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LETTER TO THE EDITOR

Scleredema accompanied by IgG-λ monoclonal gammopathy

Dear Editor,

Scleredema is a rare fibromucinous connective tissue disease, firstly reported by Buschke in 1902[1], and is characterized by thickening and hardening of the skin on the face, neck, and trunk. Although the etiology is unknown, it is revealed that scleredema is related to infection, diabetes mellitus, and paraproteinemia.[2] Here, we report a rare case of scleredema accompanied by IgG-λ monoclonal gammopathy.

A 53-year-old Japanese man presented with a 20-year history of progressively hardened skin of the neck, upper arms, and upper trunk. He had no history of diabetes mellitus or preceding infection. Physical examination showed a symmetrical induration and nonpitting edema of the neck, upper trunk, and upper arms (Figure 1A,B). Raynaud's phenomenon or sclerodactyly was not observed. A skin biopsy specimen obtained from the upper back showed remarkably thickened dermis with separation of enlarged collagen bundles (Figure 1C,D). Slight deposition of mucin was seen between the collagen bundles (Figure 1E). Laboratory evaluation showed normal complete blood cell counts and comprehensive metabolic panel. The patient's antinuclear antibody and autoantibodies to centromere, topoisomerase 1, RNA polymerase III, and RNP were all negative. His hemoglobin A1c level

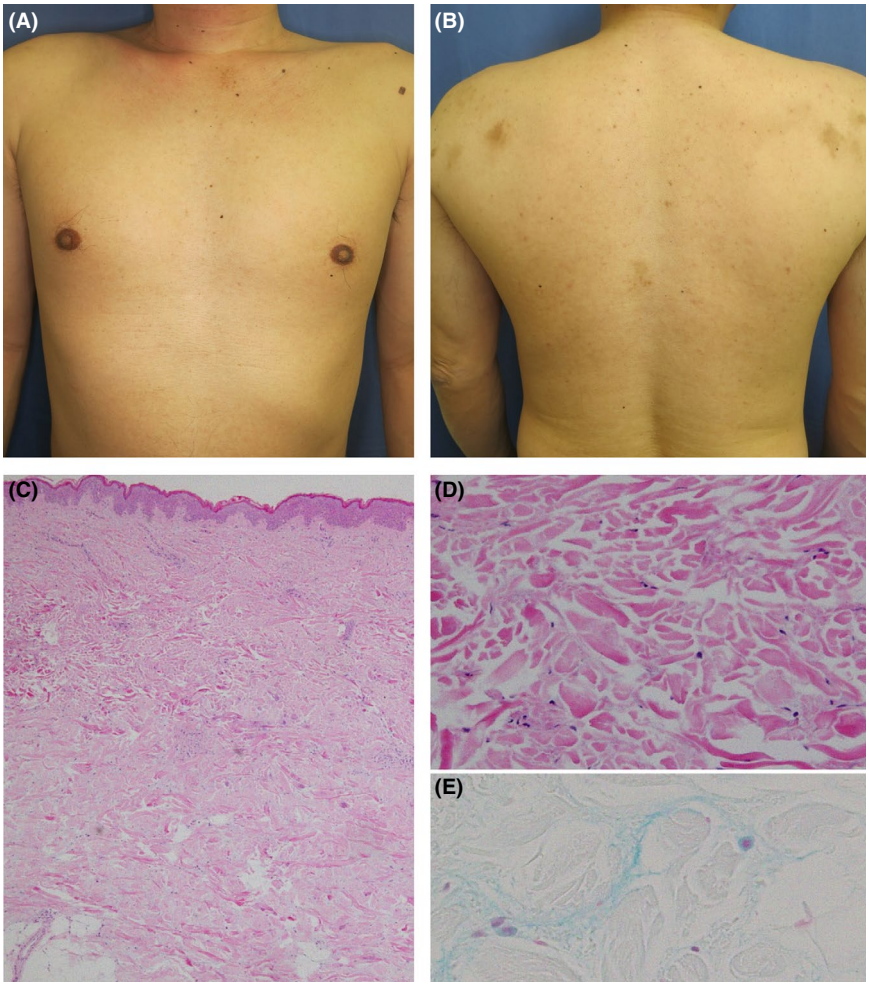

FIGURE 1 Clinical and histopathological features. A,B, Thickening and hardening of upper trunk. C,D, Hematoxylin-eosin-stained biopsy specimen from the upper back, showing thickened dermis with large collagen bundles that appear separated from one another (original magnifications: [B] ×40; [C] ×200). E, Alcian blue staining showing interstitial mucin deposition (original magnifications ×400)

was normal. Electrophoresis test of serum protein revealed an abnormal peak of γ-globulin. The serum IgG level was elevated (2184 mg/dL; normal range, 861-1747 mg/dL), and serum immunoelectrophoresis confirmed the presence of monoclonal protein (M-protein) of an IgG-λ band. Bone marrow aspiration negated any abnormalities in myeloid cells. Based on these findings, we diagnosed his condition as scleredema accompanied by IgG-λ monoclonal gammopathy, which was considered as monoclonal gammopathy of undetermined significance.

In previous studies, scleredema is classified into three types. The first group (the classic type described by Buschke, 55% of cases) has a preceding illness (particularly upper respiratory tract infection) and resolves in several months to years. The second group is associated with paraproteinemia, multiple myeloma, and amyloidosis. This group makes up about 25% of cases of scleredema. The third group (20% of cases) has been called diabetic scleredema which has association with diabetes mellitus.[2] According to Rongioletti, five cases (11%) in 44 patients of scleredema were accompanied by monoclonal gammopathy and the most common comorbidity were metabolic diseases particularly type 2 diabetes.[3] IgG, particularly IgG-κ, paraprotein is the commonest in patients with scleredema associated with paraproteinemia. Half of the patients with paraproteinemia developed myelomatous disorder including multiple myeloma.[4]

Several treatments have been reported, including phototherapy, systemic glucocorticoids, cyclosporine, and methotrexate.[2] Phototherapy (UVA1 or PUVA) is the common treatment although it shows partial response.[3] Furthermore, it is reported that the treatment of intravenous immunoglobulin was successful for a case of scleredema with paraproteinemia.[5]

There is no established standard therapy for scleredema, and further studies are required. Scleredema may appear as a skin symptom of diabetes or monoclonal gammopathy and leads to early diagnosis of the underlying disease. It is kept in mind that we consider scleredema with infection, diabetes mellitus, and paraproteinemia as a diagnosis of hardened skin of the neck and upper trunk.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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