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CASE REPORT

Primary localized amyloidosis in nasopharynx: A Case Report

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ABSTRACT

Nasopharyngeal amyloidosis is an extremely rare benign tumour. It is divided into localized or systemic amyloidosis. It is more common in men. Clinical presentation includes nasal blockage, epistaxis and reduced hearing. Classical positive Congo red stain and appearance of apple green birefringence on polarized microscopy confirms the diagnosis of amyloidosis. We present a case of nasopharyngeal amyloidosis in a 44-year old lady who presented with acute hearing loss for 1 week with epistaxis mimicking nasopharyngeal carcinoma. Clinical examination showed a nasopharyngeal mass with biopsy proven AA amyloidosis. She is now cured of amyloidosis following endoscopic transnasal excision of tumour. We discuss on the similarity of presentation between nasopharyngeal carcinoma; the commonest malignant tumour in our region and the much rarer nasopharyngeal amyloidosis as well as highlighting the importance in early recognition of the latter in view of its known risk of systemic involvement.

Keywords: Amyloidosis, Nasopharynx, Localized, Nasopharyngeal carcinoma, Congo red stain

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INTRODUCTION

Amyloidosis rarely affects head and neck region. It comprises of 2 types; which are are the localized amyloidosis or it may represent a constituent of systemic amyloidosis. Commonest site of involvement in the head and neck region are in the larynx (60%) (1). Primary nasopharyngeal amyloidosis is very rare. It is a slow growing benign nasopharyngeal tumour but can become locally aggressive. There are 32 published cases of localized nasopharyngeal amyloidosis described in literature (2). Presenting complaints in nasopharyngeal amyloidosis include epistaxis and reduced hearing; which is also the common presentation in nasopharyngeal malignancy. Long term clinical outcome of isolated nasopharyngeal amyloidosis is not well documented in view of its rarity.

CASE REPORT

A 44-year-old Malay lady presented with reduced hearing on both sides for one-week. She had minimal self limiting epistaxis. She has had nasal symptoms for many years; treated as allergic rhinitis. There was no recent worsening of rhinitis. There was no facial pain, headache, blurring of vision or neck swelling. She worked as a teacher; neither smoked nor consumed alcohol.

On examination, she was comfortable. There were no neck nodes. Otoscopy showed retracted tympanic membrane on both sides. Nasoendoscopy revealed a mass in nasopharynx; involving the left more than the right (Fig. 1). Throat examination was unremarkable.

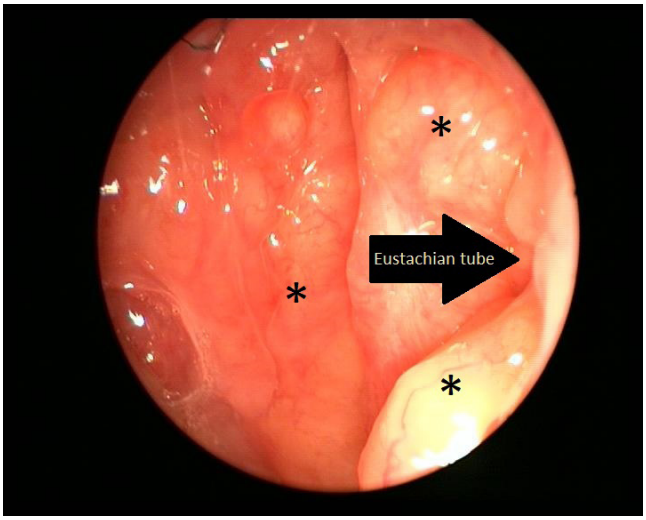

Figure 1: Endoscopic view of mass in nasopharynx seen from the left side. (Arrow = Eustachian tube, * = mass in the nasopharynx)

Biopsy of the nasopharyngeal mass confirmed the diagnosis of AA amyloidosis by immunohistochemistry. The histopathological examination showed fragments of amorphous material covered partly by respiratory epithelium (Fig. 2). There was scattered stromal cells admixed with some inflammatory infiltrates amidst the amorphous tissue. Congo red staining was positive (Fig. 3) and exhibited apple green birefringence on polarization (Fig. 4).

