

Multiple Myeloma-associated Light Chain Amyloidosis Presenting as Verrucous Lingual Masses

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Sir,

Multiple myeloma is a monoclonal B-cell neoplasm, which is characterized by autonomous proliferation of immunoglobulin-secreting plasma cells, capable of synthesizing amyloidogenic light chains (AL). AL amyloidosis is characterized by extracellular deposition of eosinophilic, homogeneous, amorphous material composed of fibrillary protein, containing light chain fragments (1). The oral clinical features of patients with AL amyloidosis include papules, nodules, plaques and macroglossia (2). To our knowledge, verrucous masses on both lateral sides of the tongue, without macroglossia, have not been described in the literature so far. Herein, we report such a case.

CASE REPORT

A 68-year-old man presented with waxy verrucous masses on both lateral sides of the tongue for 6 months. He also complained of pain in both hip joints and back for 5 months. An examination of the oral cavity revealed dark reddish verrucous masses on both lateral sides of the tongue without macroglossia (Fig. 1A). The biopsy specimen from lingual mass revealed eosinophilic amorphous materials in the dermis and fibromuscular tissue. The deposited materials gave a positive result with Congo red stain (Fig. 1B) and apple-green birefringence on polarized microscopy. Electron micrograph of dermal deposits showed randomly oriented, fine, rigid, non-branching fibrils of variable length. Twenty days later, he visited the emergency room due to abdominal pain

accompanied by vomiting and constipation which had lasted for 7 days. Abdominal computed tomography scan and barium enema revealed mild obstruction of the descending colon, and multiple erythematous patches were seen in the rectum and sigmoid colon by colonoscopy. The biopsy specimen from the colonic lesion revealed the deposition of pinkish amorphous material in the dermis, which stained positively with Congo red. Laboratory data revealed mild normochromic, normocytic anaemia (Hb 9.4 g/dl, Hct 27.0%), and the albumin-globulin ratio (2.8:3.6) was reversed. On bone marrow biopsy, plasma cells occupied 50% of the proportion. Serum electrophoresis showed increased beta-globulin fraction, and immunoelectrophoresis disclosed λ -type light chain disease. Urine electrophoresis showed M-peak protein in gamma area, and immunoelectrophoresis was positive for λ chain. On skull X-ray, several lytic lesions were seen. Finally, the patient was diagnosed with multiple myeloma-associated AL amyloidosis and received four cycles of chemotherapy consisting of vincristine, doxorubicin and dexamethasone. The patient showed low response to the chemotherapy and died 5 months after the diagnosis of amyloidosis.

DISCUSSION

AL amyloidosis is composed of insoluble fibrillar protein, amyloid with subsequent severe tissue dysfunction (3). AL is associated with multiple myeloma, monoclonal gammopathy, or macroglobulinaemia (4, 5). Serum amyloid A amyloidosis results from chronic

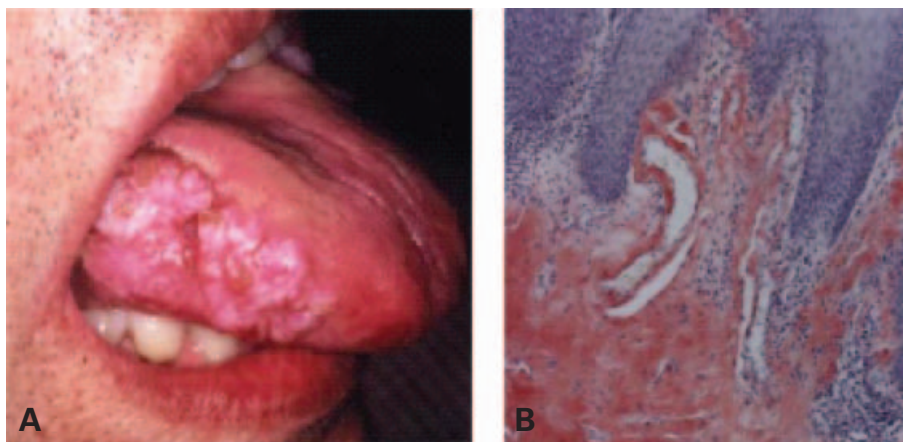


Fig. 1. (A) Waxy verrucous masses along the lateral sides of the tongue. (B) Dermal deposits showed a positive result with Congo-red stain (magnification $\times 100$).

inflammatory diseases such as Crohn's diseases, tuberculosis, rheumatoid arthritis, etc.

Amyloid fibrils in patients with multiple myeloma are derived from monoclonal immunoglobulin light chains termed Bence Jones proteins. The actual mechanism of deposition of Bence Jones proteins in the extracellular compartment is not completely understood (6).

The oral manifestations have been reported in 39% of patients with AL amyloidosis (6). The tongue is the most affected site and may be enlarged in all directions and completely fill the mouth, or in extreme cases it can extrude from the mouth at rest (6, 7). The tongue may be firm, dry, stony, hard, fissured, ulcerated, haemorrhagic and painful with or without lateral tooth indentation. In 1994, Reinisch et al. (8) compiled a summary of patients with oral amyloidosis as a result of multiple myeloma. In this literature, most of the patients presented macroglossia except one (8). To our knowledge, verrucous masses on both lateral sides of the tongue without macroglossia have not so far been described in the literature.

When the diagnosis of amyloidosis is made in patients with multiple myeloma, the survival time is shorter than without amyloidosis, and these patients deteriorate rapidly (6, 9). The median survival time is about 4 months (10).

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