

Diffuse Palmoplantar Keratoderma Associated with Acrocyanosis and Livedo Reticularis Two Sporadic Cases

Sir,

The association of diffuse palmoplantar keratoderma with acrocyanosis is very rare, and it has been described in only one pedigree (1) to our knowledge. We report two sporadic cases of diffuse palmoplantar keratoderma with this association.

Case 1. A 26-year-old Japanese woman had had keratoderma of the palms and soles from an early age and cutaneous colour abnormality of the hands and feet for 3 years. She did not know whether the keratoderma had been present at birth. The discoloration was persistent all the year round. She had no history of Raynaud's phenomenon, perniosis or cold injury. Her family history was non-contributory for either keratoderma or discoloration.

On physical examination, her palms and soles were light reddish, rather than reddish-blue, and showed symmetrical, diffuse, smooth hyperkeratosis. However, the dorsa of her hands and feet were cyanotic with diffuse, reddish-blue discoloration, which easily blanched on pressure. Her fingers and toes were slightly swollen without pain. There was also reticular, reddish-blue livedo on her forearms and whole legs. As soon as her arms and legs were raised, the cyanosis and livedo completely disappeared and the skin reassumed a normal colour.

The following laboratory findings were normal or negative: full blood cell counts, liver and kidney function tests, serum electrolytes, erythrocyte sedimentation rate, C reactive protein, amylase, IgA, IgE, IgG, IgM, C3, C4, cryoglobulins, cold hemagglutinins, Paul-Bunnell test, anti-streptolysin O antibodies, anti-streptokinase antibodies, antinuclear antibodies, LE factor, rheumatoid factor, serological tests for syphilis, and anti-cardiolipin antibodies. The ECG and chest X-ray were also normal.

Histologically, a skin specimen from the left foot showed hyperkeratosis, acanthosis without acantholysis, dilation of the small blood vessels in the upper dermis and an infiltration of a few lymphocytes around the blood vessels.

Case 2. A 36-year-old Japanese man had developed slightly reddish, diffusely keratotic palms and soles at an early age. The keratosis has become aggravated over the last 10 years. On the other hand, the skin colour on the distal extremities had become reddish-blue since adolescence. The discoloration was seldom affected by alterations in temperature. He had no past history of Raynaud's phenomenon, perniosis or cold injury, and no family history of keratoderma or discoloration.

On physical examination, his palms and soles were light

reddish and showed symmetrical, diffuse hyperkeratosis with surrounding erythematous bands. The soles were more keratinized than the palms. However, the dorsa of his hands and feet were cyanotic with diffuse, reddish-blue discoloration. His forearms, legs and lower back had reticular, reddish-blue livedo.

On laboratory investigations, there were no abnormalities, as in Case 1.

Histologically, a skin specimen from the left palm showed hyperkeratosis, acanthosis without acantholysis and the infiltration of a few lymphocytes in the papillary dermis. An acrocyanosis lesion from the left forearm showed perivascular infiltration of only a few lymphocytes in the dermis.

Although the exact mechanisms of acrocyanosis are unknown, a vasospasm (2) or blood high viscosity (3) may be related to a phenomenon of acrocyanosis. In our cases, acrocyanosis was complicated by livedo reticularis as well. However, such poor blood circulation of acrocyanosis does not usually cause hyperkeratosis on unwounded skin. Moreover, the onset of diffuse palmoplantar keratoderma is generally earlier than that of acrocyanosis, so acrocyanosis may not directly induce diffuse palmoplantar keratoderma.

As patients with diffuse palmoplantar keratoderma or acrocyanosis frequently have a family history, and their association has also been reported as inherited, we suggest that mutation occurred in genes responsive to diffuse palmoplantar keratoderma and acrocyanosis in our cases. If different genes mutated at the same time, the genes may have been related to each other in location or may have had other common characteristics in mutation.

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