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Relato de Caso

Laryngeal amyloidosis – case report

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ABSTRACT

Laryngeal amyloidosis is a rare disease characterized by the accumulation of amyloid protein in the larynx, mainly occurring in the supraglottic region. We present a case of a 65-year-old woman, in regular follow-up with an otorhinolaryngologist due to cough and throat discomfort. The diagnosis of laryngeal amyloidosis was made after 7 years of follow-up through biopsy of a supraglottic lesion. The treatment instituted was surgical resection of the lesion and after 11 months of postoperative a new surgical approach was performed due to disease recurrence. So far, there is no gold standard for the treatment of isolated laryngeal amyloidosis, and surgical resection is the method adopted by several specialists to improve the patient's quality of life.

Keywords: laryngeal amyloidosis; case report; supraglottic.

INTRODUCTION

Amyloidosis is a group of diseases characterized by the deposition of extracellular amyloid proteins in various organs and tissues. The etiology of amyloidosis is not yet defined, and it can be divided into systemic and localized amyloidosis[1].

Amongst them immunoglobulin light chain (AL) amyloidosis is the most common type, which occurs when the bone marrow or plasma cells produces unfolded proteins (amyloid light chain proteins) that are deposited in the various organs of the body[2].

Clinical signs are varied, depending on the location and amount of protein deposits. Among the most affected organs are the heart and kidneys, but several combinations of affected organs have already been described. In the case of continued deposit of amyloid protein, complete organ failure may occur. There are several diseases with amyloid characteristics, but not called amyloidosis, such as Alzheimer's Disease and Parkinson's Disease[3].

Although localized amyloidosis occurs mainly in the larynx, this diagnosis is rare, corresponding to 1% of benign laryngeal diseases[4].

The initial complaint of patients with laryngeal amyloidosis is discomfort in the throat, evolving with dysphagia and coughing, which may direct the physician to diagnose laryngopharyngeal reflux, thus delaying the diagnosis and ideal therapeutic approaches.

The aim of this study was to describe a case of laryngeal amyloidosis, from the initial symptoms to diagnosis and treatment.

CASE PRESENTATION

A 65-year-old female, Asian descent patient was in otorhinolaryngological follow-up since 2014, complaining of intermittent discomfort in the throat and cough. The patient had a complex health history, which included hypertension, non-insulin dependent diabetes mellitus, dyslipidemia, breast carcinoma treated in 2003, brain aneurysm treated in 2012 and thyroid carcinoma treated in 2019. All these comorbidities were under control and under specialized medical follow-up.

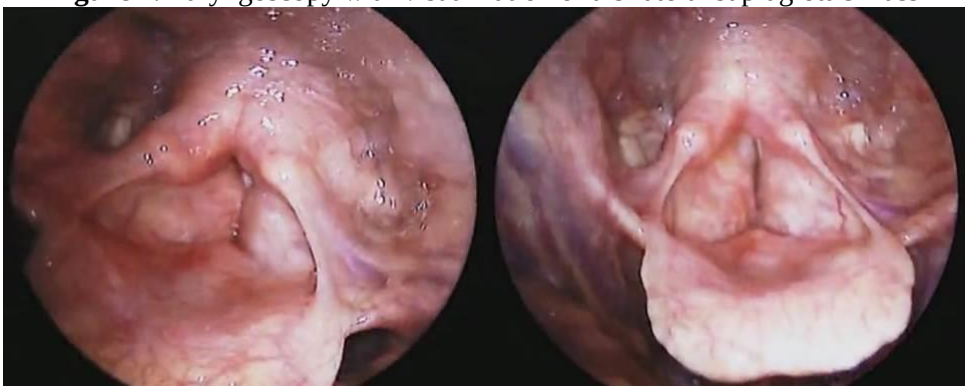
She had received many treatments for an upper airway infection, sporadically with antibiotic therapy. The symptoms progressed with gagging, and the suspicion of Laryngopharyngeal Reflux was raised, therefore a proton-pump-inhibitor was prescribed, without significant improvement.

Three years later, a swallowing videoendoscopy exam was requested due to the maintenance of dysphagia complaint, and the exam was only performed in August/2018, in which it was possible to observe a decreased laryngeal sensitivity. The patient was them guided to perform speech therapy. Despite the correct follow-up of the guidelines, the patient maintains a progressive worsening of dysphagia and due to the previous history of breast adenocarcinoma, the patient underwent neck computed tomography in 2019, in which no lesions detectable by the method were identified in the larynx, in the cervical path of the vagus nerves or the laryngeal recurrent nerves.

By the year of 2020, it was requested a laryngoscopy exam with stroboscopy, but the exam was only performed in June/2021.

The stroboscopy identified fixation of the bilateral vestibular folds and a solid supraglottic lesion (figure 1).