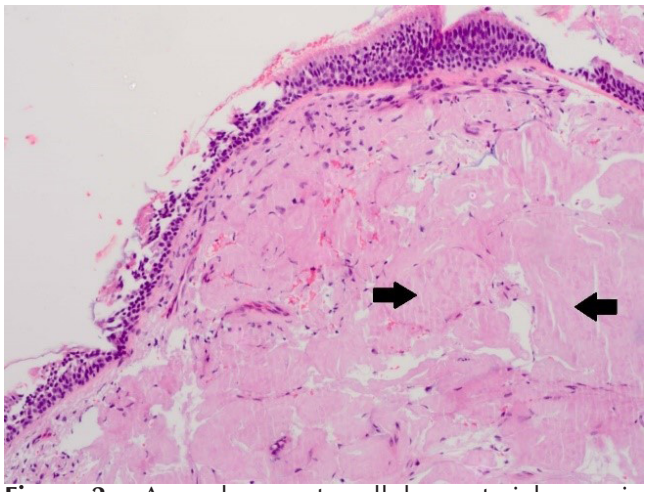

Figure 2: Amorphous extracellular material seen in subepithelium (arrow) seen on Hematoxylin and Eosin (H&E) stain

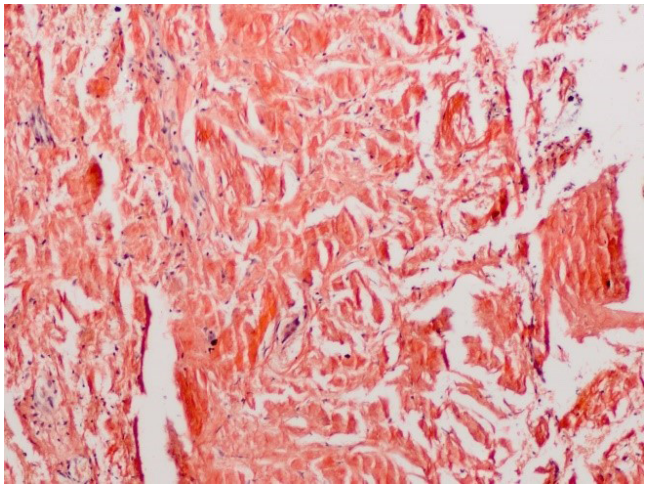

Figure 3: Amyloid stains red in Congo stain

Computed Tomography (CT) scan of Neck showed partial obliteration and thickening of the fossae of Rosenmuller bilaterally with asymmetrical appearance, worse on the left side. There was no bony erosion of the adjacent skull base (Fig. 5).

Her full blood count as well as renal, liver, coagulation and urine profiles were normal. Chest X-ray was normal. She underwent endoscopic excision of the nasopharyngeal mass under general anaesthesia. Intraoperatively, there was polypoidal mucosa at the postnasal space extending laterally to the Eustachian

