SUCCESSFUL TREATMENT OF PROGRESSIVE CUTANEOUS SCLEREDEMA ASSOCIATED WITH INDOLENT MGUS: FROM « UNDETERMINED » TO CLINICALLY SIGNIFICANT GAMMOPATHY

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INTRODUCTION:

Scleredema of Buschke is a rare connective tissue disorder characterized by progressive, non-pitting skin induration caused by dermal mucin deposition separating collagen bundles. Type II scleredema, particularly associated with IgG subtype monoclonal gammopathy, most often follows the evolution of the underlying plasma cell dyscrasia.

OBSERVATION:

A 62-year-old woman came to our clinic for progressive skin tightening since 2012, involving the face, neck, arms, abdomen, and legs. Three biopsies were performed over time (Fig.1). Videocapillaroscopy remained normal. Laboratory results included elevated Electrocyte Sedimentation Rate (ESR) (26 mm/h), normal C-ReactiveProtein (CRP), polyclonal hypergammaglobulinemia (Ig 16 g/L; normal 8–13 g/L), and a minor IgG lambda and kappa component. Pulmonary function tests showed fluctuating lung diffusion capacities (DLCO 65–88%) and reduced total lung capacity (45–69%) over the years.

Numerous treatments failed or provided minimal benefit with clinical worsening over time. Bone marrow examinations revealed rising plasma cell percentages (Fig.1). Immunophenotyping in 2025 identified a small (0.16%) monoclonal lambda plasma cell population with abnormal phenotype (CD19−, CD56+, CD20−, CD27+ dim, CD81+, CD117−, CD200+), consistent with Monoclonal Gammopathy of Undetermined Significance (MGUS). Based on lab tests, PET-CT and bone marrow biopsy, no hypercalcemia, renal dysfunction, anemia, bone involvement (CRAB) criteria for multiple myeloma were present.[1]

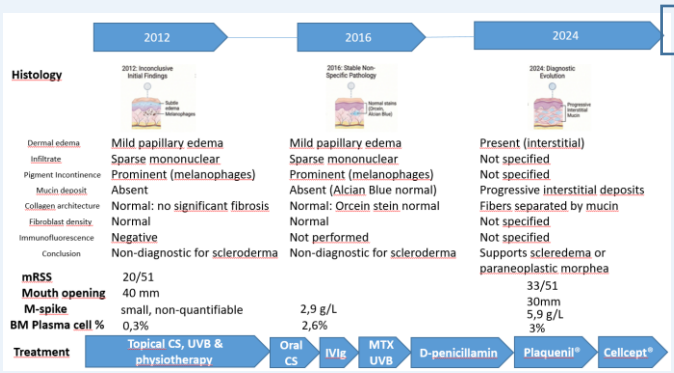

Figure 1. Clinical and histological evolution from 2012 to 2024. mRSS: modified Rodnan Skin Score

DIAGNOSIS: SCLEREDEMA OF BUSCHKE TYPE II ASSOCIATED WITH IgG LAMBDA MGUS

A multidisciplinary discussion persuaded the hematologists to initiate a myeloma-based treatment consisting of daratumumab, lenalidomide and dexamethasone. Four months later, impressive skin improvement was observed: mRSS fell to 1/51, mouth opening returned to 40 mm, and mobility normalized with no remaining limitations. The M-spike decreased to 0.7 g/L, indicating good therapeutic response of the underlying plasma-cell dyscrasia.

Improved mobility

Improved mRSS

DISCUSSION

- Exact pathophysiological mechanisms of type II scleredema remain incompletely understood: monoclonal immunoglobulin or plasma cell–derived cytokines may stimulate fibroblasts and promote excessive mucin production within the dermis
- Our patient fulfilled the diagnostic criteria for MGUS. Despite the lack of overt multiple myeloma, the progressive increase of the M-protein paralleled the worsening of the cutaneous disease, suggesting a biologically relevant relationship.
- When conventional dermatologic and immunosuppressive treatments failed, plasma cell-directed therapy provided dramatic and rapid clinical improvement supporting the causal link between the monoclonal plasma cell clone and dermal mucin deposition.

CONCLUSION

- MGUS may have clinical significance cutaneous manifestations impacting the quality of life of patients.
- It is crucial to investigate monoclonal gammopathy in patients with sclerodermiform-like lesions.
- Early multidisciplinary collaboration between dermatologists and hematologists is essential to optimize patient outcomes.

[1] International Myeloma Working Group. Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: a report of the International Myeloma Working Group. Br J Haematol. 2003;121(5):749-757.