Laryngeal Amyloidosis – Report of 2 Cases with Literature Review

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Background:
Amyloidosis constitutes less than 1% of benign laryngeal tumours and may occasionally be associated with systemic disease. Commonly present with long-standing hoarseness or dyspnea. Diagnosis is established with histological features of acellular, amorphous, eosinophilic deposits which react positively with Congo red.

We present 2 such cases of laryngeal amyloidosis – one localized and other one with systemic involvement. Literature review, diagnostic work-up and treatment options are discussed.

Materials:
Case 1:
63 years old man, non-smoker, presented with hoarse voice for 2 months. Flexible laryngoscopy showed diffuse fullness on left hemilarynx. Microlaryngoscopy revealed submucosal lesion in anterior left vocal cord which on deep biopsy confirmed AL type amyloid. Detailed work-up by haematologist ruled out systemic disease. Patient is under follow-up with no clinical recurrence.

Case 2:
74 years old moderate smoker presented with 6 months of hoarse voice and burning sensation in tongue. He had mucosa-covered growth in right false vocal cord which on biopsy confirmed amyloidosis (AL type). Further work-up revealed systemic disease with elevated serum free light chains, IgG Kappa paraprotein and positive abdominal fat biopsy. He had treatment with CDT chemotherapy and is under haematologist surveillance. Past history includes carpal-tunnel decompression.

Discussion:
1. There have been a variety of classifications of amyloidosis: according to its distribution (localization), clinical type, and the presence or absence of underlying disease and by its precursor protein (immunocytochemical nature) and patterns of extracellular deposition.
2. The three forms of systemic amyloidosis include the primary form, the reactive form, and the familial form. The localized form is rare, with the larynx affected more frequently than any other single site.
3. Although immunophenotyping has become universally available, the "gold standard" for the diagnosis of amyloid remains a tissue biopsy demonstrating characteristic hematoxylin and eosin changes and Congo red birefringence.
4. Systemic amyloidosis is caused by plasma-cell dyscrasia and is treated with chemotherapy and/or radiotherapy.
5. LA may develop from a localized monoclonal immunoproliferative disorder and is best treated with cold steel resection or CO2 laser excision and may require repeated removal in some patients.
6. Investigation for LA are tailored to each patient based on clinical presentation and include search for lymphadenopathy, radiographic imaging (chest x-ray, skeletal survey, urinary and digestive tract evaluation), bone marrow biopsy, and clinical laboratory studies (FBC, ESR, liver chemistries, renal function, quantitative immunoglobulin assay, urine analysis, urine and/or serum electrophoresis, Bence-Jones protein analysis).
7. Furthermore, as indicated clinically, a rectal, lip, gum, kidney, spleen, liver, skin, or small-bowel biopsy and/or abdominal fat aspirate may be performed to exclude systemic disease.

Conclusion:
Endolaryngeal treatment is sufficient with localized disease. Systemic disease needs more extensive treatment and those with associated myeloma have poor prognosis.

Points to remember
1. Laryngeal amyloidosis (LA) is an uncommon disorder and usually a localized disease. However, LA may rarely be associated with multifocal and/or systemic disease.
2. Conservative surgical intervention to preserve laryngeal function and long-term clinical follow-up for recurrent/residual disease is the current recommendation.
3. Although the possibility of systemic disease is low, appropriate clinical, radiographic, and laboratory investigation to rule out systemic disease and to subclassify type of amyloidosis is recommended.

Bibliography

Amyloid deposition in the lamina propria, sparing the overlying epithelium

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

A. perivascular deposition of amyloid in the subepithelial region.
B. a periglandular deposition of amyloid