Two Young Adults with Laryngeal Amyloidosis—A Localized "Super-Low-Grade" B-Cell Lymphoproliferative Disease

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Background and Purpose

Laryngeal amyloidosis is believed to be a self-limiting, localized B-cell plasmacytoma that primarily affects adults in their late 50s. The current approach to treatment is surgical removal of the amyloid plaques and surrounding tissue--however, disease tends to be recurrent. This case report presents 2 patients with a much younger onset compared to recently published medians/averages and demonstrates the need for further study of localized amyloidosis and the pursuit of non-surgical treatment options.

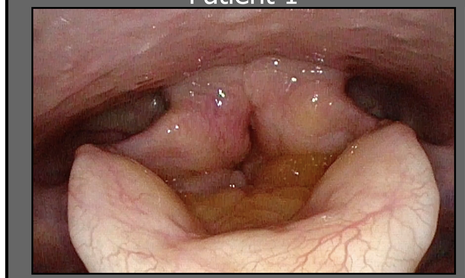
Case 1

Patient 1, a 29-year-old woman who presented to Otolaryngology with intermittent aphonia and sore throat over the course of 2 years. She noted pain when able to speak and denied dysphagia. Laryngeal videostroboscopy was performed with the salient findings being: large subepithelial infiltrative lesions encompassing the entire glottis, supraglottis, and stretching to the petiole of the epiglottis, a large glottic gap, complete vibrational absence of the vocal cords, and identical infiltrate noted in the soft palate. Later biopsy revealed apple green birefringence on Congo red stain and further characterization using liquid chromatography tandem mass spectrometry (LS MS/MS) showed the deposits to be amyloid-lambda (AL)-type amyloidosis. The lesion was partially removed from the right side; however, resection was limited due to concerns of stenosis of the supraglottis. Workup revealed no indications of systemic amyloidosis. She has currently regained about 50% of her ability to speak.

Case 2

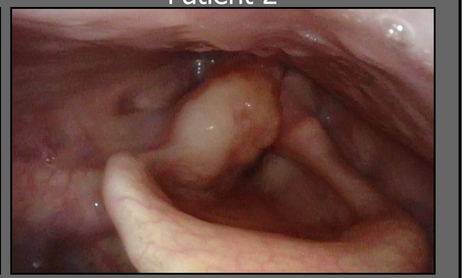
Patient 2 is a 26-year-old woman who was referred to Otolaryngology 2 years ago for laryngoscopy as follow-up when a 1.25cm mass on the right arytenoid process was noted as an incidental finding during upper endoscopy for ongoing gastroesophageal reflux disease (GERD); she presented without dysphonia or dysphagia. Her follow-up laryngoscopy reaffirmed the presence of a >1 cm lesion that was described as cauliflower appearing and it was noted that this lesion did not interfere with vocal cord vibration. Biopsy of the lesion was characterized using LS MS/MS and determined the deposits to be AL-type amyloidosis. Workup for systemic amyloidosis, including positron emission tomography-computed tomography scan (PET-CT) and bone marrow biopsy, were negative. Without symptoms, surgical treatment was not indicated and yearly follow-up was ordered. Initial follow-up revealed no changes.

Patient 1



An image of the larynx taken via laryngoscopic examination of a 29 year-old woman who presented with dysphonia. The orange lesions on her glottis, supraglottis, and epiglottis were later determined to be amyloid-lambda (AL) type amyloid deposits using liquid chromatography tandem mass spectrometry (LS MS/MS).

Patient 2



An image of the larynx taken via laryngoscopic examination of a 26 year-old woman who presented without symptoms. Laryngoscopic examination was performed due to the incidental finding of a cauliflower appearing lesion on the right arytenoid (seen above). The lesion was later determined to be AL-type amyloid deposits LS MS/MS.

Discussion

These patients are outliers within our current understanding of localized amyloidosis due to their age of onset, as this is a disease that typically affects older individuals. Given that laryngeal amyloidosis is typically recurrent, it's likely that these young patients will require future procedures and could develop new/worsening dysphonia. These cases demonstrate the severity of some of the lifelong complications that can arise due to laryngeal amyloidosis. Patient 1 has already experienced a 50% decrease in her ability to speak despite surgical intervention and, although an uncommon symptom of laryngeal amyloidosis, cases like hers are at a high risk of further complications due to the recurrent nature of the disease. Extremely low mortality rates and prognoses that have no effect of average life-expectancy have made laryngeal amyloidosis an often overlooked condition medically; however, it is here where efforts should be made to prevent recurrence and future symptoms. With the current understanding of localized amyloidosis being a self-limiting neoplastic B-cell process, the development of medical therapies similar to those used to prevent progression of indolent B-cell lymphomas should be explored to prevent recurrence and development of worsening symptoms.

References

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