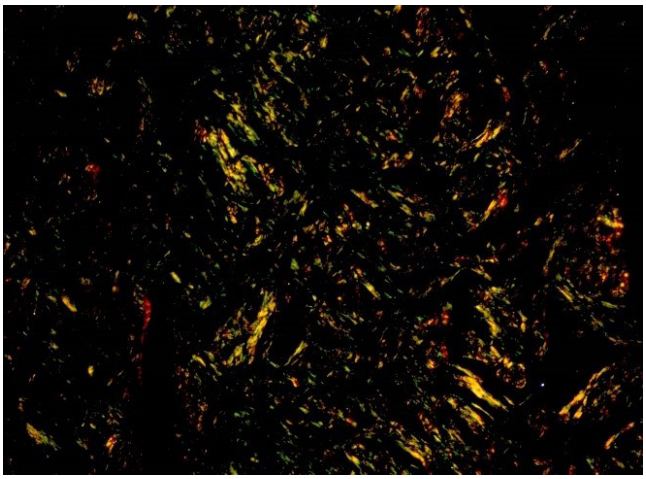

Figure 4: Produce apple green birefringence on polarized light microscopy

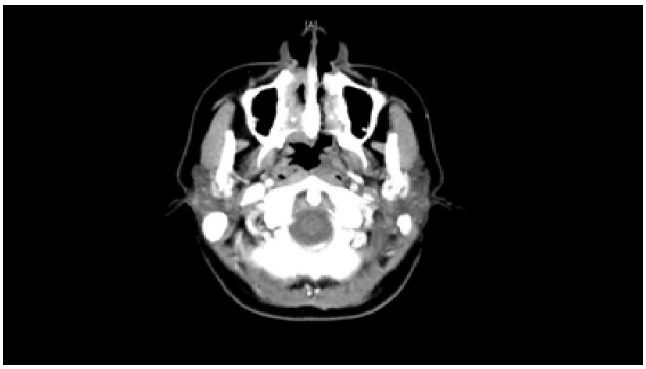

Figure 5: Axial CT showing asymmetry of the fossa of Rossenmuller

tube opening on both sides.; anteriorly to the posterior end of the inferior and middle turbinates bilaterally. The tumoural mucosa was friable with presence of contact-bleeding. Microdebrider was used to excised the tumour until healthy normal mucosa was seen. Haemostasis was achieved using diathemy method. Closure was not done as to allow adjacent mucosa to reepithelialize. Tympanostomy tubes were inserted on both sides during the surgery.

Postoperatively, there was no significant complication except intranasal synechiae.

At 4 years' of surveillance follow up, there was no recurrence of disease. No revision of surgery or medical therapy given in view of negative surveillance nasoendoscopy for residual or recurrence of amyloidosis.

DISCUSSION

Amyloidosis is a condition in which there is deposition of protein fibrils in extracellular space of various tissues. It has unique histochemical findings which demonstrates an apple green birefringence fibrils on Congo red stained upon viewing under polarizing microscope. Amyloid deposition was first described by Rokitansky in

1842. However, the pathogenesis remains uncertain. It predominantly affects men (male to female ratio = 3:1) and affected adults are typically aged between 50 to 70 years old (3).

There are 3 types of amyloidosis; comprising of immunoglobulin light chain derivative (AL) amyloidosis, amyloid associated (AA) amyloidosis and familial amyloidosis. AL amyloidosis is associated with primary systemic amyloidosis and multiple myeloma. AA amyloidosis which is also known as secondary amyloidosis is the most frequent form. AL amyloidosis can be considered as the worst type as it can affect vital organs such as the kidney, heart, liver and gastrointestinal tract leading various symptoms ranging from nephrotic syndrome, heart failure, cholestatic hepatitis and occult bleeding, respectively (4). The presence of amyloid deposits destroy the normal architecture of the tissue or organ; therefore leading to the impairment of normal physiological mechanism.

The spectrum of clinical presentation of amyloidosis depends on which tissues or organs that are involved by the amyloid deposition. Head and neck amyloidosis is rare. As far as we know, there are 32 published cases of localized nasopharyngeal amyloidosis described in literature (2).

Nasopharyngeal amyloidosis presentation can mimic nasopharyngeal carcinoma (NPC) as both diseases can present with epistaxis, nasal obstruction and otitis media with effusion. The clinical presentation in our patient favours NPC in view of its higher incidence although it is a relatively a rare disease worldwide. NPC is the fifth most common cancer in Malaysia based on Malaysian National Cancer Registry Report 2007-2011. In view of the similarity in their clinical presentation, it is crucial to exclude rule out NPC as this malignant tumour has a high incidence in our local population.

Nasopharyngeal amyloidosis has been reported to be associated with nasopharyngeal carcinoma. A large study by Prathap K et al (1984) has reported amyloid deposits in 12% of 434 primary nasopharyngeal carcinoma, more commonly seen in non-keratinizing type (5). Therefore, surveillance nasoendoscopy is required to identify possible malignant occurrence and recurrence of disease.

CONCLUSION

It is of utmost importance to rule out nasopharyngeal carcinoma (NPC) in a patient presented with acute epistaxis and otitis media with effusion in view of its high incidence in our local population. Progression of NPC can lead to intracranial invasion. Nasopharyngeal amyloidosis on the other hand is slow growing, benign and rare tumour. It must be considered in differential diagnosis of any nasopharyngeal mass. Nasopharyngeal amyloidosis can be a local manifestation of a major systemic disorder that may also affecting the heart and kidney; as delay in management may leads to life-threatening complications and death.

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