Skin changes in POEMS syndrome

GIUSEPPE LONGO, GIOVANNI EMILIA, UMBERTO TORELLI
Department of Medical Sciences, Section of Internal Medicine, University of Modena , Italy

The so-called POEMS syndrome is a multisystemic disorder characterized by the association of polyneuropathy, organomegaly, endocrinopathy and skin changes. These disorders seem to be secondary to a plasma cell dyscrasia leading to production of a monoclonal component. Several other signs can occur, e.g. anasarca, pyrexia, finger clubbing, sweating and hematologic disorders.

The syndrome has been described mainly in Asians, although some Caucasian cases have been reported.[1,2] We report the images of skin changes which appeared in a 39-year-old Caucasian male; all the other above mentioned signs were also present. The diagnosis of POEMS syndrome was established in April 1987. The patient developed a progressive peripheral polyneuropathy with demyelination, hepatosplenomegaly, sclerotic bone lesions, scleroderma and IgGλ monoclonal gammopathy. Bone marrow examination showed about 9% plasma cells of apparently normal morphology. Two years later the patient had a myocardial infarction with transient thrombocytosis together with papilledema. A consistent improvement was obtained with plasmapheresis, chemotherapy (melphalan) and high dose prednisone. After a further three years severe polyneuropathy reappeared, with symmetrical motor and sensory deficiencies in the limbs, peripheral edema, pleural effusion, hypogonadism and hypothyroidism. Treatment with melphalan, cyclophosphamide and plasmapheresis together with prednisone was not very effective. Since September 1997 the patient has had rapid, progressive appearance of multiple skin angiomas (50-60, several tuberous), together with dermal fibrosis with sclerodactylia, pleural and myocardial effusions, pyrexia and capillary leak syndrome. A slight improvement was obtained using high dose dexamethasone and this has been maintained up to now with intermittent dexamethasone treatment.

Correspondence: Giuseppe Longo, M.D., Department of Medical Sciences Section of Internal Medicine, University of Modena, via del Pozzo 71, 41100 Modena, Italy. Phone: international +39-059-422782 – Fax: international +39-059-424549 – E-mail: longo@unimo.it

References

1. Soubrier MJ, Dubost JJ, Sauvezie BJM, the French Study Group on POEMS Syndrome. POEMS syndrome: a study of 25 cases and review of the literature. Am J Med 1994; 97:543-53.
2. Ropper AH, Gorson CK. Neuropathies associated with paraproteinemia. N Engl J Med 1998; 338:1601-7.