INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

SUCCESSFUL TREATMENT OF SCLEROMYXEDEMA WITH INTRAVENOUS IMMUNOGLOBULIN: REPORT OF THREE CASES.

I Rosan [1] - C Y Maibashi [1] - J Martins [1] - D Miyashiro [1] - G A Martinez [2] - J A Sanches [1]

University Of São Paulo, Dermatology, São Paulo, Brazil [1] - University Of São Paulo, Hematology, São Paulo, Brazil [2]

Background: Scleromyxedema is a rare fibromucinous disorder of unknown origin that presents with generalized popular and sclerodermoid eruption. It is associated with the characteristic pathologic triad: dermal mucin accumulation, increased collagen deposition and fibroblast proliferation. This disease is typically associated with monoclonal gammopathy and may be associated with other systemicsymptoms, such as neurologic, cardiac, gastrointestinal, respiratory, and articular.
The monoclonal gammopathy is found in more than 88% of the patients, whereas the extracutaneous manifestations are found in over 60% of the cases. Some of these extracutaneuos manifestations may lead to great morbidity and mortality. Scleromyxedema tends to have a chronic and progressive course and it its spontaneous resolution is rare.
Because of its rarity, most information has been obtained from case reports or small series from the last decade, and only a few discuss its treatment. Therefore, it is difficult to obtain data about physiopathology, prognosis and therapeutics options. Reported therapies include intravenous immunoglobulin, thalidomide, autologous stem cell transplantation, systemic corticosteroids, with no defined standart treatment.

Observation: We report a case series of three female patients with scleromyxedema and paraproteinemia (one of them was also diagnosed with multiple myeloma). Important regression of cutaneous lesions and sustained response was achieved by intravenous immunoglobulin therapy with no side effects.

Key message: The current series supports the use of intravenous immunoglobulin as a safe and effective treatment for scleromyxedema, with good clinical responses and great improvement on patients quality of life.