Localized Amyloidosis of the Tongue: A Review of the Mayo Clinic Experience

Ashley O'Reilly, M.D.[1], Anita D'Souza, M.D.[2], Daniel Price, M.D.[1]
Division of Otorhinolaryngology, Division of Hematology
Mayo Clinic, Rochester, MN

Abstract

Objectives:
-Review Mayo Clinic experience of localized tongue amyloidosis
- Evaluate the work-up of patients with localized amyloidosis, particularly method of excluding systemic amyloidosis
-Review any associated symptoms of patients with localized tongue amyloidosis
-Review treatment and outcomes for patients with localized tongue amyloidosis, particularly if patients went on to develop systemic amyloidosis

Methods: Cases of localized tongue amyloidosis were identified from the tumor registry at the Mayo Clinic in Rochester, Minnesota from 1986 to 2011. Electronic records were reviewed with focus on presenting symptoms, laboratory results (i.e., serum or urine immunoelectrophoresis, bone marrow biopsy and fat aspirate analysis), treatment modality, and status of disease at follow-up.

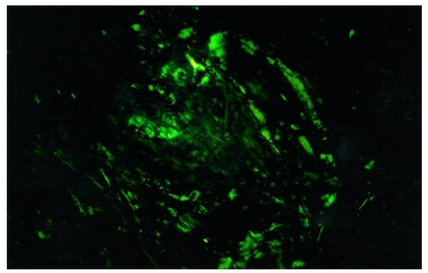
Results: Six cases of localized tongue amyloidosis presented to the Mayo Clinic between 1986 and 2011. Mean patient age was 69 years (range, 43 to 90 years). Patients presented with asymptomatic tongue mass(es). Biopsy of the tongue mass in all patients showed amyloid on Congo red stains. Workup for systemic amyloidosis, including bone marrow biopsy, fat aspiration and serum and urine protein immunoelectrophoresis, was negative for all six patients. Two patients underwent resection of the lesions and the remaining patients elected for observation. Recurrence requiring repeat excision occurred in one of the patients that underwent resection. Repeat evaluation for systemic involvement was performed in three patients 1 to 3 years after the initial diagnosis. Work-up for systemic involvement continued to be negative in these patients

Conclusions:
- Localized tongue amyloidosis remains a rare diagnosis
- Systemic amyloidosis evaluation is necessary in all patients with amyloidosis of the tongue
- Once systemic involvement is ruled out, amyloid lesions may be observed
- Resection of amyloid lesions may result in recurrence
- Patients with localized tongue amyloidosis do not appear to be at increased risk of developing systemic involvement

Background

Amyloidosis is a disease characterized by deposits of proteinaceous material in various organs. These deposits are identified by apple-green birefringence when stained with Congo red and viewed under polarized light (figure 1). There are three forms of amyloidosis: primary systemic amyloidosis, secondary systemic amyloidosis, and localized amyloidosis. Mean survival of patients with the systemic forms is between 5 to 15 months. Patients with localized forms, however, have excellent prognosis and are not at increased risk of developing systemic involvement. Although localized disease in the head and neck is common, particularly in the larynx, involvement of the tongue is almost always associated with the systemic forms of amyloidosis. Amyloidosis localized to the tongue, with no other systemic manifestations, is extremely rare with fewer than 10 cases reported in the literature.

Figure 1



Amyloid demonstrating apple-green birefringence with polarized light with Congo red.

Thompson LDR, Derringer DA, Wenig BM. Amyloidosis of the Larynx: A Clinicopathologic Study of 11 Cases. Modern Pathology (2000); 13: 528-535

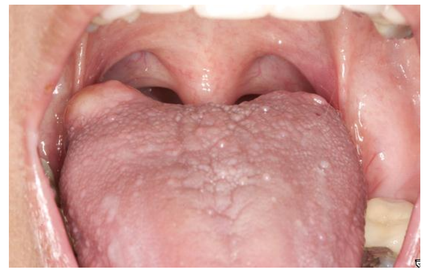
Methods

Six cases of localized tongue amyloidosis were identified from the tumor registry in the department of Hematology at The Mayo Clinic in Rochester, Minnesota from 1986 to 2011. After institutional review board approval, data was gathered using the institutional electronic records. Clinical data included presenting symptoms, laboratory results (i.e., serum or urine immunoelectrophoresis, bone marrow biopsy and fat aspirate analysis), treatment modality, and status of disease at follow-up.

