

## CASE REPORT

# Localized Amyloidosis of Tongue: A Rare Case from India

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## ABSTRACT

Involvement of the tongue by amyloid is mostly secondary to systemic amyloidosis, with less than 9% of all types of amyloidosis presenting as isolated amyloidosis of the tongue. This is a case report of a 56-year-old patient who presented with mass lesion in the posterior part of tongue since 2 years duration. Magnetic resonance imaging revealed nodular area on posterior aspect of tongue without focal signal intensity and no contrast enhancement. On histopathological examination, the lesion exhibited homogeneous extracellular eosinophilic material. Congo red staining revealed amyloid material as red color under light microscopy and as apple green birefringence under polarized microscope. A definitive diagnosis of localized isolated amyloidosis of tongue was made after detailed workup to rule out the systemic form of amyloidosis. Isolated and localized amyloidosis of tongue is very rare and often mimics benign tumors, thus making the job of the treating doctor more challenging. The foremost task in management of these patients is to rule out the possibility of systemic amyloidosis because the localized forms of the disease may be treated with wide local surgical excision.

**Keywords:** Amyloidosis, Apple green birefringence, Polarized light, Tongue.

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## INTRODUCTION

Amyloidosis is a disorder defined by the deposition of proteinaceous material in extracellular areas. These deposits have a characteristic apple green birefringence with polarized light after staining with Congo red. Three major forms of amyloidosis are primary systemic amyloidosis, secondary systemic amyloidosis, and localized amyloidosis. The mean survival of patients with systemic

forms is between 5 and 15 months, whereas those with the localized form have an excellent prognosis.<sup>1,2</sup>

Involvement of the tongue by amyloid deposits is mostly seen secondary to systemic disease. The aim of presenting this report is to highlight a case of the localized, nodular form of amyloidosis in tongue, which is very rare. Complete systemic workup for amyloid is stressed as this could be a cornerstone in setting treatment protocols and thus can markedly change the morbidity and mortality.

## CASE REPORT

A 56-year-old father of a serving soldier presented to the Department of ENT, INHS Asvini, Mumbai (India), with history of mass lesion in the posterior part of tongue since 2 years duration. The patient had no history of pain and difficulty in deglutition. No history of fever, malaise, and weight loss was found. There was no history of breathing difficulty, chest pain, and bone pains. The tongue was not enlarged. On examination, no cervical lymphadenopathy was found. Complete hemogram, liver function tests, and renal function tests were essentially within normal limits. No history suggestive of heart disease was noted. Resting electrocardiogram, transthyretin levels, serum, and urinary protein electrophoresis were within normal limits. Magnetic resonance imaging revealed nodular area on posterior aspect of tongue without focal signal intensity and no contrast enhancement. Abdominal fat biopsy was done, which appeared normal on histopathology. Bone marrow examination for plasma cell dyscrasias was negative. Wide local excision of the lesion was done, and the tissue was sent to pathology department for histopathological examination (HPE). Gross examination revealed area of depression in posterior part of tongue postoperatively (Fig. 1A). Resected specimen showed 2 × 2 cm whitish mass with yellowish discoloration (Fig. 1B). In addition, HPE on hematoxylin and eosin (H&E) sections of the tissue revealed amorphous eosinophilic deposits in the dermis upper one-third region, suggestive of amyloid (Fig. 2A). Special stain (Congo red) revealed characteristic apple green birefringence in polarized microscopy, confirming amyloid deposits in tongue (Fig. 2B). After exclusion of systemic amyloidosis by detailed laboratory investigations, definitive diagnosis of isolated amyloidosis of tongue was given.

