

Case Report

A case report on localised laryngeal amyloidosis

Marija Agius^{1,2*}, Tara Grima³, Maja Cvorovic¹, Darko Babic⁴, Charles Borg¹

¹Department of Otorhinolaryngology, Head and Neck Surgery, ³Department of Surgery, ⁴Department of Pathology, Mater Dei Hospital, Malta

²Faculty of Medicine and Surgery, University of Malta, Malta

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*Correspondence:

Dr. Marija Agius

E-mail: marija.agius@gov.mt

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ABSTRACT

Amyloidosis is a rare disease which involves deposition of amyloid in the tissues. Very rarely, amyloid can be deposited in a single organ giving rise to localised amyloidosis. In the present study, a rare case of localised amyloid deposition in the larynx in a young patient was presented. It is a slow progressive disease and often diagnosed late and is confirmed by biopsy results.

Keywords: Amyloid, Deposition, Larynx, Dysphonia

INTRODUCTION

Amyloidosis is a rare disease and is characterized by the deposition of amyloid protein in various target organs of the body.¹ Very rarely, amyloid can be deposited in a single organ giving rise to localised amyloidosis. We present a rare case of a young patient with localised amyloid deposition in the larynx.

CASE REPORT

MN is a 38 year old lady, with a history of smoking, who presented with an 8 month history of dysphonia associated with dysphagia, the latter of which improved.

The patient denied any weight loss or anorexia. Initial examination findings showed a fixed left hemilarynx on indirect laryngoscopy which was confirmed by a flexible nasendoscopy performed at the outpatient department. Subsequently the patient was scheduled for a computed tomography scan inclusive of the neck and trunk, which showed thickening of the left side of the larynx characterized by loss of the fat plane between the

left side of the larynx and the thyroid cartilage anteriorly. No loco-regional lymphadenopathy was noted and the chest and abdomen were reported as normal. Figure 1 below shows the thickened left hemilarynx on imaging.

The patient was admitted for an urgent direct laryngoscopy and biopsy under general anaesthesia. Intra operatively a well circumscribed lesion was seen on the left false cord. The true vocal cord did not seem to be involved. Multiple biopsies were taken and sent for histopathological analysis. The histology result showed mild chronic inflammation and the presence of abundant amyloid in stroma, which was demonstrated by the staining of amorphous material with Congo red that displayed apple-green birefringence when viewed under high- intensity cross-polarised light. Amyloid deposits were present throughout the provided samples. Immunohistochemistry showed that the amyloid stained with antibodies to lambda light chains thereby confirming the Amyloid of AL type (lambda subtype). Refer to Figures 2-4 below which show the histological appearance of amyloid.

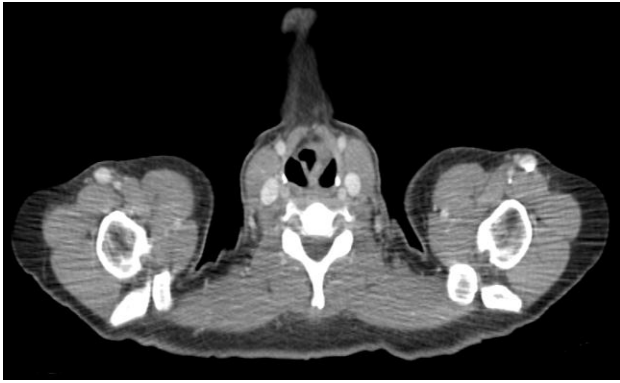


Figure 1: Computed tomography axial view at the level of the glottis which shows thickening of the left hemilarynx.

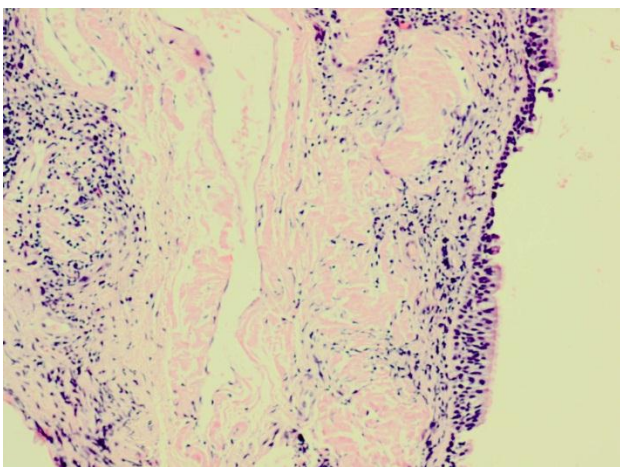


Figure 2: Laryngeal mucosa with amyloid deposited in stroma and around blood vessels.

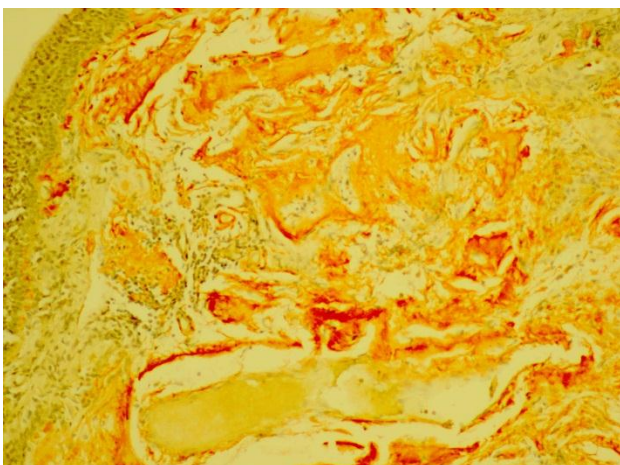


Figure 3: Same area showing red coloured amyloid after congo red staining, without polarization.

