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Treatment of scleromyxedema Arndt-Gottron with a novel intravenous immunoglobulin preparation

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Dear Editor,

Scleromyxedema is known as a rare, severe mucinosis with characteristic waxy skin papules along with sclerodermiform induration of the skin and rare systemic manifestations.[1] Monoclonal gammopathy of undetermined significance (MGUS; mostly monoclonal immunoglobulin G [IgG] type lambda) can be detected in up to 90% of patients.[1] Diagnosis of scleromyxedema is made according to the criteria of Rongioletti and Rebora.[2] Without adequate treatment, scleromyxedema may progress and take a severe and potentially lethal course.[1] First-line treatment involves the use of high-dose intravenous immunoglobulins (IVIgs).[1]

We report on a 36-year-old female patient with a histologically confirmed diagnosis of scleromyxedema Arndt-Gottron and monoclonal gammopathy IgG type lambda. She presented with disseminated skin-toned papules on the extremities (Figure 1); other symptoms included pruritus, edema, intermittent flushing, joint pain, and fatigue, impairing her ability to manage her job and daily activities. Therapy with high-dose IVIgs (Intratect® 100 g/L, Biotest Pharma GmbH) every four weeks was initiated in January 2019. Cutaneous symptoms and impaired joint mobility in the fingers gradually improved during therapy, and further progression of the disease was halted. Due to a lack of continued cost coverage, IVIg administration had to be paused for three months at the beginning of 2020, leading to a rapid worsening of the patient's symptoms. Disease improved only after resumption of IVIg therapy. A transient switch to subcutaneous immunoglobulins in March 2023 resulted in increasing fatigue, edema, loss of joint mobility, and poor tolerability, including local reactions at the injection site and abdominal pain, which demanded returning to IVIg administration at a dose of 2 g/kg every 4 weeks (Igymune® 100 mg/mL, LFB Biomedicaments S.A.). From July 2023, IVIg therapy at 4-week intervals (Intratect® 100 g/L, Biotest) was continued in our dermatology department. Although therapy was well tolerated, worsening of skin symptoms occurred (development of new indurated papules, fatigue, and mental impairment). Hence, from October 2023, IVIg was administered every three weeks. In January 2024, we decided to switch to the new IVIg formulation Yimmugo® (Biotest Pharma GmbH) at a dosage of 2 g/kg body weight administered over two days at 3-week intervals, which was well tolerated. The double-modified Rodnan skin score was 14 when Yimmugo® was started, then decreased to 6 in April 2024. Following two 4-week intervals, the double modified Rondan skin score increased to 17 in August 2024. After resuming 3-week intervals, it decreased to 8 in October 2024. Therapy with Yimmugo® every three weeks was continued, yet the patient complained about fatigue and new papules at the beginning of 2025; hence a hematological assessment was intended to evaluate additional therapy options.

For patients with scleromyxedema, we support the assessment of treatment response with the double modified Rodnan skin score, previously suggested by Mecoli *et al*.[3] In our patient, continuous IVIg therapy was necessary to achieve disease stabilization with an additional benefit from shorter therapy intervals. Individual adjustment of therapy intervals to patients' treatment response is feasible.

In general, it remains to be determined how IVIgs exert their effects in scleromyxedema. Some evidence suggests that IVIgs affect the process of matrix turnover and fibrosis by altering the levels of matrix metalloproteinases.[4-7] It is widely believed that the efficacy of different IVIg products is comparable, yet studies are missing. Commercially available IVIg products are distinguished by stabilizing agents (*e.g.*, glucose, sucrose, maltose, glycine, D-sorbitol, or L-proline). Sugar-free IVIg formulations are generally preferred in patients with renal insufficiency, as sucrose-containing products have been associated with an increased risk of renal dysfunction.[8,9] Different IVIg products differ with regard to individual tolerability.[10] Subcutaneous IVIgs may show fewer side effects than intravenous preparations. However, in our patient, these approaches were insufficient to achieve adequate disease control; therefore, a prompt return to IVIg therapy was required. At present, switching between IVIg preparations is primarily driven by availability or cost considerations, but it may also be a viable option in cases of adverse effects or reduced efficacy.[8] In our patient, transitioning between IVIg formulations was well tolerated and did not result in any adverse events or practical difficulties.

In conclusion, further reports and studies assessing the side effects and efficacy of various IVIg preparations in scleromyxedema are of interest.

References

1. Hoffmann JHO, Enk AH. Scleromyxedema. J Dtsch Dermatol Ges 2020;18:1449-67.
2. Rongioletti F, Rebora A. Updated classification of papular mucinosis, lichen myxedematosus, and scleromyxedema. J Am Acad Dermatol 2001;44:273-81.
3. Mecoli CA, Talbot CC, Fava A, et al. Clinical and Molecular Phenotyping in Scleromyxedema Pretreatment and Posttreatment With Intravenous Immunoglobulin. Arthritis Care Res (Hoboken) 2020;72:761-7.
4. Harper RA, Rispler J. Lichen Myxedematosus Serum Stimulates Human Skin Fibroblast Proliferation. Science 1978;199:545-7.
5. Ferrarini M, Helfrich DJ, Walker ER, et al. Scleromyxedema serum increases proliferation but not the glycosaminoglycan synthesis of dermal fibroblasts. J Rheumatol 1989;16:837-41.

6. Yaron M, Yaron I, Yust I, Brenner S. Lichen myxedematosus (scleromyxedema) serum stimulates hyaluronic acid and prostaglandin E production by human fibroblasts. J Rheumatol 1985;12:171-5.
7. Karim A, Lawlor F, Black MM. Successful treatment of scleromyxoedema with high dose intravenous immunoglobulin. Clin Exp Dermatol 2004;29:317-8.
8. Feldmeyer L, Benden C, Haile S, et al. Not All Intravenous Immunoglobulin Preparations are Equally Well Tolerated. Acta Derm Venereol 2010;90:494-7.
9. Chérin P, Cabane J. Relevant Criteria for Selecting an Intravenous Immunoglobulin Preparation for Clinical Use. BioDrugs 2010;24:211-23.
10. Ameratunga R, Sinclair J, Kolbe J. Increased risk of adverse events when changing intravenous immunoglobulin preparations. Clin Exp Immunol 2004;136:111-3.

Figure 1. Clinical presentation and histopathology. Disseminated skin-toned waxy papules up to a diameter of 3 mm in the left gluteal region (**A**) and on the left forearm (**B**). Compact stratum corneum, epithelium free from atypical cells, numerous interstitial round cells, fusiform cell elements between collagen fibers and dilated vessels (hematoxylin-eosin staining, **C**). Distinct mucin deposition in the upper dermis (periodic acid-Schiff staining, **D**).