SKIN MANIFESTATIONS OF INTERNAL DISEASE

POEMS SYNDROME: A DIAGNOSIS NOT TO BE IGNORED

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Background: POEMS syndrome is a rare multi-visceral systemic disorder associating polyneuropathy (P), organomegaly (O), endocrinopathy (E), monoclonal gammapathy (M) and skin lesions (S). The polymorphism of the clinical manifestations makes the diagnosis of this disease difficult and might delay its management.

Observation: We report a case of a 50-year-old man who had a diabetes type 2 evolving for 5 years. He was hospitalized for pain and functional impotence of the 4 limbs, associated with dermatological disorders and evolving in a context of deterioration of the general state. Clinical examination revealed a disorder of motility and sensitivity with areflexia of the 4 limbs, a hepatosplenomegaly, big hands and feet, a facial lipoatrophy, a hyperpigmentation predominant on the extremities, a distal cutaneous sclerosis and a leuconychia. Electromyogram showed demyelinating sensitivo-motrice neuropathy of the 4 limbs. Laboratory tests revealed hypothyroidism and monoclonal gammapathy IgA lambda type. The diagnosis of POEMS syndrome was than evoked.

Key message: POEMS syndrome would be more common in Japan. Male predominance has been reported. The average age of onset is between 40 and 50 years. Some authors require the presence of two major criteria (neuropathy and monoclonal gammapathy) with a minor criterion among the following criteria: osteoconductive lesion, Castleman's disease, organomegaly, edema (peripheral, pleurisy, ascites), endocrinopathy (adrenal, pituitary, parathyroid, thyroid, diabetes), cutaneous manifestations (hyperpigmentation, cutaneous sclerosis, hypertrichosis, angiomas, facial lipoatrophy, white nails ...). Polyneuropathy remains the constant element and most often revealing. Apart from the five main manifestations of the POEMS syndrome, the clinical presentation is highly polymorphic and may include: febrile syndrome with deterioration of general state, renal failure, diarrhea , pleural effusions, heart failure, thrombocytosis, polycythemia, bilateral papilledema ... Treatment with thalidomide, lenalidomide and melphalan-dexamethasone combination has been successful. The marrow transplant would be indicated in forms refractory to corticosteroid therapy, chemotherapy and radiotherapy.