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LETTER TO THE EDITOR

Reversal of skin changes in smoldering myeloma with clinical presentation of POEMS syndrome with a lenalidomide-based regimen

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Dear editor,

POEMS (Takatsuki or Crow-Fukase syndrome), the acronym for polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, and skin changes, is a rare multi systemic disorder [1]. It affects men more often than women and usually presents in the 6th decade of life. Skin changes are prominent, with hyperpigmentation being the most frequent alteration [2]. Sclerodermoid skin changes are also very common, and white nails (leukonychia) are characteristic of the disease. Cutaneous angiomas can be present as well as vasculitis, hypertrichosis, and acrocyanosis. Hepatomegaly, splenomegaly, and/or enlarged lymph nodes can also be seen. Hypothyroidism, adrenal insufficiency, and diabetes mellitus represent the most common endocrine system–related disorders. Usually a monoclonal band is seen in serum protein electrophoresis; however, the number of plasma cells that infiltrate the bone marrow can be only slightly elevated. The rarity of the disease delays the diagnosis [3].

A 72-year-old previously healthy male was admitted to the hospital, because of fatigue, muscle weakness, skin changes, and respiratory distress. His symptoms started approximately 6 months earlier, with muscle weakness and skin changes as the most prominent findings. At the time of admission, the patient was confined to a wheel chair. He was very weak, and he had shortness of breath. Physical examination revealed scleroderma-like skin changes which consisted of thickening and induration of the skin, mainly in the extremities, dark skin discoloration, white fingernails (Fig. 1, panels a, b, and c), and edema of the lower extremities. Splenomegaly (16 cm) was confirmed with an ultrasound. Laboratory evaluation disclosed hypothyroidism, a monoclonal (M = 4.7 g/L) IgG-λ band in serum protein electrophoresis, and increased free λ chains (5.1 mg/dL, normal values 0.57–2.63 mg/dL). The free kappa chains were 1.97 mg/dL (normal values 0.33–1.94 mg/dL). A bone marrow examination revealed only slightly increased (15%) plasma cells in the bone marrow. Electrophysiological testing showed both changes of demyelination and axonal degenerative neuropathy.

Based on the clinical and laboratory findings, the diagnosis of smoldering myeloma was made, despite the clinical presentation with features of POEMS syndrome. Treatment with cyclophosphamide (500 mg on days 1, 8, and 15 in a 21-day cycle), corticosteroids (40 mg dexamethasone/week), and lenalidomide (25 mg/day) was initiated [4]. The patient received six cycles of this regimen (with 1 week off between cycles), followed by lenalidomide only as maintenance treatment. Two months after the initiation of treatment, the patient's muscle weakness greatly improved. His skin color, thickness, and nails also improved (Fig. 1, panel d). Six months later, the improvement was prominent; the skin was soft, and nails were not white any longer (Fig. 1, panel e). No monoclonal band was detected in the protein electrophoresis, and free light chains were within normal limits.

A year later, skin changes returned to normal (Fig. 1, panel f), and the patient remains on a continuous low dose of lenalidomide, as maintenance therapy. The patient now is capable of driving by himself to the hospital for his follow-up sessions.

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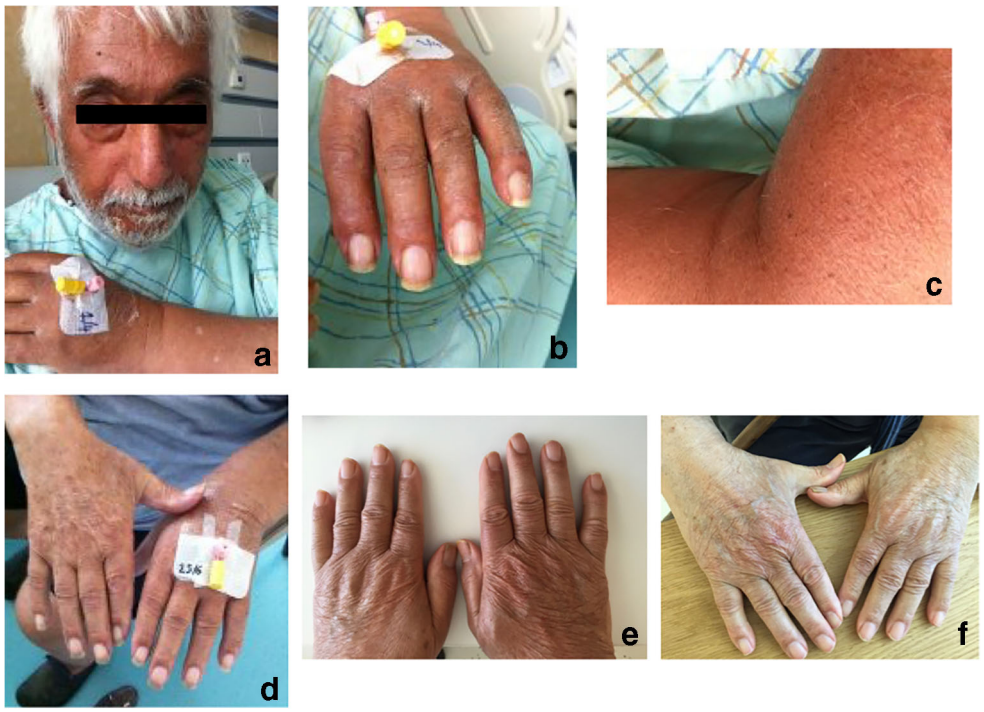

Fig. 1 Skin changes in POEMS syndrome. **a**, **b**, **c** Scleroderma-like skin changes, edema, and white fingernails at the time of diagnosis. **d** Two months later, skin changes improved. **e** After six months, the skin was soft and the nails had a normal color. **f** Complete remission of skin changes a year after diagnosis

The pathophysiology of POEMS syndrome is not fully understood, but VEGF is implicated in the disease. The rarity of the disease makes it difficult to consider in the differential diagnosis. Any patient with the above symptoms and described skin changes should be thoroughly evaluated if fulfills criteria for the disease or if represents a plasma cell dyscrasia associated with features of POEMS syndrome [5].

Compliance with ethical standards

All procedures performed in this manuscript were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Written informed consent was obtained from the individual participant described herein to be included in this study.

Conflict of interest The authors declare that they have no conflict of interest.

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