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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

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Dear Editor:

Scleredema is a rare connective tissue disease characterized by the deposition of collagen and mucin, mainly involving upper back and posterior neck. Skin findings include non-pitting edema, skin hardening, and movement limitation[1]. It is categorized based on underlying diseases, such as streptococcal infection (type 1), monoclonal gammopathy (type 2), and diabetes mellitus (DM; type 3). Other diseases including primary hyperparathyroidism, ankylosing spondylitis, Sjögren syndrome, and dermatomyositis have also been described to be associated[2]. Treatment options including immunosuppressive agents, antibiotics, systemic glucocorticoids, and phototherapy have been tried, but no standard treatment protocol has yet been established[1].

A 53-year-old male presented with a 1-year history of progressive skin hardening on the posterior neck and upper back. He also reported decreased range of motion. Physical examination revealed non-pitting induration and erythema on the posterior neck and back (Fig. 1A, B). He had hypertension, obesity, and poorly controlled type 1 DM. There was no history of a preceding infection. A skin biopsy performed on his back showed thickened collagen bundles separated by clear spaces in the reticular dermis (Fig. 2). Laboratory tests including a com-

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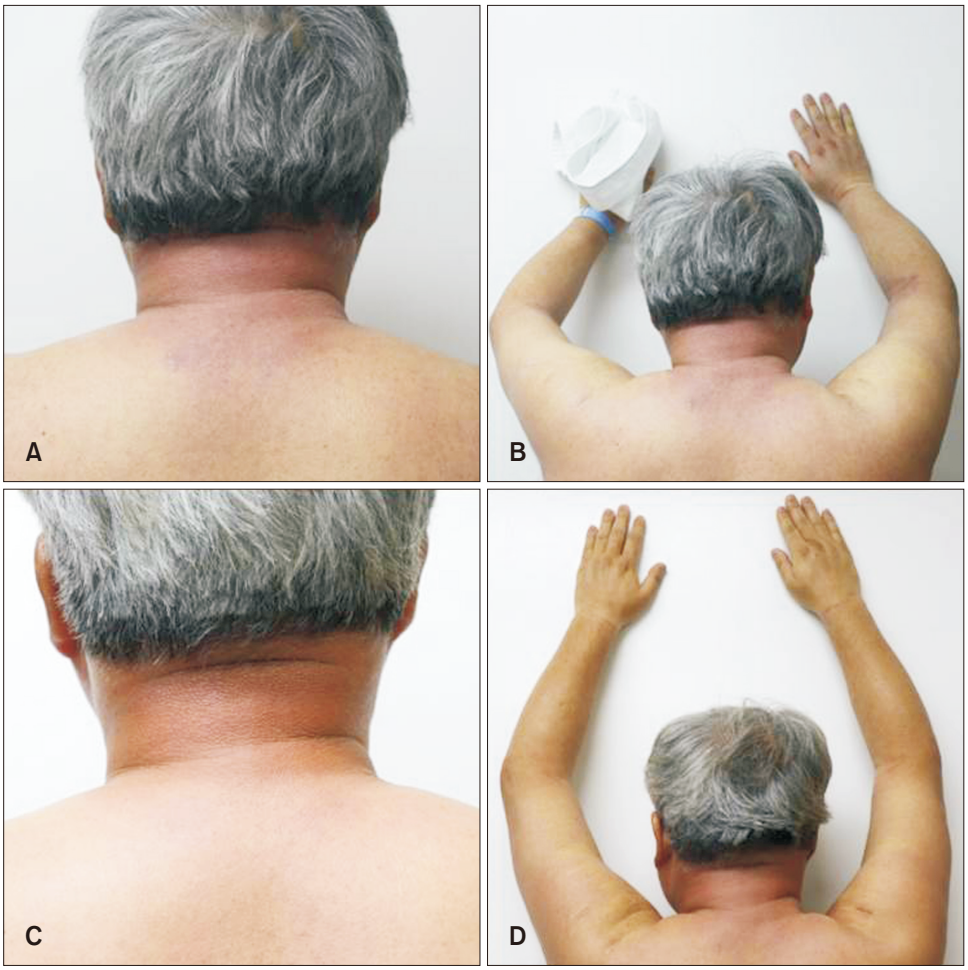

Fig. 1. Before intravenous immunoglobulin (IVIG) treatment, non-pitting edema, hardening of the skin and limited motion on the posterior neck (A) and both shoulders (B). After 4 cycles of IVIG treatment, improvement with motion on the posterior neck (C) and both shoulders (D).