Results

Mean patient age for this six patient case series was 69 years (range, 43 to 90 years). All six patients presented with asymptomatic tongue mass(es) (Figure 2) and no other constitutional symptoms. Biopsy of the tongue mass in all patients showed amyloid on Congo red stains. Workup for systemic amyloidosis was performed on the six patients, including bone marrow biopsy, fat aspiration and serum and urine protein immunoelectrophoresis. These studies were uniformly negative in the six patients. Two patients underwent resection of the lesions and the remaining patients elected for observation. One patient in which resection was performed developed a recurrence in the lesion within one year, requiring repeat excision. Three patients underwent repeat evaluation for systemic involvement 1 to 3 years after the initial diagnosis. Work-up for systemic involvement continued to remain negative in these patients (Table 1).

Figure 2



43-year-old woman presented with a 1-year history of a 1.5-cm right-sided dorsal tongue mass

Table 1

Patient	Age	Presenting symptom	Systemic Work-up[1]	Recommendation	Follow-up[2]
Patient 1	43 yo female	1.5-cm right-sided dorsal tongue mass	Negative	Resection of mass	none
Patient 2	72 yo male	2-cm central dorsal tongue mass	Negative	Observation	none
Patient 3	73 yo female	Central dorsal tongue mass	Negative	Observation	3 years: negative systemic work-up
Patient 4	74 yo male	2 1-cm masses on left base of tongue	Negative	Observation	none
Patient 5	64 yo male	Central dorsal tongue mass	Negative	Observation	2 years: negative systemic work-up
Patient 6	90 yo female	Several lesions on tongue base	Negative	Resection of lesions	1 year: Recurrence of lesions, negative systemic work-up

[1]Systemic work-up at initial diagnosis: bone marrow biopsy, fat aspirate analysis, urine and serum immunoelectrophoresis
[2]Systemic work-up at follow-up: Urine and serum immunoelectrophoresis

Discussion

Clinically, amyloidosis represents a diverse disease process owing to the wide range in organs that can be affected. Systemic versus local involvement further varies the clinical manifestations and prognosis of this disease. Patients with systemic involvement typically have a poor prognosis with a mean survival of 5 to 15 months. Patients with localized forms appear to have excellent prognosis and are not at increased risk of developing systemic involvement.[1]

Head and neck involvement is commonly seen in both the localized and systemic forms of the disease, with the tongue and larynx as the most frequently affected subsites. The tongue is considered the most common head and neck site of involvement in cases of systemic amylodosis.[2,3] In cases of localized amyloidosis, however, the tongue is very rarely involved. The larynx, subglottis and thyroid are the most commonly encountered head and neck sites in localized amyloidosis.[4]

Cases of localized tongue amyloidosis are extremely rare with fewer than 10 cases reported in the literature[1,2]. As with the previously reported cases of localized tongue amyloidosis, the 6 patients in this case series presented with a rubbery to firm mass or masses on the tongue rather than macroglossia. Observation is typically undertaken, particularly if the mass is asymptomatic. In our limited experience, resection of the mass may result in recurrence. Although we had limited follow-up, our case series confirmed that localized forms of amyloidosis, including the tongue, result in an excellent prognosis with no increased risk of developing systemic amyloid involvement.

References

1) KS Fahrner, CC Black and BJ Gosselin. "Localized amyloidosis of the tongue: a review," American Journal of Otolaryngology, vol. 25, no. 3, pp. 186-189, 2004.
2) MM Kerner, MB Wang, G Angier, et al: Amyloidosis of the head and neck – A clinicopathologic study of the UCLA experience, 1955-1991. Arch Otolaryngol Head Neck Surg (1995) 121: 778-782
3) RA Kyle, ED Bayrd: Amyloidosis: Review of 236 cases. Medicine (1975) 54: 271-782
4) M Pentenero, L Davico Bonino, C Tomasini, et al. Localized oral amyloidosis of the palate. Amyloid (2006) 13: 42-46