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Letter to the Editor

AMYLOIDOSIS OF THE TONGUE AS-AN INSIGHT VIEW OF A SYSTEMIC DISEASE

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Amyloidosis represents a heterogeneous cluster of disorders characterized by abnormal extracellular deposition of insoluble fibrillar proteinaceous materials. Three forms of amyloidosis are defined by the presence or absence of systemic disease: primary systemic amyloidosis, secondary systemic amyloidosis, and localized amyloidosis (1).

Primary systemic amyloidosis is a condition with an unknown underlying cause, different from secondary systemic amyloidosis, which occurs associated with other known diseases, such as tuberculosis, rheumatoid arthritis, and mainly multiple myeloma (1). Localized amyloidosis consists of a nodular amyloid deposit mass without association with a systemic disease (2). Although the mean survival of patients with systemic forms is between 5 to 15 months, those with the localized form have an excellent prognosis (2).

The head and neck region is affected in about 12–90% of the cases, typically involving the larynx and tongue (2). Although oral amyloidosis involves the tongue, buccal mucosa, and gingivae, tongue amyloidosis (TA) is a rare and benign disease (3). The most reported features of TA are multiple soft nodules accompanied by yellowish, red, blue, or purple colour changes in the mucous membrane. TA may be linked to systemic disease. A complete systemic workup for amyloid is necessary in the case of TA because this can markedly change the expected morbidity and mortality. To the best of our knowledge, only 53 well-documented cases of TA have been previously described in the English-language literature (3).

TA most commonly affects individuals between 50 and 70 years of age, with a male-female predominance of 3:1 to 3:2. The mean age of the patients with TA is 57.5 years (range 10–90 years), with a female predilection (3).

TA typically results in macroglossia, manifested by increased tongue volume, tongue protrusion beyond the alveolar ridge, speech impairment, and dysphagia. In TA, yellow nodules or raised white lesions occurring predominately along the lateral border are also common; the mucosal surface is usually intact, and the underlying lesion may be nodular or flat, with a yellow, pink, or bluish colour.

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The diagnosis of TA can be easily made with a biopsy. Criteria for diagnosis of TA include eosinophilic extracellular deposits of protein fibrils that exhibit apple-green birefringence on polarized light microscopy when stained with Congo red (4). Once the diagnosis has been made, an extensive workup for systemic amyloidosis should be undertaken, including an abdominal fat or rectal biopsy. These tests are positive in 75% to 90% of patients with systemic involvement. A bone marrow biopsy may be performed to rule out plasma cell dyscrasia. Urine and serum electrophoresis is necessary to detect the presence of a monoclonal paraprotein composed of amyloid light chains. Serum or urine "Bence Jones proteins" will be found in up to 88% of patients with primary systemic amyloidosis and 100% with multiple myeloma-associated systemic amyloidoses. An echocardiogram should be done to evaluate the myocardium for signs of amyloidosis. Other tests may be useful in evaluating amyloidosis, as well as dynamic magnetic resonance imaging, Tc-99m phosphate radionuclide imaging, and 123I serum amyloid P scintigraphy.

There is a broad differential diagnosis with TA that should be considered in patients presenting with macroglossia or nodular tongue lesions. The differential diagnosis for generalized macroglossia, tuberculosis, lymphangioma, hypothyroidism, acromegaly, lingual infarction caused by giant-cell arteritis, idiopathic muscular hypertrophy, and Beckwith-Wiedemann syndrome should be considered. Furthermore, TA differential diagnosis must be considered with other nodular lesions such as fibroma, lipoma, granular cell tumor, sarcoma, and salivary gland tumors (5).

There is still no consensus regarding the management of TA, although numerous therapies have been proposed, including surgical excision and pharmacological treatment. Unfortunately, however, lesions often persist or recur. In addition, the prognosis is uncertain due to the rarity of the condition, requiring regular follow-up and monitoring (5).

In summary, TA is rare. Despite this, TA should be considered in the differential diagnosis of multiple or single yellowish nodules in the oral cavity.

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