Amyloidosis of the Nasopharynx and Nasal Cavity Presenting as Severe Epistaxis and Sphenoid Sinus Dehiscence

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Abstract

Objectives: To review the presentation and management of amyloidosis of the nasal cavity and nasopharynx and to present a case with unusual radiographic findings.

Study design: Retrospective chart review.

Methods: Case report and review of the literature.

Results: Localized amyloidosis involving the head and neck is uncommon. Nasopharyngeal amyloidosis is even rarer, with only a few case reports in the literature. On Computed Tomography (CT) scan, these lesions usually present as a well-defined, homogenous, smooth soft tissue mass without any evidence of bony erosion. We present a case of a 33 year old female transferred to our institution from another hospital with a longstanding history of nasal obstruction and epistaxis. On endoscopic examination, the nasopharynx and nasal cavity were noted to have a nonobstructive smooth, well-circumscribed tumor-like mass along the roof of the nasopharynx, which was also biopsied. There was no clear source of epistaxis found, and bilateral nasal septal splits were placed to prevent further nasal obstruction. The patient was discharged on POD #2 without any significant recurrent epistaxis.

Discussion

Amyloidosis is characterized by the extracellular deposition of an insoluble fibrillar proteinaceous matrix into a highly organized nonimmunoglobulin precursor proteins, with their resultant polymerization into an insoluble amyloid fibril which deposits in the extracellular space. Its symptoms vary depending on whether the pathology is systemic or localized, and on the organ systems involved. While localized amyloidosis is rare, constituting 10–20% of amyloidosis cases, it most commonly affects the head and neck. The larynx is most frequently affected, followed by the oropharynx (23%), trachea (9%), and orbit (4%). Nasopharyngeal and nasal cavity involvement is rare, comprising 3% of cases in one study, and with only 14 % of case reports in the literature. Due to its rarity, amyloidosis is not routinely considered in the differential of nasal cavity and nasopharyngeal masses.

Case Report

The patient was a 33 year old female with a past medical history of iron deficiency anemia and sickle cell trait who presented with a several month history of epistaxis, nasal congestion, and headaches. She was originally scheduled for sinus surgery by an outside Otolaryngologist, however had worsening epistaxis requiring admission and nasal packing at an outside hospital. She was taken to the operating room for a septoplasty and biopsy of a right lateral nasal wall mass, however the surgery was terminated due to excessive bleeding. A posterior nasal pack was placed intraoperatively for hemostasis, and the patient was transferred to our institution for further management. Upon presentation to our institution, the patient had a right sided posterior nasal pack, and a left sided anterior nasal pack with no evidence of persistent epistaxis. Noncontrast CT of the sinuses performed one month prior to admission noted bilateral anterior nasal cavity soft tissue masses, as well as a well circumscribed soft tissue mass along the roof of the nasopharynx. There was associated dehiscence of the sphenoid sinus floor and unilateral sphenoid sinus opacification. (Figures 1,2) Retrospective review of a CT one year prior to presentation for complaints of nasal obstruction revealed the same radiographic finding. The patient’s preoperative labs were normal except for a mild leukocytosis with WBC of 11,600.

Pathology

Amyloidosis is characterized by the extracellular deposition of an insoluble fibrillar proteinaceous matrix into a highly organized nonimmunoglobulin precursor proteins, with their resultant polymerization into an insoluble amyloid fibril which deposits in the extracellular space.

Pathologic examination of the right lateral nasal wall and nasopharyngeal biopsies revealed amyloid fibrillar material in the subepithelial stroma, consistent with amyloid. The presence of amyloid was confirmed by Congo Red stain, and the presence of apple green birefringence with polarized lens. (Figures 3-5)

Figure 1: noncontrast CT sinus demonstrating soft tissue mass along nasopharyngeal roof with sphenoid sinus floor dehiscence and opacification

Figure 2: Case report

References

4) Lim JS et al. Primary amyloidosis presenting as a nasopharyngeal mass. American Journal of Otolaryngology 2010; 31; 130-131
13) Pambuccian S. Nasopharyngeal amyloidoma: CT, MR, and pathologic findings. Ear Nose Throat J 2001; 80: 802-3

Figure 3: CT & E. 400X. Amyrophous eosinophilic material in subepithelial struma, consistent with amyloid. The presence of amyloid was confirmed by Congo Red stain, and the presence of apple green birefringence with polarized lens. (Figures 3-5)