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CASE REPORT

Scleredema in a patient with rheumatoid arthritis and monoclonal gammopathy of undetermined significance: case report

Escleredema em doente com artrite reumatoide e gamopatia monoclonal de significado indeterminado: caso clínico

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Abstract

Scleredema is a rare connective tissue disorder that typically causes non-pitting induration of the posterior neck and upper back. It most often follows upper respiratory tract infections and is also linked to diabetes and monoclonal gammopathies. Association with rheumatoid arthritis is rarely reported. We present a 63-year-old male with long-standing rheumatoid arthritis who developed progressive cervical and upper back induration over 12 months. Skin biopsy revealed thickened dermal collagen bundles separated by interstitial mucin with a mild superficial perivascular lymphocytic infiltrate. Blood tests identified an immunoglobulin G-κ monoclonal component consistent with monoclonal gammopathy of undetermined significance. He received intravenous immunoglobulin, phototherapy, and thalidomide with an unsatisfactory response. This case highlights scleredema in the setting of rheumatoid arthritis and monoclonal gammopathy of undetermined significance and the ongoing challenge of selecting effective therapy.

Keywords: Rheumatoid arthritis. Scleredema. Monoclonal gammopathy of undetermined significance.

Resumo

O escleredema é uma doença rara do tecido conjuntivo que cursa tipicamente com espessamento cutâneo não depressível da região cervical posterior e dorso superior. Associa-se mais frequentemente a infeções respiratórias superiores, mas está também relacionado com diabetes mellitus e gamopatias monoclonais. A associação com artrite reumatoide é rara. Apresentamos o caso de um homem de 63 anos com artrite reumatoide de longa data, que desenvolveu espessamento cutâneo progressivo ao longo de 12 meses, na região cervical e dorso superior. A biópsia cutânea revelou feixes espessados de colagénio dérmico separados por mucina intersticial, com discreto infiltrado linfocítico perivascular superficial. As análises identificaram um componente monoclonal IgG-κ consistente com gamapatia monoclonal de significado indeterminado. O doente realizou imunoglobulina endovenosa, fototerapia e talidomida, com resposta insatisfatória. Este caso destaca o escleredema no contexto de artrite reumatoide e gamapatia monoclonal de significado indeterminado, e o desafio terapêutico da patologia em questão.

Palavras-chave: Artrite Reumatóide. Escleredema. Gamapatia monoclonal de significado indeterminado.

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Introduction

Scleredema is a rare connective tissue disorder characterized by progressive, non-pitting skin thickening, preferentially involving the upper back, shoulders, and posterior cervical region.[1] It typically occurs in three clinical settings: post-infectious, most commonly following upper respiratory tract infections; in association with diabetes mellitus; and in the context of monoclonal gammopathies.[2] As the available literature consists mainly of case reports and small case series, no standardized treatment has been established. Reported therapeutic options include phototherapy,[3] intravenous immunoglobulin (IVIG),[4] and immunomodulatory agents such as thalidomide, all of which show variable responses. The coexistence of scleredema and rheumatoid arthritis (RA) has rarely been described in the literature.[5] We therefore report the case of a patient with long-standing RA and a recently identified monoclonal gammopathy of undetermined significance (MGUS) who developed scleredema.

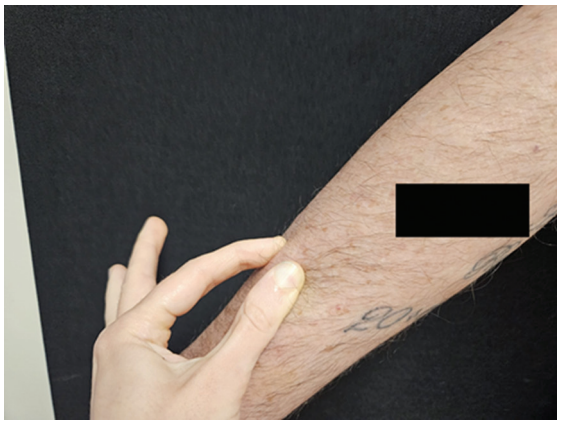

Figure 1. Non-pitting skin thickening and edema.

