Skleromyxedema - treatment options and comorbities

Bullet points

- Scleromyxedema is a skin disorder characterized by waxy, firm papules and plaques with mucin deposition, fibroblast proliferation, and fibrosis.
- It can also lead to extracutaneous manifestations affecting different organs, including neurologic, rheumatologic, cardiovascular, gastrointestinal, pulmonary, and renal manifestations, which can be fatal.
- Currently, there is no standardized method for grading the severity of cutaneous involvement and no effective treatment has been identified. Intravenous immunoglobulin (IVIG) is the preferred treatment.

Scleromyxedema is a skin disorder characterized by waxy, firm papules and plaques with mucin deposition, fibroblast proliferation, and fibrosis. The exact pathophysiology of this disease is unknown, but cytokines such as interleukin (IL)-1 and tumor necrosis factor-alpha (TNF-α), Th2-skewed T cell responses, as well as a deficiency of interferon-gamma (IFN-γ) and IL-17 may play a role. There is also conflicting data regarding the role of serum paraproteins in stimulating fibroblast proliferation and mucin overproduction. Other proposed theories include increased glycosaminoglycan synthesis and an adjuvant-induced autoimmune syndrome.

The condition presents with various skin symptoms, such as shiny and indurate skin, erythema, and edema. The mucous membranes are spared, and sclerodactyly and decreased motility of the mouth and joints may occur. It can also lead to extracutaneous manifestations affecting different organs, including neurologic, rheumatologic, cardiovascular, gastrointestinal, pulmonary, and renal manifestations, which can be fatal.

Currently, there is no standardized method for grading the severity of cutaneous involvement and no effective treatment has been identified. Intravenous immunoglobulin (IVIG) is the preferred treatment, but glucocorticoids and immunomodulatory drugs (e.g., thalidomide or lenalidomide) are also used. Autologous stem cell transplantation may be considered for poor responders. Other therapies like antibiotics, cyclosporine, phototherapy, and total skin electron beam therapy may improve skin symptoms but not systemic involvement.

Scleromyxedema is a chronic and progressive disease that requires long-term maintenance therapy. No definitive cure has been identified, and relapses commonly occur when treatment is discontinued.