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# Palmoplantar keratoderma as the preceding cutaneous manifestation of juvenile-onset dermatomyositis

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A 6-year-old girl presented with skin lesions over the soles for last 4 months. These lesions were initially red and itchy and later became hyperkeratotic with scaling. For these symptoms, the 6-year-old girl was treated elsewhere and was prescribed topical corticosteroids. However, she showed no improvement with this therapy. After 4 months, she started developing progressive difficulty in walking and swallowing along with nasal intonation of voice, and she gradually became non-ambulatory.

On examination, she had heliotrope rash over the eyelids; Gottron's papule over the dorsum of the proximal interphalangeal joints; hyperpigmented dry, scaly, and hyperkeratotic lesions present over the soles (Figure 1a); erythema; and dryness over the palms (Figure 2). She had proximal muscle weakness (power grade 3/5) and poor gag reflex. Laboratory investigations revealed aspartate aminotransaminase level to be 202 IU/L (normal <44 IU/L), alanine aminotransaminase level to be 185 IU/L (normal <44 IU/L), creatine kinase level to be 3,290 U/L (normal <130 U/L), and serum lactate dehydrogenase level to be 1,310 U/L (normal <500 U/L). Extended immunoblot (Euroimmun) panel for myositis-specific antibodies showed positivity for anti-PM/Scl7 and anti-PM/Scl100, whereas antibodies for Mi2Aα, Mi2β, TiF-1γ, MDA5, NXP2, SAE1, Ku, Jo-1, SRP, PL-7, PL12, EJ, OJ, and Ro-52 were negative. Nail fold capillaroscopy revealed loss of capillaries, thrombosis, microhemorrhages, and arborization of the capillaries. T2-weighted fat-suppressed magnetic resonance imaging revealed hyperintense signals in the thigh muscles (Figure 3). A clinical possibility of juvenile dermatomyositis (JDM) based on the modified Bohan and Peter criteria with palmoplantar keratoderma was considered.[1] She was given (initiation) intravenous methylprednisolone pulse therapy (30 mg/kg/day for 5 days) followed by oral prednisolone in tapering doses (initial dose 2 mg/kg/day), subcutaneous methotrexate (15 mg/m$^2$/week), hydroxychloroquine (5 mg/kg/day), and keratolytic agents (salicylic acid). She showed marked clinical improvement in muscle weakness and palmoplantar cutaneous lesions (Figure 1b).

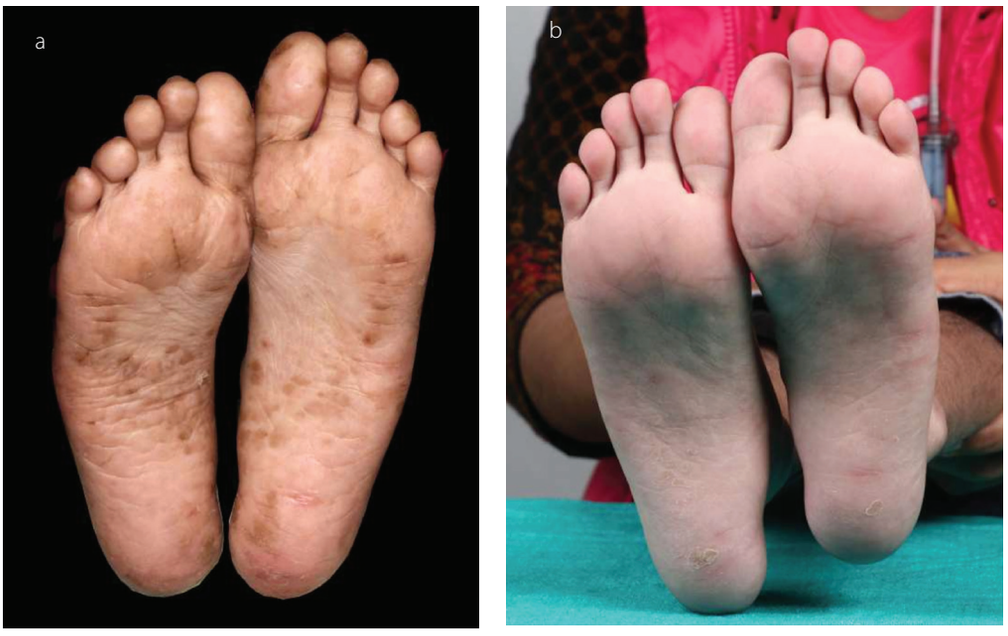


**Figure 1. a, b.** Showing palmoplantar keratoderma over the soles (a); Improvement of the lesions following the therapy (b).

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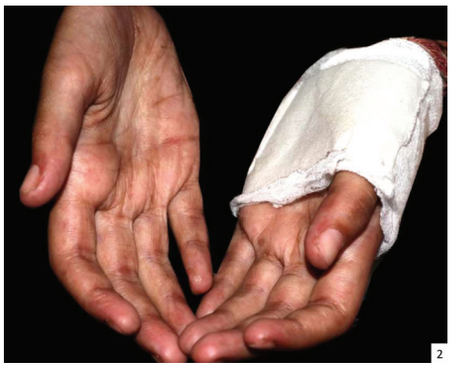

**Figure 2.** Erythema and dryness over the palms.

Heliotrope rash and Gottron's papule are the hallmark cutaneous manifestations of JDM. However, a variety of cutaneous lesions have been reported in these patients.[2] Palmoplantar keratoderma has rarely been reported in children with JDM. It has also been reported that palmoplantar keratoderma may precede the onset of JDM by few months as was seen in our case.[3,4] The pathogenesis of this cutaneous manifestation in patients with JDM is not known. However, it may be considered to be an exaggerated form of mechanic's hand. It has also been observed that palmoplantar keratoderma is associated with the presence of anti-PM/Scl in patients with JDM. Prevalence of anti-PM/Scl in patients with JDM is low. In adults, this has been found to be associated with anti-synthetase syndrome characterized by interstitial pulmonary fibrosis, arthritis, and Raynaud phenomenon.[5] Index patient showed anti-PM/Scl positivity; however, the manifestation of anti-synthetase syndrome was not seen.[6]

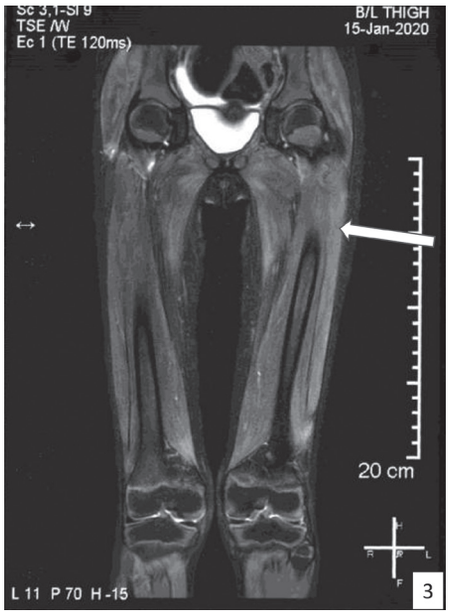


**Figure 3.** T2-weighted fat-suppressed magnetic resonance imaging showing diffuse hyperintense signals in the thigh muscles (arrow).

Palmoplantar keratoderma has been found to improve with the standard treatment given for patients with JDM.

To conclude, palmoplantar keratoderma may occasionally precede the clinical manifestations of JDM by several months. Therefore, children with palmoplantar keratoderma should be carefully observed on follow-up.

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