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CASE REPORT

A 34-year-old man presented progressive generalized hardening of the body for 3 years. Three months ago, he experienced difficulty in swallowing and extending his tongue, and several hard nodules were noted progressively on the forehead (Figure 1A). Physical examination revealed that the skin on his face and distal extremities was indurated (Figure 1B). On the trunk and proximal extremities, the skin was normal in elasticity, but the underlying muscles were indurated, and almost all the joints were restricted in motion. No regional lymph nodes were palpable. Laboratory examination showed no abnormality in routine tests. Serum and urine immunofixation showed a characteristic pattern of monoclonal IgG gammopathy, particularly of λ paraprotein. Some cardiac abnormalities including left ventricular outflow obstruction and right ventricular dysfunction were noted upon echocardiography. Positron emission tomography computed tomography (PET-CT) scan showed multiple areas of abnormal uptake in almost all the joints and some muscles. Bone marrow aspirate revealed active myeloid proliferation and 3% of plasmacyte observed. Flow cytometry showed that $CD38^{birght}$ $CD45^{dim/-}$ cells occupied 6.5% of all the nucleated cells and the expression of antigen was CD19 (0.7%), CD56 (1.9%), CD20 (1.7%), CD54 (99.9%), CD138 (99.8%), CD49e (2.6%), intracytoplasmic IgM (3.5%), intracytoplasmic IgD (0.5%), intracytoplasmic IgG (12.6%), intracytoplasmic κ paraprotein (0.3%) and intracytoplasmic λ paraprotein (98.5%). There were no remarkable findings in various other studies, including abdominal ultrasonography and chest computer tomography (CT) scan. The skin and muscle biopsy specimens obtained from the nodule on the forehead and inner thigh showed a deposition of amyloid materials in the dermis, subcutaneous tissues, vessel walls and connective tissue sep-

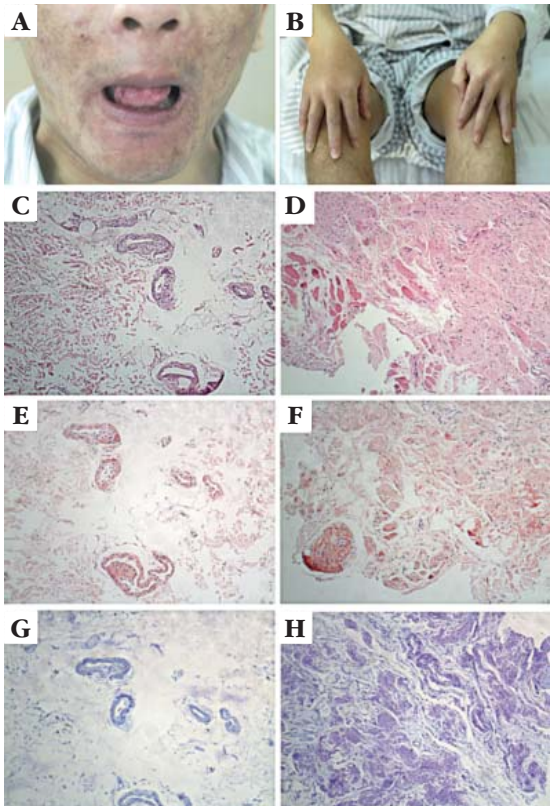

FIGURE 1: **A.** Indurated skin on the face and macroglossia; **B.** Indurated skin on the distal extremities; **C.** HE: deposition of amyloid materials in the dermis and vessel walls; **D.** HE: deposition of amyloid materials in the muscles; **E.** Red Congo stain highlighting the amyloid substance in the vessel walls; **F.** Red Congo stain highlighting the amyloid substance in the muscles; **G.** Crystal violet stain revealing the amyloid substance in the vessel walls; **(H)** Crystal violet stain revealing the amyloid substance in the muscles

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tae of the skeletal muscle, by Congo red and crystal violet stain (Figure 1C, 1D, 1E, 1F, 1G and 1H). The diagnosis was immunoglobulin light chain (AL) amyloidosis with scleroderma-like manifestation.

DISCUSSION

Amyloidosis refers to a heterogeneous group of disorders characterized by extracellular deposition of proteinaceous fibrillar materials (termed amyloid) in various tissues and organs. Clinically, the most commonly affected organs resulting in symptoms are the heart, kidneys, skin, peripheral nerves, autonomic nerves, and liver. Their respective means of presentation are: 1) normal ejection fraction with diastolic dysfunction, left ventricular hypertrophy with low voltage electrocardiogram; 2) nephrotic syndrome with preserved glomerular filtration rate; 3) purpura, most notably around the eyes and neck, as well as macroglossia; 4) small fiber peripheral neuropathy characterized by dysesthesia; 5) orthostatic hypotension; 6) hepatomegaly, often with a cholestatic rise in liver function tests.[1] Mucocutaneous alterations are present in 20-40% of cases, frequently as the first signs of amyloidosis.[2] In this patient, however, it has rarely been seen only with scleroderma-like skin manifestation.

At present, all treatments are aimed at destroying the underlying plasma cell clone, which, in turn, reduces or eliminates the amyloidogenic clonal immunoglobulin light chain. Currently, there are no approved drugs that directly attack and/or dissolve the amyloid. The approach of using molecules and/or antibodies directed against serum amyloid protein, or antibodies directed at the tertiary structure of the amyloid, may be treatments for the future.[3] ❑

Abstract: Immunoglobulin light chain amyloidosis is the most common acquired systemic amyloidosis. Its presentation is often insidious and progressive, which may delay diagnosis. The authors describe a rare case of immunoglobulin light chain amyloidosis in a 34-year-old man with scleroderma-like manifestation substantiated by multifarious laboratory investigations and the histopathologic feature of involved skin lesions stained with Congo red and crystal violet. This helps to maintain a high clinical suspicion of the disease when confronting similar skin presentation.
Keywords: Amyloidosis; Scleroderma, diffuse; Skin manifestations

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