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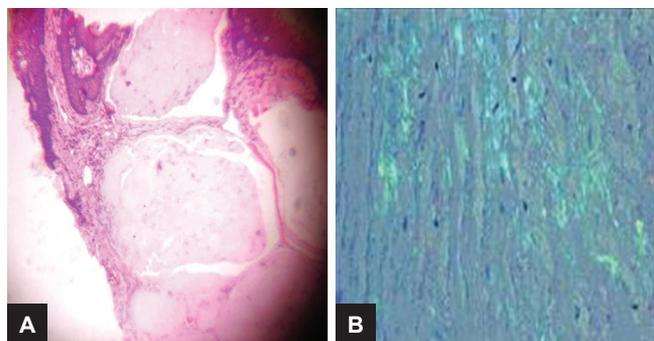
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**Figs 1A and B:** (A) Area of depression seen in posterior part of patient's tongue after mass lesion resection; and (B) Diffuse yellow discolored area noted on gross examination of resected specimen



**Figs 2A and B:** (A) Hematoxylin and eosin (H&E) stain showing eosinophilic, amorphous, hyaline, and acellular material deposition, extracellularly in the dermis – amyloid deposits (H&E, 10×); and (B) Congo red stain showing apple green birefringence of amyloid on polarized light (Congo red, 40×)

## DISCUSSION

In 1854, Rudolph Virchow named it amyloid based on color after staining these proteins with iodine and sulfuric acid. In approximately 12 to 90% of the amyloidosis patients (both systemic and localized form of the disease), head and neck region is known to be affected.<sup>3</sup> In oral cavity, involvement of tongue is the most frequent site, followed by the floor of the mouth, buccal mucosa, and lower lip region.<sup>4</sup> It also affects pharynx, orbital sinuses, larynx, and salivary glands.<sup>5</sup>

It has been documented that most patients with tongue involvement have systemic amyloidosis;<sup>6</sup> however, localized amyloidosis is a rare type of amyloidosis which is not associated with either multiple myeloma or systemic amyloidosis and which does not usually progress to systemic amyloidosis. Cases of localized tongue amyloidosis are extremely rare, with not more than 30 cases being reported in the literature so far.<sup>7</sup>

In the review of 236 cases of amyloidosis, Kyle and Bayrd<sup>2</sup> found only 22 cases (9%) of localized form. Kerner et al<sup>1</sup> observed that localized amyloidosis did not evolve to primary or secondary systemic amyloidosis or multiple myeloma. Our present case also showed similar features as were reported in the above studies.

Macroglossia, difficulty in swallowing, respiratory difficulty, and tongue protrusion beyond the alveolar ridge are the typical presenting features of localized amyloidosis of tongue.<sup>8</sup> This is in contrast to our patient who did not have these presenting features and presented with swelling in back part of tongue, which shows variation and diversity in the modes of presentation of these patients.

Symptoms of systemic amyloidosis vary from constitutional symptoms like fatigue, unexplained weight loss, and loss of appetite to the features mimicking other medical conditions like swelling of the ankles and legs, shortness of breath, unexplained bruising around the eyes, numbness or tingling in the hands and feet.<sup>9</sup> It is of

utmost importance to distinguish systemic amyloidosis from localized amyloidosis, as complete clinicopathological and management profile varies. The former usually has a good prognosis and recurrence is rare following excision.<sup>10</sup> Conversely, most patients diagnosed with systemic amyloidosis die within a year.<sup>3</sup>

The differentiation between these forms of amyloidosis can be done by carrying out extensive laboratory investigations, which include serum and urine electrophoresis to detect Bence-Jones proteins and by doing a bone marrow examination for plasma cell dyscrasias.<sup>11</sup> According to Dr G. Gallo's experience, abdominal fat biopsy gave 94% success rate in cases with presence of systemic amyloidosis.<sup>12</sup> Our patient did not show any of the above features, thus ruling out the possibility of systemic amyloidosis.

## CONCLUSION

Isolated and localized amyloidosis of tongue is very rare and often mimics benign tumors, thus making job of the treating doctor more challenging. The foremost task in management of these patients is to rule out the possibility of systemic amyloidosis because the localized forms of the disease may be treated with wide local surgical excision. The prognosis of the patients with localized amyloidosis is not fully clear, considering the rarity of the disease. So careful follow-up of these patients is advised to monitor its progression. Contribution of new cases is required to study larger sets of these patients to establish the treatment protocol.

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