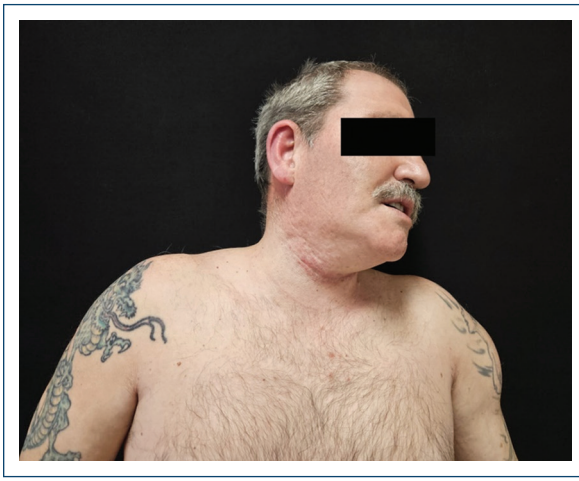

Figure 2. Limitation of cervical mobility due to skin edema and thickening.

Clinical case

We present the case of a 63-year-old male with an 11-year history of seronegative RA who was not receiving conventional medical treatment and was being followed exclusively with alternative therapies, with apparently adequate symptomatic disease control. He was referred to the dermatology department for progressive skin thickening with a 1-year duration. The patient also reported stiffness and marked limitation of cervical mobility. On physical examination, diffuse, symmetrical, non-pitting skin thickening was observed (Fig. 1), involving the posterior cervical region, the upper half of the trunk, the proximal segments of the upper limbs, and the face. A marked reduction in cervical flexion, extension, and rotation was also noted (Fig. 2), as well as limited abduction of both upper limbs (Fig. 3). The patient denied symptoms suggestive of systemic sclerosis, including Raynaud phenomenon, dysphagia, and dyspnea, and physical examination revealed no features such as sclerodactyly, digital ulcers, pitting scars, periungual or mat telangiectasias, pigmentary changes or microstomia. A skin biopsy revealed thickened dermal collagen bundles separated by interstitial mucin, with a mild superficial perivascular lymphocytic infiltrate (Fig. 4), findings consistent with a diagnosis of scleredema. Laboratory evaluation was unremarkable, except for the presence of a monoclonal peak in the gamma region on serum protein electrophoresis. Serum immunofixation identified an immunoglobulin G-κ (IgG-κ) monoclonal protein (IgG 1,987 mg/dL; κ light chains 545 mg/dL). The patient was referred to the hematology department, where a diagnosis of MGUS was established. Given the significant functional impairment, treatment with IVIG was initiated, and the patient completed 3 monthly cycles (2 g/kg per cycle), with poor clinical response. He subsequently underwent PUVA therapy twice weekly due to professional constraints, starting at 1.5 J/cm^2 with weekly increments of 0.5 J/cm^2 up to 3.5 J/cm^2, which was then maintained until completion of a total of 48 sessions, resulting in a slight improvement in cervical and upper limb mobility. Given the suboptimal clinical response to prior therapies, thalidomide was initiated. This decision was supported by its immunomodulatory and anti-fibrotic properties.

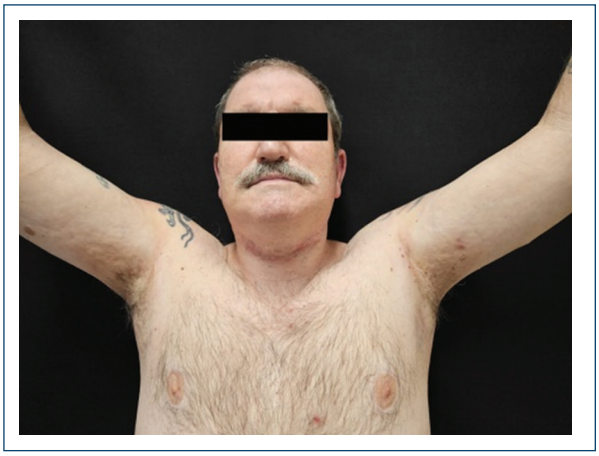

Figure 3. Limitation of upper limb abduction due to skin edema and thickening.

