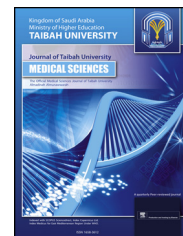




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Case Report

Isolated primary amyloidosis of the epiglottis presenting as a long-standing foreign body sensation in throat: A case report



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المخلص

داء النشواني الأولي للحنجرة هو مرض حميد نادر غير معروف السبب. وتعرض لسان المزمار فقط بداء النشواني هو من أمراض الحنجرة الحميدة غير المألوفة التي تظهر كإحساس بجسم غريب في الحلق. ويجب وضع هذا المرض في الاعتبار في حالة الإحساس بجسم غريب بالحلق لمدة طويلة. الملاحظة والاستئصال بالمنظار باستخدام ليزر ثاني أكسيد الكربون هو طريقة العلاج الرئيسية. هناك حاجة أيضا للمتابعة على المدى الطويل بسبب طبيعة المرض البطيئة والمتفرقة.

الكلمات المفتاحية: النشواني؛ لسان المزمار؛ الإحساس بجسم غريب بالحلق

Abstract

Primary amyloidosis of the larynx is a rare benign disease of unknown aetiology. Isolated epiglottic involvement with amyloidosis is an extremely uncommon benign laryngeal pathology and can present with a foreign body sensation in the throat. This lesion should be kept in mind in cases with a long-standing foreign body sensation in the throat. Observation and endoscopic carbon dioxide laser excision are the main methods of treatment. Long-term follow-up is also required because of the slow progressive nature of this disease.

Keywords: Amyloidosis; Epiglottis; Foreign body sensation in throat

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Introduction

Isolated laryngeal amyloidosis is an extremely rare lesion representing 1% of all benign laryngeal tumours.¹ Amyloidosis was first described by Rokitansky in 1842. The term amyloid was coined by Virchow, who found that it stained violet with iodine and sulphuric acid; he attributed this to its cellulose or starch-like nature.² Laryngeal amyloidosis was first documented in 1875 during a post-mortem examination.³ It is characterized by the deposition of homogenous eosinophilic extra-cellular materials in the laryngeal tissue; this material is protein in nature. Laryngoscopic findings are not very characteristic, and the diagnosis is based on histopathology. Staged microlaryngeal excision using a carbon dioxide (CO₂) laser is the treatment of choice. Because of its rarity and clinical interest, here, we report a case of localized amyloidosis involving the epiglottis in a 45-year-old female patient who presented with an unusual symptom described as a long-standing foreign sensation in the throat.

Case report

A 45-year-old female came to the outpatient department of otorhinolaryngology and presented with a foreign body sensation in the throat that had persisted for three years.

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Indirect laryngoscopic examination of the larynx showed a smooth swelling over the lingual surface of the epiglottis. The laryngostroboscopic video examination confirmed the same findings (Figure 1). The false vocal cords, arytenoids, ventricles, true vocal cords and subglottis appeared normal. The mobility of the vocal cords was normal. She had no history of shortness of breath or stridor. The general examination showed a good general condition, with no breathlessness or dysphagia. Her vital signs were normal, and she had no palpable neck nodes. The ear, nose, pharynx and oral cavity examinations were normal. Complete blood counts (CBC), thyroid functions test (TFT), liver functions test (LFT), C-reactive proteins, ECG and chest X-ray were normal. A computed tomography (CT) scan of the neck with omnipaque contrast revealed a localized enhanced soft tissue mass measuring 1.5 cm × 1 cm in diameter over the lingual surface of the epiglottis (Figure 2). Microlaryngoscopy was performed under general anaesthesia, and the lesion was firm in consistency. The tissue from the mass lesion was sent for histopathological examination. In histopathological examination, the excised polypoidal mass revealed amorphous deposits of eosinophilic, proteinaceous material within the subepithelial stroma (Figure 3a), and this was confirmed as amyloidosis after positive staining with Congo red stain (Figure 3b). The full blood count and erythrocyte sedimentation rates were normal. With a confirmed diagnosis of the mass over the epiglottis, a carbon dioxide-assisted microlaryngeal surgery was performed under general anaesthesia. The epiglottic amyloidosis was resected with a CO₂ laser. The patient is now under regular follow-up care every six months and performs video laryngostroboscopy to rule out any evidence of local recurrence in the larynx. To date, she has had no complaints of a foreign body sensation in her throat.

