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CASE REPORT

Laryngeal amyloidosis presenting as false vocal fold bulging: clinical and therapeutic aspects[☆]



Amiloidose laríngea apresentando-se como abaulamento em prega vestibular: aspectos clínicos e terapêuticos

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Introduction

The larynx is the main affected site in upper aerodigestive tract amyloidosis, which predominates in men in the fifth decade of life^{1,2} and represents approximately 1% of benign tumors in this organ.¹ Dysphonia is the main symptom^{3,4} and histopathological diagnosis is the gold standard.

This study describes a case of laryngeal amyloidosis in a young female patient, followed at this service from November of 2011 to October of 2013, and aims to demonstrate the clinical diversity of the disease, as well as to draw attention to the differential diagnosis and follow-up.

Case presentation

A 35-year-old black female patient, born and living in São Paulo, SP, Brazil, complained of odynophagia (worse on the

right), throat clearing, globus pharyngeus, and dysphonia for four years. She denied respiratory, gastric, or nasal symptoms. She underwent treatment with omeprazole 40 mg/day for an extended period, but showed no symptom improvement. She denied smoking, alcoholism, or relevant personal and family history.

She presented with a rough voice; anterior oroscopy and rhinoscopy showed no alterations. Telelaryngoscopy showed evidence of submucosal bulging in right vestibular fold, with no alterations in mobility, and edema/hyperemia of the interarytenoid space (Fig. 1A).

The hypothesis of a saccular cyst was raised, with lesion excision followed by postoperative speech therapy recommended: these were performed without complications.

Histopathological analysis result was laryngeal amyloidosis (Fig. 1B and C) and, therefore, renal and hepatic function tests were requested, as well as an electrocardiogram, which showed normal results. The patient was referred for follow-up at the rheumatology department.

The patient did not undergo otorhinolaryngology/rheumatology follow-up and came to the clinic one year after surgery, asymptomatic. New renal, hepatic, and cardiovascular function tests, as well as rigid telelaryngoscopy were performed; the results were normal. Semi-annual outpatient follow-up was indicated and patient was once again referred to the rheumatology department.

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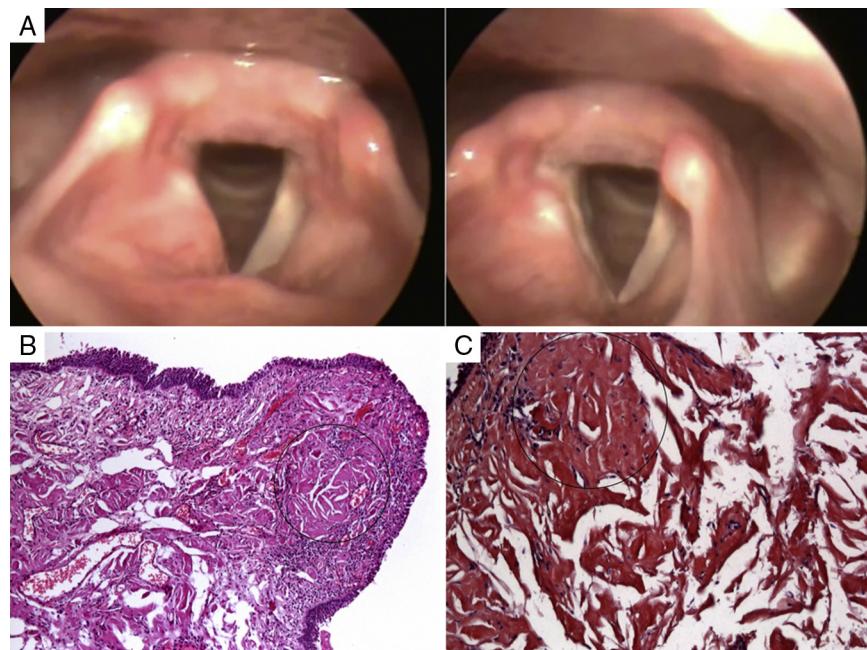


Figure 1 (A) Telelaryngoscopy: bulging in the right vestibular fold, without glottic or subglottic involvement. (B) Fragment showing respiratory-like epithelium with mild lymphomononuclear infiltrate in the stroma and apparent deposition of amorphous eosinophilic material at hematoxylin-eosin staining, in the outlined space (H&E, 200×). (C) Same area shown in B with paler appearance than the rest of the sample, when stained with Congo red (Congo red, 400×).

Discussion

Laryngeal involvement usually results from the localized form of amyloidosis⁴ and, contrary to what occurs in other head and neck sites, it is rarely involved in systemic disease cases.³

The lesion occurs mainly in the vestibular fold (55%), presenting as subepithelial edema or nodular formation.¹ The diagnosis is rarely suspected and is often only attained after anatomopathological analysis, which discloses typical positive birefringence under polarized light after Congo red staining, showing a greenish color.⁵ In the present case, a saccular cyst was suspected, a disease characterized by obstruction/atresia of the laryngeal saccule orifice, with consequent mucus retention and submucosal bulging. Sarcoidosis, metastatic tumors, polyps, malignant neoplasms, and salivary gland tumors are other possible differential diagnoses.^{4,6}

Some authors suggest the assessment of hepatic and renal functions, as well as electrocardiography and endoscopic assessment of multifocal amyloidosis of the respiratory tract in the initial investigation of systemic involvement.⁴ The normal results of these tests in this patient, in addition to the asymptomatic postoperative course, suggest a case of localized amyloidosis. However, the importance of a full rheumatological assessment in these patients is noteworthy.

Treatment varies from observation to surgical excision.³ Immunosuppressive agents and radiotherapy have shown to be ineffective and can accelerate the amyloid deposition.⁵ In localized and symptomatic laryngeal

pictures, endoscopic surgical excision is the treatment of choice.³ In contrast, in extensive cases without imminent airway obstruction, the expectant conduct can be chosen, considering the slowly progressive nature of the disease.²

Prognosis is excellent and the long-term follow-up should be conducted for at least five to seven years, due to the possibility of late recurrence and systemic involvement.^{4,5}

Final comments

Laryngeal amyloidosis is a disease whose diagnostic suspicion is difficult, considering its clinical diversity and similarities to other more relevant diseases in the population. The prognosis is excellent; however, long-term follow-up should be performed, due to the possibility of late recurrence of the disease and systemic involvement.

Conflicts of interest

The authors declare no conflicts of interest.

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