CASE REPORT

Amyloidosis of the Nasopharynx: An Unexpected Cause of Unilateral Middle Ear Effusion

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ABSTRACT

Amyloidosis is an idiopathic disease that is characterized by the extracellular deposition of fibrillar proteins. The disease can be categorized as primary or secondary where deposits occur in conjunction with chronic diseases such as rheumatoid arthritis or tuberculosis. The deposits can be localized or systemically distributed. It can mimic, and also be associated with underlying malignancy. Primary amyloidosis is a rare cause of a nasopharyngeal lesion, and less so of a secondary middle ear effusion. Its association with underlying chronic and malignant disease must not be over-looked if serious complications are to be avoided. It is, therefore, important to consider this as a differential diagnosis in such patients.

Key words: *Amyloidosis. Nasopharynx. Middle ear effusion.*

INTRODUCTION

Amyloidosis is an idiopathic disease that is characterized by the extracellular deposition of an abnormal proteinaceous substance in many tissues and organs of the body.[1] The disease can be categorized as primary, where spontaneous deposits of amyloid occur, or secondary, where deposits occur in conjunction with chronic diseases such as rheumatoid arthritis or tuberculosis. The deposits can be localized or systemically distributed. It can mimic, and also be associated, with underlying malignancy. It rarely presents to the otolaryngologist but if the diagnosis is missed then there can be potentially serious complications.

We present a rare case of amyloidosis of the nasopharynx, resulting in a unilateral middle ear effusion. This highlighted some important otolaryngologic implications of the disease.

CASE REPORT

A 45 years old healthy Caucasian woman with no relevant past medical history presented to the otolaryngology clinic with a 5 years history of worsening, but not disabling, left sided deafness. Examination of the ears demonstrated a small left sided middle ear effusion with mild conductive hearing loss on audiometry. A routine fibreoptic examination of the nasopharynx demonstrated a yellow-colored, flat plaque-like thickening of the posterior surface of the upper soft palate, close to the left eustachian tube orifice.

Concern for malignancy promted further investigation. Endoscopic trans-nasal examination was performed under anaesthesia, at which time, biopsy of the lesion was also carried out. Subsequent histopathological analysis demonstrated replacement of normal minor salivary gland tissue with eosinophilic acellular nodules which demonstrated green birefringence with polarized light on Congo Red staining - diagnostic of amyloidosis (Figure 1).

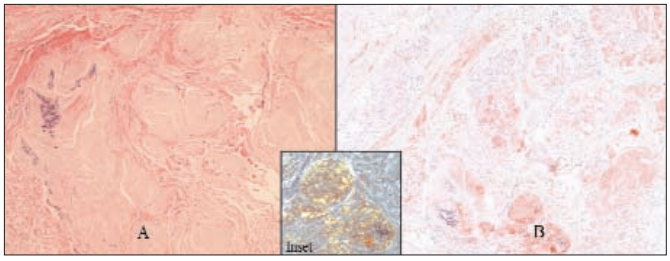

Figure 1: A: H&E appearances of small nodules of amyloid deposited in palatal mucosa. **B:** positive staining of amyloid with congo red, showing amyloid nodules surrounding and adjacent to benign seromucinous glands. **Inset:** apple green birefringence of congo red positive amyloid.

The soft palate swelling was endoscopically dubulked using the microdebrider and the patient made a good recovery. The soft palate and the post-nasal space appeared normal at 3 weeks postoperative follow-up. The left middle ear effusion resolved spontaneously.

Based on the unexpected histopathological diagnosis, the patient was reviewed by the Rheumatologist with special interest in amyloidosis. The autoimmune screen, immunoglobulin levels, and routine haematology and biochemistry investigations were all normal. No evidence of systemic amyloidosis was found clinically.

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At 3 years follow-up, the patient remained well without any symptoms to warrant further investigations. After multidisciplinary team discussion, the patient was believed to have a localized deposit of amyloid in the post-nasal space with unknown etiology, and no systemic involvement. As the patient never developed any systemic symptoms, she was discharged from follow-up with an advice to seek medical help, if needed in future.

DISCUSSION

Amyloid is a pathological pertinacious substance, which may be deposited in various tissues and organs throughout the body in a variety of clinical settings. Amyloidosis is not considered as a single disease entity but rather a group of diseases that have in common the deposition of amyloid proteins. When stained with Congo red and visualized under a polarizing microscope, these proteins exhibit a characteristic green birefringence.[1] The accumulation of these proteins produces pressure atrophy of surrounding cells.[1] Amyloidosis can present as part of a systemic disease, a familial disorder or as a localized finding. It was previously classified according to the clinical setting in which it presented. Advances in immunohistochemistry means that amyloid can now be subdivided into around 15 distinct forms based on the chemical nature of the fibril protein.[1] Light chain amyloid (AL) is seen in primary systemic amyloidosis, myeloma-associated amyloid and is the most common subtype found in localized amyloid deposition.[2] Amyloid A (AA) is implicated in systemic amyloidosis secondary to chronic inflammation or infection.[2] A variety of rare familial amyloidoses have also been described.[1] Amyloidosis localized to the upper aerodigestive tract is rare.[2] When this does occur, the location is most commonly larynx.[2]

Primary amyloidosis of the nasopharynx is very rare, with only a few documented cases.[3-5] It is, therefore, often not considered in the differential diagnosis. In this patient, it manifested as a unilateral middle ear effusion. Assessment of adults with middle ear effusion includes an examination of the nasopharynx to look for pathology obstructing the eustachian tube orifice. This is how the amyloidosis plaque on the upper surface of the soft palate was detected. The significance of this finding is that biopsy must be carried out to rule out localised malignancy, which this condition can mimic.[5] The patient also requires further investigation to rule out amyloidosis secondary to other systemic diseases, in particular rheumatoid arthritis, tuberculosis and myeloma. A diagnosis of any of these clearly has significant implications for the patient. In this patient, the investigations for causes of secondary amyloidosis were performed by rheumatology colleagues. However, this patient remained well clinically without evidence of systemic amyloidosis. In cases of primary amyloidosis, some departments also advocate radio-labeled amyloid scanning[6] to search for other deposits. Since it involves exposing the patient to radiation, the general opinion is to conduct this investigation only if there is a strong suspicion of further deposits. Biopsy of the suspected area is important for diagnosis.[7]

Once other systemic diseases have been ruled out or treated, the management of localized amyloid is dependent on whether it is causing any clinical problems due to pressure effects.[8] In this case, the patient developed a middle ear effusion. Following biopsy, the patient was followed-up and the effusion resolved spontaneously. However, had it persisted then grommet insertion would have been offered to the patient. Deposits elsewhere in the nasopharynx can potentially cause breathing and swallowing difficulties if the deposit is large enough to cause obstruction, or interfere with the function of the soft palate. In these cases, excision of the deposit may be warranted.

Amyloidosis is a rare cause of a nasopharyngeal lesion, and less so a secondary middle ear effusion. Its association with underlying chronic and malignant disease must not be over-looked if significant complications are to be avoided. It is, therefore, important to consider amyloidosis as a differential diagnosis in patients with a middle ear effusion and be vigilant to potentially significant systemic disease.

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