Discussion

Amyloidosis is characterized by the extracellular deposition of proteinaceous material in the tissue. The incidence of amyloidosis is estimated to be 5 to 9 cases per million per year throughout the world. It is more frequently observed in developed countries.⁴ The amyloid is stained with iodine, metachromatic dyes, methylviolet and Congo red. It has a 3:1 male to female predominance and is common between the 5th and 7th decades of life.⁵ Amyloidosis can be genetic or acquired.⁶ Out of 15 biochemical forms of amyloidosis, the common three types are: i. AL (light chain): derived from plasma cells and contains kappa and lambda immunoglobulin light chains; ii. AA (amyloidosis associated): amyloidosis is a nonimmunoglobulin protein synthesized by the liver. It is a reactive systemic amyloidosis. iii. AB: amyloidosis found in Alzheimer's disease and sometimes familial cases.

Clinically, amyloidosis can be classified into two types: primary amyloidosis, in which there is spontaneous development of an amyloid deposit, and secondary amyloidosis, which is found in conjunction with some other systemic diseases, such as Rheumatoid arthritis or tuberculosis.⁷ Primary amyloidosis can be further divided into localized and generalized forms. The larynx is rarely

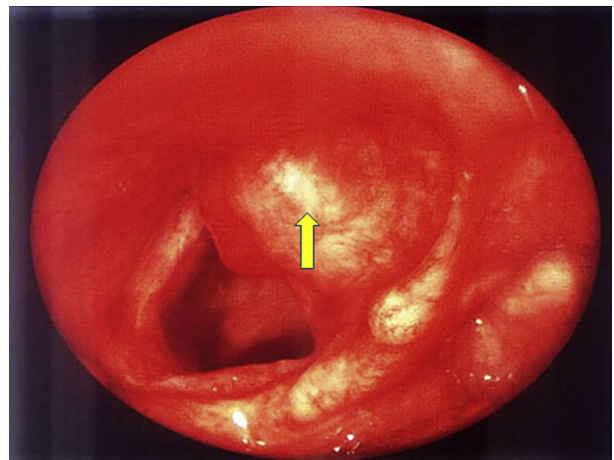


Figure 1: Video laryngostroboscopic picture showing the isolated mass over the laryngeal surface of the epiglottis (marked by the arrow).

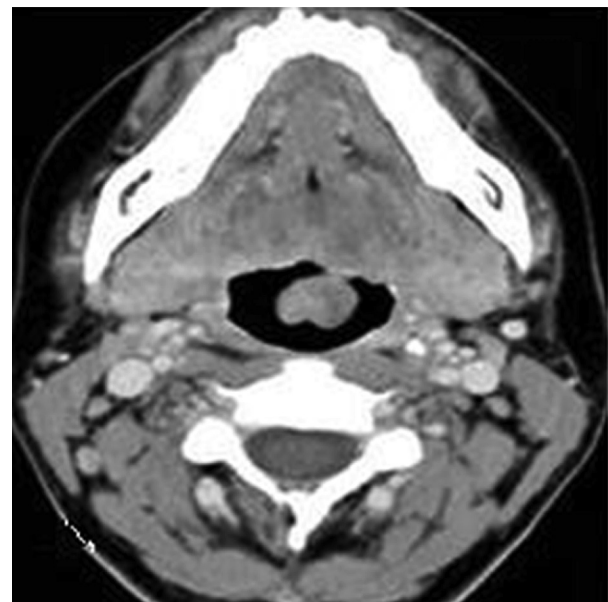


Figure 2: Axial cuts of the CT scan of the neck showing a mass in the epiglottis.

affected with amyloidosis. The most common part of the larynx that is affected is the vestibule, followed by the false cords, the aryepiglottic folds and the subglottic region.⁸ The false vocal cords and the ventricles of the larynx are most commonly affected, and they show smooth, submucosal nodular to diffuse deposits under an intact surface epithelium. In the larynx, the amyloid can be observed as subepithelial extracellular deposits of acellular, homogenous and amorphous eosinophilic materials displaying apple-green birefringence with polarized light when stained with Congo red or metachromatic with crystal violet or methyl violet.⁹ Our case showed primary involvement of the epiglottis without affecting the glottis or laryngeal airway.