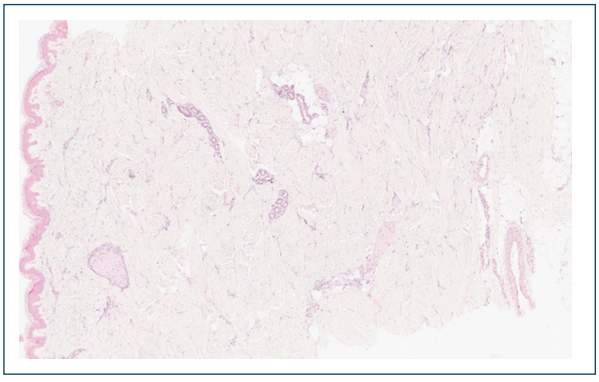

Figure 4. Thickened dermal collagen bundles with mucin deposition and a mild superficial perivascular lymphocytic inflammatory infiltrate.

Although evidence remains limited to case reports, thalidomide has been described as a potential therapeutic alternative in refractory cases, especially in the context of paraproteinemia-associated disease.[6] In this patient, thalidomide was initiated at a dose of 50 mg daily, with a suboptimal clinical response at the most recent evaluation.

Discussion

Scleredema is a rare entity, but it is likely underdiagnosed. It most commonly occurs in association with diabetes mellitus or following upper respiratory tract infections, but it may also be associated with monoclonal gammopathies and, less frequently, with autoimmune diseases[1]. In the present case, the presence of an IgG-κ monoclonal gammopathy represents the most well-established associated condition. Although the patient had long-standing untreated RA, its role in the development of scleredema is less clearly defined. Nevertheless, previous reports have described an association between RA and scleredema, suggesting that it may also have acted as a contributory factor in this case.[5] Therapeutic evidence remains limited to case reports and small case series. Phototherapy has been shown to improve skin thickening and, consequently, mobility;[3] IVIG has demonstrated variable benefit and immunomodulatory agents such as thalidomide are used selectively. In our patient, IVIG provided minimal clinical improvement, PUVA resulted in only modest benefit, and low-dose thalidomide had an unsatisfactory effect, highlighting the heterogeneity of responses to available treatments. The coexistence of RA and MGUS may have influenced the response to the different therapeutic approaches employed. Given the rarity of this condition, further studies are needed to better evaluate the efficacy of the various therapeutic options across the different subtypes of scleredema.

Conclusion

This case highlights the diagnostic and therapeutic challenges of scleredema associated with MGUS and long-standing rheumatoid arthritis. The limited response to multiple treatment modalities underscores the need for individualized management and further studies to establish evidence-based therapeutic strategies for this rare condition.

Funding

None.

Conflicts of interest

None.

Ethical considerations

Protection of human subjects and animals. The authors declare that no experiments on humans or animals were performed for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from all patients, and secured approval from the Ethics Committee. SAGER guidelines have been followed as applicable to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that ChatGPT was used for grammatical correction.

References

1. Hong KR, Hong JY, Chung EH, Lee SH, Lee SW, Kim JE. Monoclonal gammopathy-associated scleredema adultorum of Buschke in a patient with diabetes mellitus successfully treated with intravenous immunoglobulin and narrow-band ultraviolet B phototherapy: a case report. Ann Dermatol. 2021;33:586-8.
2. Pereira M, Ramos Pinheiro R, Lencastre A, Bártolo E. Scleredema diabeticorum. Dermatol Rep. 2022;14:9477.
3. Yoshimura J, Asano Y, Takahashi T, Uwajima Y, Kagami S, Honda H, et al. A case of scleredema adultorum successfully treated with narrowband ultraviolet B phototherapy. Mod Rheumatol. 2016;26:302-6.
4. Eastham AB, Femia AN, Velez NF, Smith HP, Vleugels RA. Paraproteinemia-associated scleredema treated successfully with intravenous immunoglobulin. JAMA Dermatol. 2014;150:788-9.
5. Hartley K, Huang C, Sykes A, Lamont D, Wlodek C. Scleredema in the setting of rheumatoid arthritis. Australas J Dermatol. 2020;61: e110-1.
6. Barnes M, Kumar V, Le TH, Nabeel S, Singh J, Rana V, et al. A case of paraproteinemia-associated scleredema successfully treated with thalidomide. JAAD Case Rep. 2020;6:1039-41.