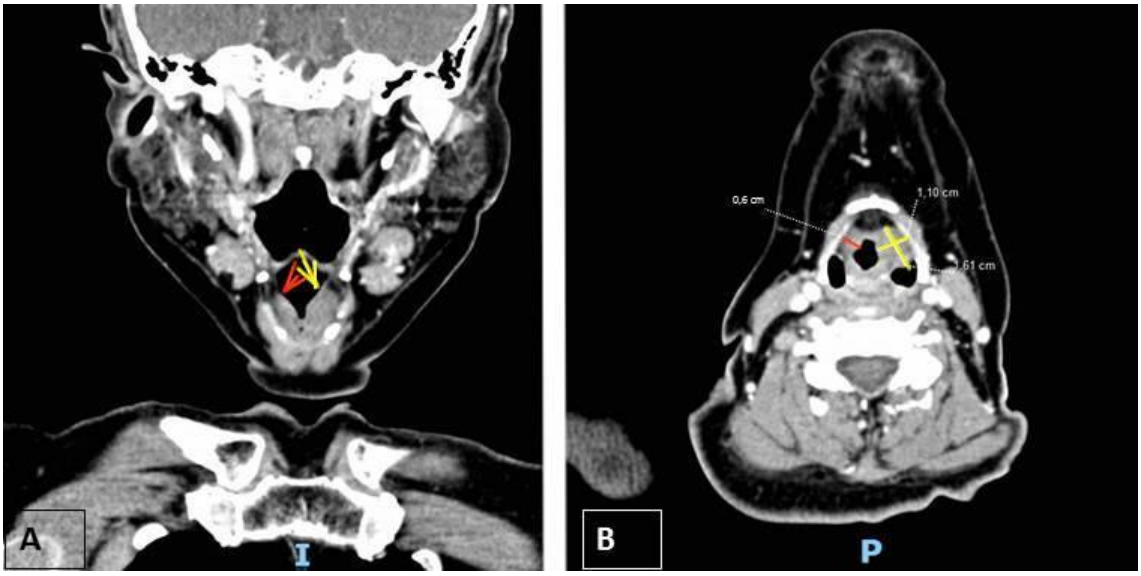
Figure 1. Laryngoscopy with visualization of a bilateral supraglottic mass



Credit source: Author data, 2023

A new neck CT scan performed in 2021 revealed volumetric increase with a grossly oval shape of the anterior two-thirds of the left ventricular band and extending to the adjacent vocal fold, compressing the left paraglottic space and in contact with the thyroid cartilage, measuring about 1. 6cm x 1.6cm x 1.1cm, and another lesion in the anterior third of the right ventricular band, with similar characteristics, but smaller, measuring approximately 1.0 cm x 0.8 cm x 0.6 cm (figure 2).

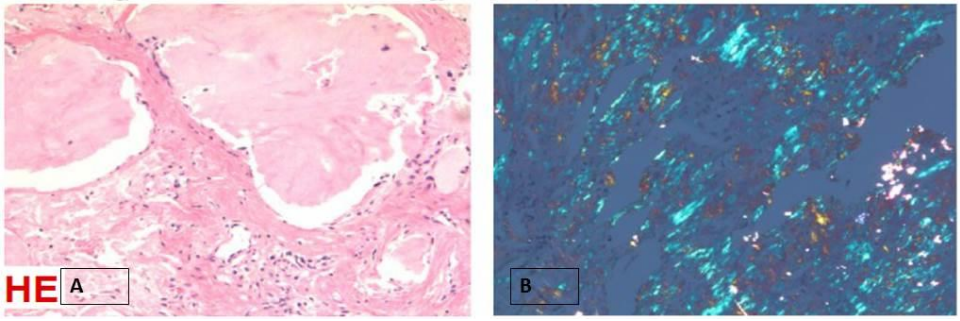
Figure 2. Contrast-enhanced neck computed tomography. A) Coronal cut left supraglottic lesion identified with yellow arrow and right supraglottic lesion identified with red arrow. B) Axial cut, dimensions of the left supraglottic lesion measured with yellow lines and dimension of the right supraglottic lesion measured with red line.



Credit source: Author data, 2023

She underwent biopsy and supraglottic tumor excision with CO2 laser in August/2021 and it was evidenced tissue with content positive to Congo red stained and to polarized light that was compatible with amyloidosis (figure 3). In the postoperative period, the patient used inhaled beclomethasone and pantoprazole. The patient progresses with improvement of dysphagia and cough.

Figure 3. Histopathological analysis. A) Staining with hematoxylin/eosin. B) Congo red coloration



Credit source: Author data, 2023

Since diagnosis, she underwent a new surgical procedure 11 months later due to disease recurrence, however, in the second approach, the lesion was removed with a Diode laser.

DISCUSSION

The rarity of laryngeal amyloidosis makes the diagnosis difficult and delays the correct approach to the patient[1-4], mainly in cases of primary laryngeal amyloidosis, without involvement of other organs. According to the International Society of Amyloidosis, the correct diagnosis and identification allows directing the appropriate treatment of the patient, since there are several therapeutic options[5,6].

There are case reports and literature reviews that are used to better understand the disease, but so far there is no defined gold standard for the treatment of laryngeal amyloidosis[6]. The main symptom associated with laryngeal amyloidosis is hoarseness, unlike the case presented here. This may have been one of the reasons for the delay in our patient's diagnosis.

According to the literature, laryngeal amyloidosis affects men and women in similar proportions, usually in the tenth decade of life[7,8]. Pai et al.,[9] described disease recurrence in 24% of cases, in a mean period of 25.4 months.

In our case, the use of computed tomography helped to exclude differential diagnoses and allowed the follow-up of the disease[10]. However, the use of tomography can result in false negatives depending on the equipment used and the method of image capture, since the lesion of laryngeal amyloidosis can be smaller than the resolution of the tomography.

The use of laser for lesion removal is reported in the literature, but there is no consensus on the best technique for lesion removal. In our case, both the use of CO_2 laser and the Diode laser obtained similar surgical results, however the patient complained of more intense dysphagia after the Diode laser surgery[11], which had

remission after 25 days postoperatively, with the use of corticosteroids..

CONCLUSION

We concluded that laryngeal amyloidosis is a rare entity, and its evolution can cause serious damage to the patient's quality of life. Diagnosis is challenging and may be delayed due to nonspecific symptoms. Due to the rarity of laryngeal amyloidosis, this work can be included in a series of cases and in epidemiological studies for the characterization and management of the disease.

CONSENT

Appropriate written consent and permission was obtained from the patient regarding the writing of case report and the work was approved by the ethics committee (Technical Report Number 5694922).

CONFLICT OF INTEREST

The authors of this publication have no conflicts of interest to disclose.

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