POEMS SYNDROME: CLINICAL AND HISTOLOGICAL DESCRIPTION OF FOUR PATIENTS

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OBJECTIVE: To describe the clinical and histological features of four patients with POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, M protein and Skin Changes), seen in our hospital during the period of time 2001-2010
METHODS: Retrospective revision of four clinical records, histopathology study of samples of sural nerve biopsy
RESULTS: Four patients, three women and one man, aged between 41 and 80 years, met criteria for POEMS syndrome: they presented a sensorimotor polyneuropathy with demyelination and secondary axonal degeneration (with rapidly progressive course in three cases) and a monoclonal plasma cell proliferation expressed as: IgA-lambda monoclonal gammopathy or solitary myeloma. In addition, patients had other systemic symptoms: hepatosplenomegaly, lymphadenopathy, ascites, pleural effusion, edema of lower extremities, endocrinopathy, skin changes in the form of hyperpigmentation and hypertrichosis and bilateral papilledema. We performed a sural nerve biopsy in three patients where there was a significant loss of myelinated fibers and axonal degeneration. Antitumor treatment was administered with chemotherapy or radiotherapy and in one case with autologous peripheral blood transplantation. In general the response to these treatments was poor, except for the case treated with autologous peripheral blood which had a stabilization and improvement. Systemic symptoms improved more than the polyneuropathy. Two patients died from complications of extreme generalized edema.
CONCLUSIONS: POEMS syndrome is a severe paraneoplastic complication associated with some lymphoproliferative syndromes that requires early diagnosis and aggressive therapeutic approach.