The patient was subsequently discussed at the multidisciplinary team meeting and the agreed consensus was to refer the patient for screening for systemic amyloidosis. The patient was again seen at outpatients; upon prompting she denied any abdominal pain, joint

pains or dyspnea. There was no relevant family history of note. She had undergone an appendectomy at the age of 12, which was reported as appendicitis. At the age of 25, the patient had had a missed miscarriage at 11 weeks gestation, for which she underwent an endoscopic removal of products of conception, histology of which showed products of conception.

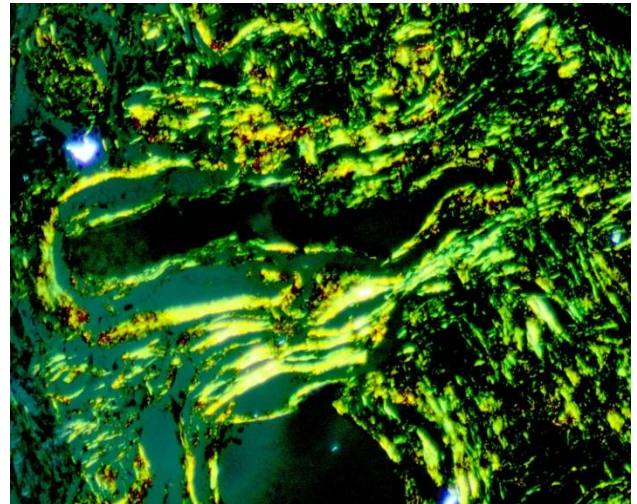


Figure 4: Intense apple green colour of birefringent areas, congo red staining, polarised areas.

The patient was then referred to the haematologist who performed a bone marrow aspirate which was reported as reactive. This was later on repeated and was reported as normal bone marrow appearances. Serum immunoglobulin as well as serum protein electrophoresis were within normal limits. Serum tests for kappa/ lambda were positive at 1.84+(0.26-1.65), while free light chains type lambda, 10.1 mg/l (5.7-26.3) and Free light chains type kappa 18.5 mg/l (3.3-19.4) were within normal limits.

The patient was followed up serially, and a repeat flexible nasendoscopy showed persistent left hemilaryngeal fixation. The patient was still able to vocalise and tolerate oral intake adequately.

In view of the rarity of such a case, the patient was referred for expert opinion abroad.

DISCUSSION

Amyloidosis is characterized by the deposition of amyloid protein in various target organs of the body.¹ Amyloidosis of the larynx is a rare and poorly understood disease of unknown etiology with limited long term studies in the literature. One study done by Thompson LD et al reported only 11 cases in 37 years.² It is a benign disease and slowly progressive. It can manifest as a local tumour or as a diffuse infiltration. It typically presents as hoarseness and can also present with dyspnea, stridor, globus and haemoptysis. If not treated laryngeal amyloidosis may lead to organ malfunction and airway compromise.^{1,3}

Localisation of lesions in the larynx is to the ventricle, false vocal cords, true vocal cords, aryepiglottic folds, and subglottis in that order of frequency. The deposits account for 0.2 to 1.2% of benign tumors of the larynx.⁴

Two theories have been proposed to explain localized amyloidosis of the larynx. The first is due to the presence of plasma cells mixed with the amyloid tissue and the reaction they generate to inflammatory antigens. This is known as secondary amyloid or amyloid of chronic inflammation (AA amyloid). Another, more likely scenario, points to the inability of the body to clear light chains produced by plasma cells located in the mucosal-associated lymphoid tissue known as primary amyloid (AL amyloid).⁴

Diagnosis is initiated by clinical suspicion and then confirmed with a histological examination of tissue.⁵ If tested with congo red staining, it will display apple green birefringence under polarized light.⁶ Following this, a workup of the patient should be done to rule out systemic amyloidosis.²

Most cases of amyloidosis of the larynx represent extension or systemic dissemination and rarely local disease where it is localized only to the larynx. If localized, treatment usually consists of local excision using laser or microlaryngeal instruments with conservation of surrounding tissue. Recent literature has advocated CO₂ laser as the treatment of choice. However treatment can even extend to a partial laryngectomy depending on how advanced it is on presentation. Other treatment options include corticosteroids, radiotherapy and agents like colchicine and melphalan.^{4,5,7}

In a study done by Pribitkin et al 16 cases of amyloidosis in the upper aerodigestive tract showed to behave in a benign, localized manner treatable by surgical resection. Regular follow up with laryngoscopy is indicated to check for recurrence.⁴

CONCLUSION

Laryngeal amyloidosis should be considered and a biopsy taken in any patient with a history of long standing hoarseness not improving on maximal treatment.

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