MEDICAL THERAPIES AND PHARMACOLOGY

SCLEREDEMA TYPE II: A NEW CASE TREATED WITH IMMUNOGLOBULIN

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Background: Buschke scleredema (BS) is a rare connective tissue disease of unknown cause. Classically, three types are distinguished: the first is acute, post-infectious and spontaneously resolving; the other two, of insidious installation and chronic evolution, are classically associated with monoclonal gammopathy or diabetes. We report a case of BS type II treated with intravenous immunoglobulin (IVIg)

Observation: A 52-year-old patient, admitted for diffuse cutaneous sclerosis evolving since 2 years . Clinical examination found diffuse cutaneous sclerosis sparing the extremities associated with hyperpigmented lesions in the neck, armpits, wrist, knees,ankles.The osteo-articular examination had objectified a limitation of the movements of flexion extension of the neck and the adduction of the two shoulders. The histological study of sclerosis and hyperpigmented lesions showed BS and acanthosis nigricans (AN) respectively. Serum protein electrophoresis with immunofixation revealed a monoclonal peak of gamma globulins with the presence of a lambda serum IgG monoclonal immunoglobulin, the myelogram was normal.The patient was put under IVIG, 2g / kg for 2 consecutive days every 6 weeks associated with UVB phototherapy, currently the patient is at his 4th treatment with beginning of desinfiltration of cutaneous sclerosis and improvement of the articular amplitudes. The decline is 6 months.

Key messages: The originality of our observation lies in the rarity of this entity as well as the good evolution under immunoglobulin. BS is characterized by sclerotic edema of pilgrim that progressively extends to the trunk and limbs, but spares the extremities. The association with AN has been reported in 2 cases in the literature without this relationship being clearly established. No treatment is consensuel : corticotherapy, ciclosporin, radiotherapy, methotrexate, thalidomide, IVIG, photopheresis and phototherapy are among the options reported in clinical cases and small series. IVIG is an interesting alternative in Bushk type II, when treatment is needed.