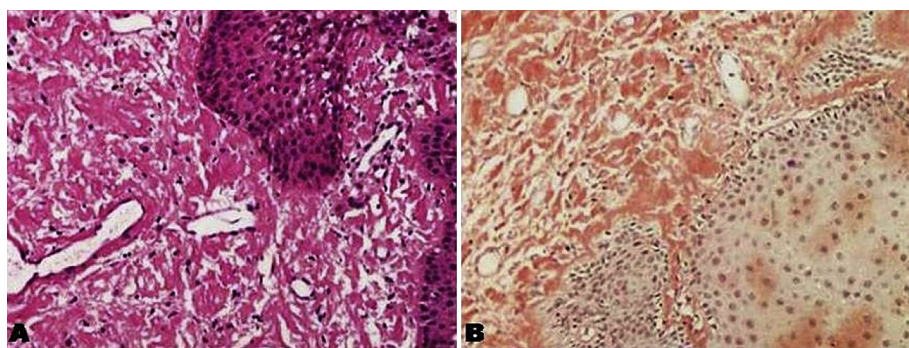


Figure 3: a: Amyloid lesion of the epiglottis showing glossy eosinophilic H and E staining at 10× magnification. b: Congo red stain of the tissue from an epiglottic mass showing the typical salmon pink staining of the amyloid at 10× magnification.

The most common head and neck clinical manifestations of amyloidosis are hoarseness, nasal congestion, odynophagia, articulation problems, mandibular deformities, deglutition difficulties, airway obstruction, speech disorders and hypogeusia.¹⁰ Herein, the patient presented with a long-standing foreign body sensation in the throat, which is rarely reported in head and neck amyloidosis.

In the CT scan, the amyloidosis appeared as a marked thickening of the laryngeal soft tissue with a high density appearance, whereas in MRI, the amyloidosis had the same signal density as skeletal muscle. This may help to differentiate amyloidosis of the larynx because tumours do not appear in this manner in MRI.¹¹

Based upon the involvement of organs of the body and histological evidence of amyloid deposits, a diagnosis is made. The outcome of the investigations differs from patient to patient. Because no blood test is diagnostic for amyloid, a final diagnosis is usually made on the basis of tissue biopsy. Biopsies can be taken from almost any organ based upon the clinical symptoms. In our case, a biopsy from the epiglottic mass was carried out after endoscopic confirmation following the patient's symptoms. Immunohistochemical stains and Congo red staining observed under polarized light microscopy or the electron microscopic findings of a laryngeal biopsy specimen can confirm the diagnosis of the amyloid.¹² Lewis et al. recommended urine and serum electrophoresis as a basic approach for diagnosis of patients suffering from amyloidosis that is observed exclusively in the larynx.¹³ Amyloid protein is detected histologically by staining the samples with Congo red stain, which imparts an apple-green colour to the amyloid protein. In polarized light, the amyloid appears birefringent.

Treatment varies from simple observation of the lesion, if the lesion is asymptomatic, to surgical excision, when the lesion is symptomatic. Presently, the most effective treatment of this lesion is CO₂ laser-assisted surgical excision. Extensive amyloidosis that compromises the laryngeal airway may be treated either with an endoscopic approach or an external approach. Radiotherapy and medical treatment with steroids are ineffective. A new therapeutic modality for amyloidosis involving an experimental small molecule drug called R-1-[6-[R-2-Carboxy-Pyrrolidin-1-Y]-6-Oxo-hexanoyl]Pyrrolidine-2-Carboxylic acid (CPHPC) that depletes the serum amyloid P component (SAP) has been developed by Pepys et al.¹⁴ SAP binds to amyloid fibrils and makes them resistant to degradation. CPHPC interferes with

SAP binding to amyloid fibrils and leads to the rapid clearance of SAP by the liver. Localized amyloidosis has an excellent prognosis in comparison to the systemic variety. Long-term follow up is required because of the slow progressive nature of the disease.¹⁵

Conclusion

Laryngeal amyloidosis is a rare and benign clinical condition. Isolated primary epiglottic amyloidosis is an extremely rare condition. It may present with a foreign body sensation in the throat. Thus, this lesion should be kept in mind in cases with long-standing foreign body sensation in the throat. An endoscopic CO₂ laser is an excellent treatment option for this type of localized laryngeal lesion. There must be regular follow ups to rule out local recurrence.

Conflict of interest

Authors declare that, they have no conflict of interests.

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