Primary Localized Amyloidosis in Nasopharynx: A rare case

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INTRODUCTION

Amyloidosis is a syndrome characterized by deposition of insoluble proteinous material in extracellular matrix. It can be systemic or localized [1]. Localized amyloidosis is a rare condition and larynx is the most common organ affected in head and neck region. There has been 25 cases of sinonasal amyloidosis published in literatures so far [2,3].

We present a case of localized nasopharyngeal amyloidosis which presented with unilateral hearing loss.

CASE

53 years old male patient applied to outpatient clinic with right sided tinnitus and hearing loss that had lasted for 6 months. He did not have any additional symptom related to upper airway structures and no history of any systemic illness or drug usage existed. Otoscopic examination and audio logical study detected right sided otitis media with effusion. Tympanometric study concluded type B tympanograph, and pure tone audiometry confirmed 35 decibel air-bone gap. Nasopharyngeal examination via endoscopy revealed a mass lesion filled Rosenmuller fossa fully and biopsy was taken. As the age of the patient was not young angiofibroma was excluded from differential diagnosis. Nasopharyngeal carcinoma, meta static disease, and lymphoid hyperplasia due to allergic rhinitis or local inflammation were the diagnosis assumed. Histopathological evaluation diagnosed amyloidosis. It was AA; subepithelial, extracellular, a cellular, amorphous, eosinophilic material which was dispersed randomly through out the lamina propria, sparing the over lying epithelium, and frequently demonstrated a perivascular and periglandular deposition, sometimes completely obliterating the seromucous glands by compression atrophy. A sparse in flammatory in filtrate noted in all cases was composed of lymphocytes and plasmacells with occasional histiocytes and a few giant cells, either at the peripheral margin of or enclosed with in the amyloid. In order to assess whether it was a part of systemic disease, internal medicine and hematology consultations were performed. Consultations revealed that there was no renal disease. It was assessed by urea creatin in levels and creatin clearance test. Multiple myeloma was excluded by protein electrophoresis. After it was confirmed that there was no suspicion of systemic amyloidosis, patient was treated by endoscopicalescision of the lesion and insertion of ventilation tube.

DISCUSSION

Amyloidosis is deposition of fibrillary proteinous material in extracellular matrix in different organs and it has 2 sub groups named AL and AA chemically [4,5]. AL is seen in primary systemic amyloidosis and AA is in secondary systemic amyloidosis [6]. Amyloidosis has systemic and localized forms clinically. Sytemic form has 3 sub groups named primary, secondary, and familial [7]. AA is the most common type seen in inflammatory and neoplasic diseases [8].

Amyloidosis is seen generally seen between 4th and 8th decades and more common in males [9]. Larynx is the most common site affected in head and neck region and followed by salivary glands, orbita, and paranasal sinuses [9,10]. In contrast to systemic forms, gender and age don't differ in localized form [11].

Nasopharyngeal amyloidosis is a slowly progressing condition which presents by epistaxis, nasal obstruction, and hearing loss due to otitis media with effusion. As these are non specific symptoms, endoscopic examination and radiologic evaluation are crucial. Certain diagnosis is made by histopathological examination [9,12]. Typical green biref ringence is detected under a polarized light microscope followings taining by congored [13].

Surgery is the definitive treatment for localized nasopharyngeal amyloidosis [4]. Open surgery is difficult to perform because of the location of nasopharynx. Therefore endoscopicales approach

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Keywords
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• Surgical excision
• Otitis media with effusion
has replaced tranpalatal surgery. Bleeding is the most common and important complication of the surgery and endoscopic approach causes less bleeding [14]. We preferred endoscopic approach in our case (Figure 1).

Effects of surgery on prognosis is not known exactly and it is not certain if localized form evolved to systemic form. Regarding the high recurrence rate and risk of development of systemic disease, close followup should be performed [4,15]. This was an important case because localized amyloidosis is very rarely seen in nasopharynx and surgical treatment enabled definitive treatment in our case.

REFERENCES


