Abstract

Nasopharyngeal amyloidosis is a rare entity described in the otolaryngology literature. It is usually isolated and does not represent a manifestation of systemic disease. Herein we discuss the first reported case of incidentally discovered nasopharyngeal amyloidosis leading to the diagnosis of systemic disease.

Localized vs Systemic Amyloidosis

Amyloidosis is important to break down into local versus systemic disease. In the head and neck, amyloidosis lesions present mostly in the larynx (60%), followed by the trachea (9%), orbit (4%), and nasopharynx (3%) and are almost always associated with local disease, except in the case of this patient. Treatment is typically excision with cold instruments or laser.

Systemic disease is categorized as primary, consisting of AL protein (immunoglobulin light chain) and falls in the spectrum of multiple myeloma and plasmacytoma, or secondary consisting of AA (serum amyloid A, acute phase reactant), associated with neoplasm. Treatment is with chemotherapy.

Conclusions

Nasopharyngeal amyloidosis has previously been described in the literature in thirteen different patients, all of whom had localized amyloidosis. This is the first case report of nasopharyngeal amyloidosis leading to the diagnosis of systemic amyloidosis, which carries significant different implications in treatment, prognosis, and followup.

Although nasopharyngeal amyloidosis can present with epistaxis, eustachian tube dysfunction, middle ear effusion, and/or a conductive hearing loss, it is usually asymptomatic. Diagnosis is possible with tissue biopsy of obvious lesions. Systemic amyloidosis is diagnosed with abdominal wall biopsy, rectal biopsy, immunoglobulin assay, or bone marrow biopsy.

Amyloidosis is associated with nasopharyngeal carcinoma although the prognostic significance of this is unknown. Nasopharyngeal amyloidosis is rare and typically occurs as an isolated manifestation of localized amyloidosis. However, the otolaryngologist must be aware of the association with nasopharyngeal carcinoma and the possibility of systemic disease.

Clinical Presentation

A 56 year old male presented to the otolaryngology clinic with complaints of otalgia and eustachian tube dysfunction. He was noted to have a unilateral middle ear effusion and subsequent nasopharyngoscopy demonstrated a right nasopharyngeal mass at the eustachian tube orifice—this was biopsied and shown to be nasopharyngeal amyloidosis.

Amyloidosis is a condition which entails the abnormal deposition of proteins in tissue (local) and organs (systemic) throughout the body secondary to an alteration in the secondary structure of the protein into an insoluble form, termed amyloid.

Amyloidosis in the head and neck most often presents in the larynx (60%), followed by the trachea (9%), orbit (4%), and nasopharynx (3%). The vast majority of these lesions represent isolated amyloidosis, which is not associated with a shortened lifespan or the sequelae associated with systemic disease.

Treatment of local disease is typically excision followed by routine surveillance for recurrence. Treatment of systemic disease is complex, and should be managed by a Hematologist/Oncologist.

Amyloidosis can be associated with malignancy, thus proper workup is important even when isolated lesions are discovered.

References


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