SKIN MANIFESTATIONS OF INTERNAL DISEASE

LICHEN MYXEDEMATOSUS ASSOCIATED WITH IGA TYPE MULTIPLE MYELOMA: A RARE CASE IN A CHINESE MAN

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Backgroun: Lichen myxedematosus (LM) is a chronic, idiopathic disorder characterized by an abnormal accumulation of mucin in the skin. It might be associated with paraproteinemia or multiple myeloma in rare times.

Observation: A 55-year-old man presented to our inpatient department, with the complaints of red papules and plaque on his whole body with mild itching. General physical examinations were normal. On cutaneous examination, there were bilaterally symmetrically densely distributed firm, infiltrative, non-scaly, non-exuding papules and plaques on face, trunk and limbs. A biopsy was taken and showed unremarkable epidermis, infiltration of chronic inflammatory cells, coarse proliferation of collagen fibers and mucin deposition in the whole dermis with positive alcian blue staining. Laboratory studies revealed the following: thyroid function tests were negative; Liver function tests showed hyper gammaglobulinemia with reversal of A/G ratio. Humoral immunity examination showed IgA 47.8g/L(normal 0.70-4.00). Bone marrow aspiration showed abnormally proliferated plasma cell system and the original and naïve plasma cells accounted for 13% of ANC. Bone marrow biopsy displayed plasmacytoid differentiation of hematopoietic cells and immunohistochemistry showed CD3(+), CD38(+), CD138(+), MPO(+), reticular fiber staining(+); Protein electrophoresis showed IgA-kappa M-protein(+); β2 microglobulin levels were elevated to 4.01 mg/L. Thus, the diagnosis of multiple myeloma was confirmed.

Key message: LM is a rare chronic metabolic disease characterized by localized or generalized form with lichenoid papules, nodules, plaques and even sclerodermoid change. The pathological features are dermal deposition of mucinous material and fibroblast proliferation with no evidence of amyloid deposition. LM can be associated with monoclonal gammopathy sometimes but overt multiple myeloma develops in only a minority of patients. Most of the times the associated monoclonal gammopathy is of IgG lambda subtype. As far as we know, this is the first report of LM associated with multiple myeloma of IgA kappa subtype.