

A Novel Approach to the Treatment of Laryngeal Amyloidosis

Abstract

Objective: To describe our experience with the non-surgical management of laryngeal amyloidosis. Management of laryngeal amyloidosis for years has consisted of excision of lesions using the CO₂ laser or cold steel- a process that is plagued with complications including hemorrhage, stenosis or persistent obstruction.

Study Design: Retrospective case series of patients with laryngeal amyloidosis treated in a tertiary care laryngology practice.

Methods: Charts from all patients with laryngeal amyloidosis between 2010 and 2016 were reviewed. Course of treatment, laryngeal imaging before and after treatment, subjective voice outcomes and symptoms were analyzed.

Results: The use of intralesional dexamethasone is an effective treatment for hoarseness and infraglottic obstruction due to amyloidosis.

Conclusions: These cases demonstrate the efficacy of intralesional steroid injection in the treatment of laryngeal amyloidosis. It is safe and effective, providing long lasting results.

Introduction

Amyloidosis is a heterogeneous family of extracellular proteinaceous deposits with characteristic microscopic, histochemical and ultrastructural features. Deposits of amyloid in the larynx are rare, accounting for 0.2 to 1.2% of benign tumors of the larynx. It has a slowly progressive growth pattern.

The laryngotracheal system is the second most common site of head and neck involvement in amyloidosis. 90% of patients with systemic amyloidosis will develop amyloid deposits in the upper aerodigestive tract. There is no evidence to suggest that patients with localized amyloid will develop systemic disease.

Amyloid in the larynx can be identified as subepithelial extracellular deposits of acellular, homogenous and amorphous, eosinophilic material displaying apple-green birefringence with polarized light when stained with Congo red. The mucosal surface is usually intact, and the underlying lesion may be nodular or flat, with a yellow, pink or bluish hue. It usually occurs in nodules or polypoid lesions and can be located anywhere in the larynx or trachea. Endoscopic appearance is described as submucosal plaques and nodules with a cobblestone appearance.

Laryngeal amyloidosis primarily impacts individuals between 40 and 60 years old. Patients with localized laryngeal amyloidosis usually present with longstanding hoarseness or dyspnea. Slowly progressive cases with cough, stridor, odynophagia, and rarely hemoptysis have been described. Symptoms are caused by the physical presence, size, and site of the tumor.

The diagnosis is made by a high degree of suspicion on the basis of history and a characteristic appearance on direct laryngoscopic examination as well as on histologic examination of a biopsy.

In treatment preservation of the airway and voice are the first priorities. To date endoscopic CO₂ laser excision of the laryngeal lesions has been the first line of therapy. However this is fraught with complications including bleeding and recurrence.

The purpose of this investigation is to describe our experience with the non-surgical management of laryngeal amyloidosis.

Methods and Materials

This study is a retrospective case series of patients with laryngeal amyloidosis treated in a tertiary care laryngology practice.

In a retrospective manner charts from all patients diagnosed with laryngeal amyloidosis between 2010 and 2016 were reviewed. Three patients were identified who were diagnosed with laryngeal amyloidosis during the study period. The course of treatment, laryngeal imaging before and after treatment, subjective voice outcomes and symptoms were analyzed.

