Primary Nasopharyngeal Amyloidosis: Unexpected Cause of Unilateral Hearing Loss

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Abstract

Primary nasopharyngeal amyloidosis (PNA) is an extremely rare condition described in the otorhinolaryngology literature, for which the standard therapy still does not exist. Herein, we discuss an atypically presented PNA case that caused a nasopharyngeal mass and the conductive-type hearing loss. A 54-year-old male patient presented with aural fullness and hearing loss in the right ear lasting for 2 weeks. A physical examination with nasal endoscopy indicated an irregular mass lesion on the right half of the nasopharynx. Otomicroscopy showed otitis media with effusion (Type B tympanogram) in the right ear. Magnetic resonance imaging (MRI) revealed the presence of a contrast-enhanced soft tissue mass arising from the lateral nasopharyngeal wall. A biopsy was performed with nasal endoscopy under local anesthesia, and histopathological evaluation was reported as primary (AL type) amyloidosis. Therefore, the patient was accepted as PNA due to the absence of systemic involvement. One year later, the patient was admitted due to hoarseness. A histopathological examination of the right ventricle was reported as primary amyloidosis. PNA is an exceedingly rare condition, and it typically occurs as an isolated manifestation of localized amyloidosis. However, the clinician must be aware of the association with nasopharyngeal carcinoma and the possibility of plasma cell dyscrasias, especially multiple myeloma and lymphoproliferative diseases. In the absence of systemic disease evidence, localized amyloidosis of the nasopharynx and larynx may be treated conservatively.

Keywords: Primary amyloidosis, nasopharyngeal amyloidosis, amyloid, hearing loss, nasopharyngeal mass

INTRODUCTION

The term amyloidosis includes a group of diseases characterized by the accumulation of extracellular amyloid fibrils that occur mostly without any known cause. Clinically, it is divided in two main groups as systemic and localized amyloidosis. Systemic amyloidosis is divided into three subsections: 1) primary amyloidosis (AL type), which is the most common type, usually associated with multiple myeloma and plasma cell dyscrasias. It is also the most common form of primary localized amyloidosis. 2) Secondary amyloidosis (AA type or reactive) occurs with chronic inflammatory diseases or malignant tumors. 3) Familial amyloidosis is an autosomal recessive transition that makes 2% of all amyloidosis cases (1). Secondary amyloidosis accompanies syndromes such as Muckle–Wells syndrome. It is associated with chronic recurrent urticaria, sensorineural hearing loss, periodic arthritis, and secondary amyloidosis. (2) Localized primary amyloidosis in the head and neck region is rarely seen, and it does not differ significantly in terms of age and sex distribution (3, 4). Also, it is pathologically benign. Clinical symptoms and findings vary according to the intensity of accumulated amyloid fibrils and the anatomical region affected by the disease. Primary amyloidosis, arising in the aerodigestive tract, is most commonly seen in the larynx (61%), oropharynx (23%), trachea (9%), and orbit (4%) (3-5). More rarely, the tongue, nasal cavity, paranasal sinuses, salivary glands, and cervical lymph nodes are involved. Localized primary nasopharyngeal amyloidosis (PNA) is extremely rare, and there are case reports described in the literature (6).

In this paper, we discussed an atypically presented case with PNA, which caused a nasopharyngeal mass mimicking nasopharyngeal carcinoma and conductive-type hearing loss. One year later, a laryngeal lesion was seen on the endoscopic examination.
A 54-year-old male patient was admitted to our clinic with aural fullness and hearing loss in the right ear lasting for 2 weeks. On the physical examination, otitis media with effusion in the right ear and irregular mass lesion on the right half of the nasopharynx were detected (Figure 1. a, b). No palpable lymph nodes were found in the head and neck region. The patient who was suspected to have nasopharyngeal carcinoma was evaluated with magnetic resonance imaging.