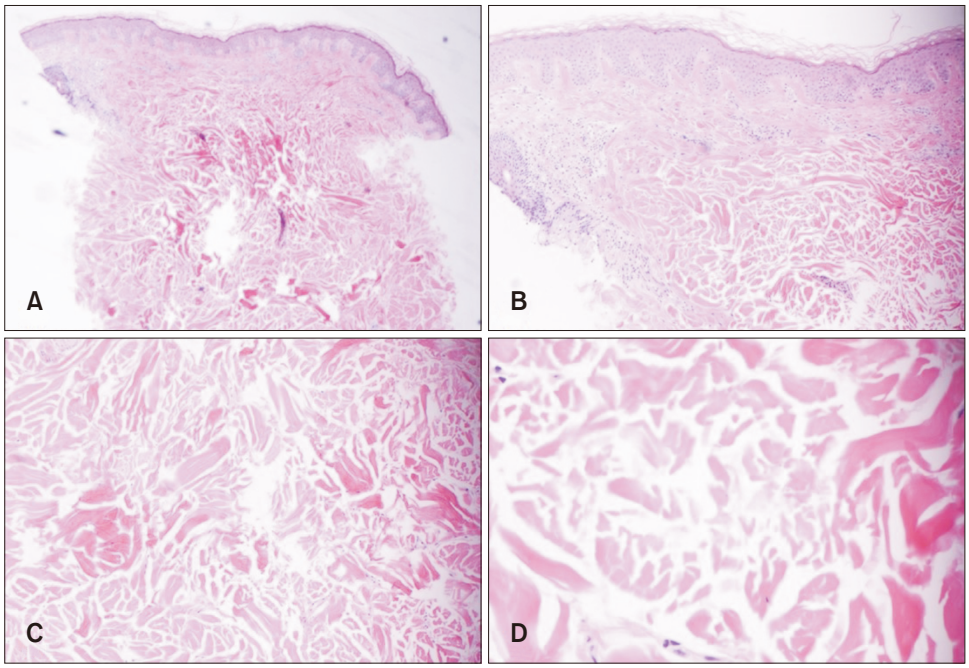

Fig. 2. Histologic figures of the specimen taken from the back. (A) Normal finding in epidermis, thickened collagen bundles in dermis (H&E, original magnification ×40). (B) Thickened collagen bundles in dermis (H&E, ×100). (C) Thickened collagen bundles and fenestration caused by mucin deposits in dermis (H&E, ×200). (D) Fenestration caused by mucin deposits in dermis (H&E, ×400).

plete blood cell count and a chemistry panel revealed elevated glucose (240 mg/dl) and mild elevated liver enzymes. Plasma electrophoresis and immunofixation showed a monoclonal peak in the gamma globulin region, suggesting monoclonal gammopathy with immunoglobulin G κ. Bone marrow biopsy and skeletal examinations for multiple myeloma were negative. The diagnosis of scleredema was made.

He was first treated with narrow-band ultraviolet B (NB-UVB), phototherapy twice a week and methotrexate 10 to 20 mg per week for 4 months. However, skin induration and range of motion worsened. Monthly intravenous immunoglobulin (IVIG) treatment was initiated at a dose of 2 g/kg over 5 consecutive days combined with NB-UVB phototherapy. After 4 cycles, he noted significant improvement. After 10 cycles, skin stiffness continued to improve without any side effects. External rotation and internal rotation of shoulders also significantly improved (Fig. 1C, D). At the latest follow-up, the patient is maintaining monthly IVIG treatment combined with NB-UVB phototherapy with showing continuous improvement.

Treatment of scleredema remains a challenge, especially when associated with diabetes or gammopathy, which tends to have a protracted clinical course and resistance to many therapies. NB-UVB phototherapy may affect keratinocyte-fibroblast interaction to inhibit fibroblast activation[3]. IVIG has been reported with successful response in some cases[4,5]. It is suggested that it may reduce glycosaminoglycans in the skin by acting on the Fc receptor and regulating innate and adaptive immunity. Furthermore, it may suppress proinflammatory cytokines and exert immunomodulatory effects by neutralization of superantigens[5].

To our knowledge, five previous reports described successful IVIG treatment in scleredema[3-5]. However, there were no reports of concurrent monoclonal gammopathy and type 1 DM associated scleredema. Herein, we described a first case of severe, progressive scleredema accompanied by both monoclonal gammopathy and type 1 DM, which showed significant improvement with IVIG and NB-UVB phototherapy. Further studies are needed to explore the exact therapeutic effects of each IVIG and NB-UVB treatment on various types of scleredema.

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CONFLICTS OF INTEREST

The authors have nothing to disclose.

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