

AMYLOIDOSIS: REPORT OF A CASE WITH AN EARLY  
AND EXTENSIVE INVOLVEMENT OF  
TONGUE

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INTRODUCTION

Amyloidosis is an irreversible disease, with unknown etiology, characterized by the deposition of amyloid in various tissues of the body. Amyloid is a complex material, generally thought to be a mixture of protein, glycoprotein, other polysaccharides, and possibly lipids (1).

There are several major clinical forms of amyloidosis that should be considered. There are the primary type, the closely related form of amyloidosis associated with multiple myeloma and the type that is associated with chronic infection and various other diseases. Classically, the primary and the myeloma associated varieties of amyloidosis involve muscles, especially in the tongue, heart, and gastrointestinal tract, whereas the secondary type affects principally the kidneys, spleen, and liver (2). However, experience with a large number of patients has shown that this sharp division is no longer valid,

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since involvement of all the organ systems in both types of amyloidosis has been documented (3,4).

Amyloid deposition in the tongue, resulting in macroglossia, and gingiva is also reported to be commonly seen. Pain in the jaw may be a complaint of these patients and usually is the result of amyloid deposition in the masseter muscles (2). The involvement of lacrimal as well as salivary glands decreases the glandular function, and the amyloid-induced xerophthalmia or xerostomia may be the initial symptoms and major source of discomfort.

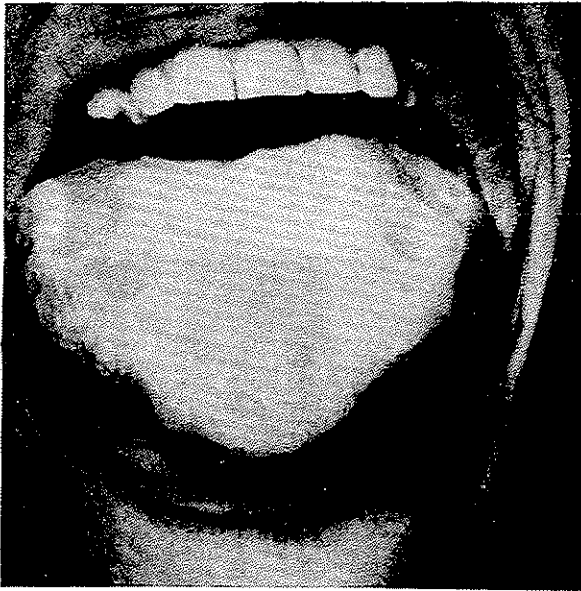
#### CASE REPORT

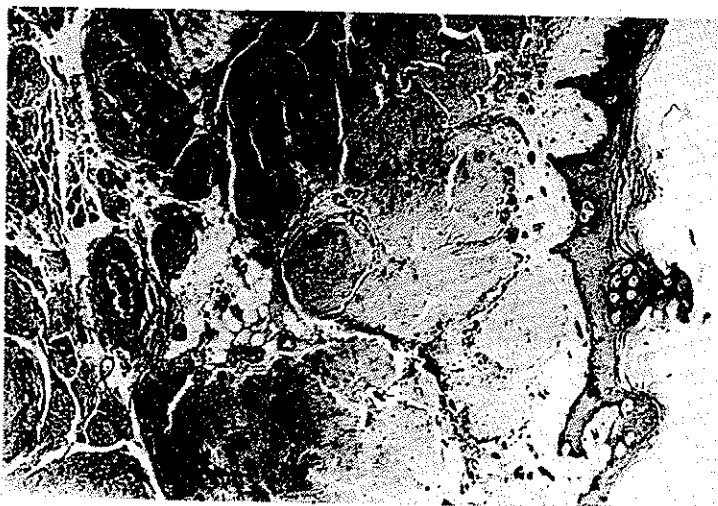
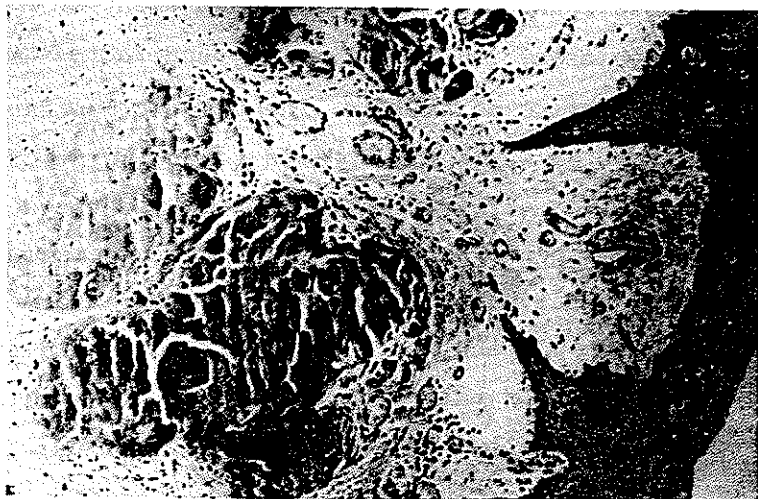
The patient is a 61-year-old white female who apparently was in a good state of health until she felt the gradual onset of pain, numbness, and tingling over the right median nerve distribution. In 1980 a right carpal tunnel syndrome was found and decompressed at Boston City Hospital. Later she noted that her tongue was enlarging and she was frequently biting it.

A biopsy of the enlarged tongue revealed "amyloidosis". (Fig. IA, B,C) At this time cervical arthrosis has been found, and also the decompression of the left carpal tunnel syndrome was done. Extensive workup of her amyloidosis disclosed: +ANA 1:2, Homogenous, ESR; 58 and IgG; 1640 mg%. Urine analysis showed proteinuria, and bone marrow biopsy revealed 15-20% plasma cell, which was not diagnostic of multiple myeloma, but a repeated biopsy was consistent with multiple myeloma. A decrease in pulmonary vital capacity was found, and the chest x-ray showed bilateral diffuse nodular involvement of the lungs. An upper GI x-ray revealed a thicken-

ing of mucosa consistent with the involvement in amyloidosis. Biopsies of skin sural nerve, and rectal mucosa all showed deposits of amyloid. The patient continued to have dyspnea as well as dysphagia to the point of being unable to swallow her food. At this time the tongue showed 4+ enlargement and was very firm and erythematous.

(Fig. 2) Bilateral partial glossectomy was performed for palliation. The patient had a downhill course and finally died of C.V.A. In this case the course of the disease was 4 years.







#### DISCUSSION

A severe multisystem amyloidosis with excessive workup was been presented. This presentation is an emphasis on the unique position of a dentist in the early diagnosis of severe systemic disorders which involve primarily the oral structures. In the case report we indicated that the enlarging tongue was one of the earlier signs of the disease, and was sufficient evidence to warrant, if not a diagnosis, at least a suspicion of a systemic disorder. We should suspect all of the abnormalities in the oral cavity which cannot be explained by the focal bases. An enlarged lymph node, unexplained jaw pain, ulcer, discoloration of soft tissue over-growth, abnormal x-ray presentation, petechia, hemorrhage, xerostomia, burning sensation, and many other signs and symptoms may be the initial and sometimes the only onset of the

severe systemic involvement. A good oral examination as well as evaluation of the patient's history may lead to an early diagnosis of many diseases and lengthening of the patient's life.

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