**CASE PRESENTATION**

A 54-year-old male patient was admitted to our clinic with aural fullness and hearing loss in the right ear lasting for 2 weeks. On the physical examination, otitis media with effusion in the right ear and irregular mass lesion on the right half of the nasopharynx were detected (Figure 1. a, b). No palpable lymph nodes were found in the head and neck region. The patient who was suspected to have nasopharyngeal carcinoma was evaluated with magnetic resonance imaging.
contrast-enhanced sino-nasal magnetic resonance imaging (MRI). MRI demonstrated a mass in the right half of the nasopharynx, which was hypointense on the T1-weighted series, mildly hyperintense on T2-weighted series (Figure 2. a, b) and in the post-contrast series, peripheral contrast enhancement revealed. A transnasal endoscopic biopsy was performed under local anesthesia for definitive diagnosis. A histopathologic examination revealed depositing of an amorphous substance within the stroma, staining pink-red with Congo red. Due to the negativity of anti-amiloid A antibody, the primary AL type amyloidosis was diagnosed (Figure 3. a, b). The patient was referred to the Internal Medicine Department for investigation in terms of systemic amyloidosis, plasma cell dyscrasias, especially multiple myeloma, and lymphoproliferative diseases. Patient was diagnosed with PNA due to the absence of systemic involvement. Because of vague symptoms and indolent and benign course of disease, we decided to closely follow up the patient. In the first control examination (1 month after diagnosis), tympanic membrane was intact, and no fluid was present in the middle ear. Also, nasal endoscopy revealed no change in nasopharyngeal findings. The first-year control MRI showed no progression at the nasopharyngeal lesion (Figure 4. a, b). One year later, the patient was admitted to our clinic with hoarseness. The polypoid lesion was seen in the right ventricle on the endoscopic examination (Figure 5). The histopathologic examination indicated primary the AL type amyloidosis. The patient gave his consent for the case report to be published.

**DISCUSSION**

Amyloidosis consists of a heterogeneous group of diseases resulting from the eosinophilic deposition of insoluble polymeric proteins at the extra-
Biopsy and histopathological examination taken from the affected tissue or organ is the gold standard method for diagnosis. A characteristic pathological finding of the disease is the accumulation of an amorphous substance in the extracellular area. With a special staining technique, using the dye Congo red, it shows the pink or red color under light microscope and reveals apple-green birefringence under polarized light that is diagnostic for amyloid deposition. Unlike the systemic form, localized amyloidosis has a very good prognosis. Therefore, systemic involvement of disease must be investigated after a definitive diagnosis of amyloidosis. Because of its rarity, there are no widely accepted treatment options or definitive therapy guides for the amyloidosis. The primary aim of the treatment is to control an underlying disease for systemic amyloidosis. In the literature, to the best of our knowledge, there are no studies that show a transformation of localized amyloidosis to systemic form. For this reason, correction of the physiology of affected tissues and organs seems to be a reasonable treatment goal for the localized form of disease. Excision with conventional surgery or using a carbon dioxide laser provides near-complete improvement in the symptoms of PNA. If the lesion is not removed completely, the likelihood of recurrence is high. Simpson et al. reported low recurrence rates with CO₂ laser resection (11). Major bleeding can occur during surgery due to increased vascularity in cases with plasma cell infiltration. Using a laser in cases of localized amyloidosis arising from the head and neck region seems to be a suitable treatment option both to reduce bleeding during surgery and to prevent recurrence.

Although there was a single case with extensive nasopharyngeal amyloidosis reported to be treated with intensity modulated radiotherapy (5), the use of radiotherapy as a treatment option is not recommended due to the risk of malignant degeneration (8). Postoperative regular follow-up with nasal endoscopy and radiological imaging tools is recommended for early recognition of recurrence (9).

CONCLUSION
Primary nasopharyngeal amyloidosis (PNA) is an extremely rare form of benign localized amyloidosis. Nearly all patients present with non-specific symptoms and signs. The definitive diagnosis is made by pathological evaluation of the material obtained with an endoscopic biopsy. Although extremely rare, it should be kept in mind in differential diagnosis of the mass arising from the nasopharynx or the postnasal region.

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REFERENCES


