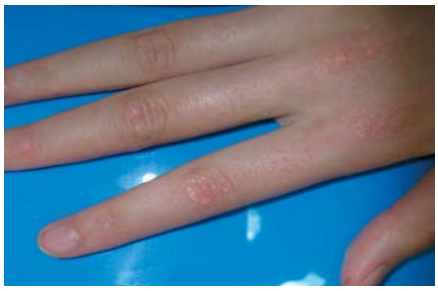

Figure 2 Sclerodactyly and Gottron's papules on PIP and DIP surface

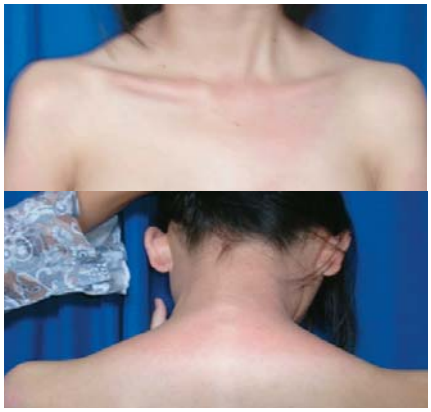

Figure 3 V sign and shawl sign