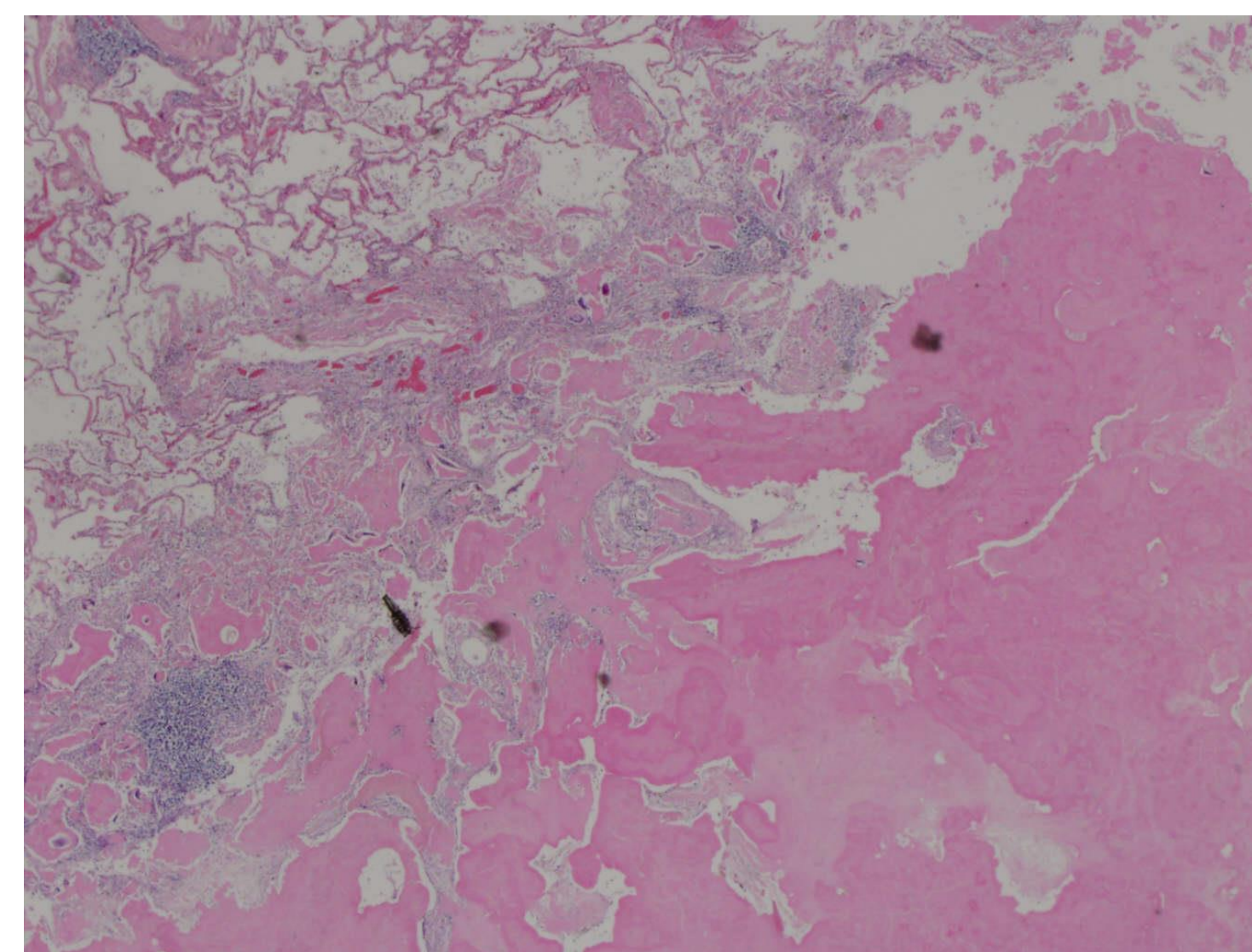


Figure 1. Patient PT laryngeal biopsy demonstrating the presence of amyloid

Results

In the six year study period three patients were diagnosed with laryngeal amyloidosis. All three of them, treated with intralesional steroid injection demonstrated improvement of airway obstruction.

The use of intralesional dexamethasone has effectively treated patients with hoarseness and infraglottic obstruction due to laryngeal amyloidosis. Intralesional dexamethasone is a safe and effective treatment, providing long lasting results including improved phonatory ability and resolution of shortness of breath due to obstruction.

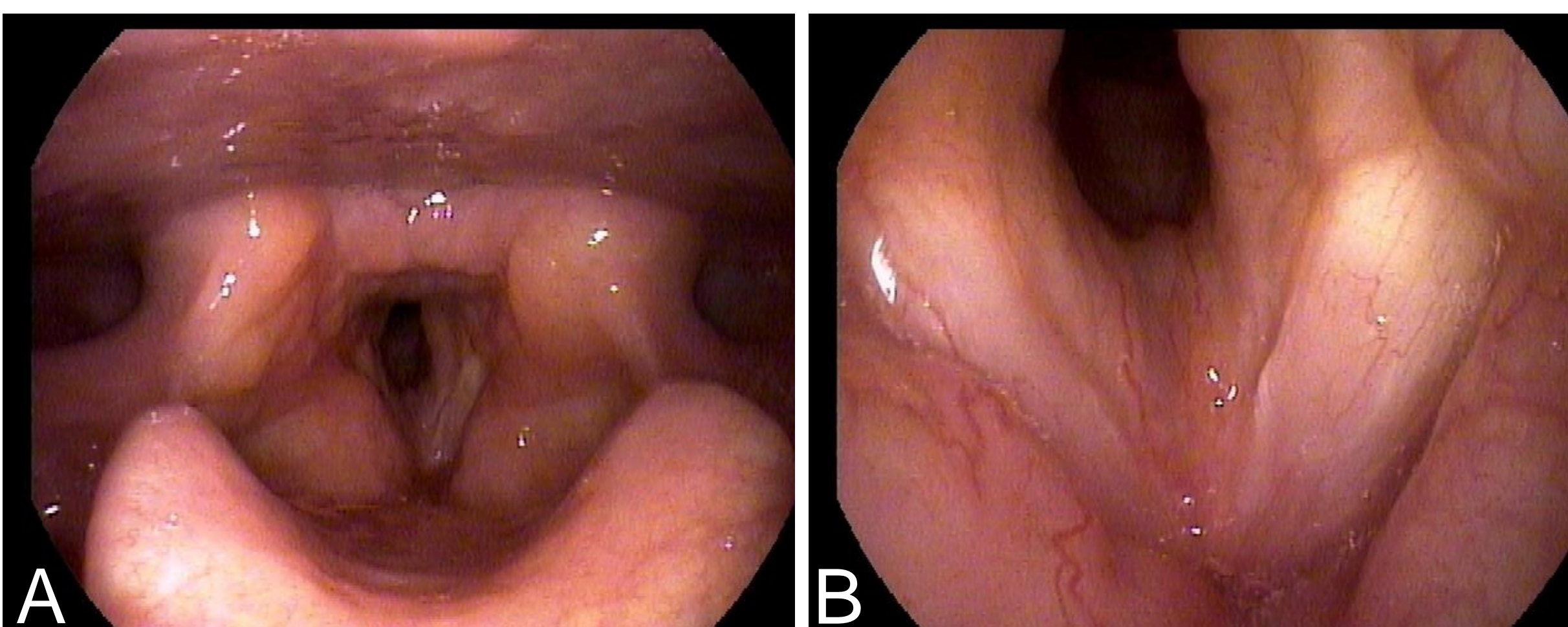


Figure 2. A: Patient PT presented with diffuse polypoid inflammation of the larynx. No mucosal waves could be observed. B: Circumferential polypoid thickening of the infraglottis with 30% narrowing of the airway and posterior glottis edema and erythema.

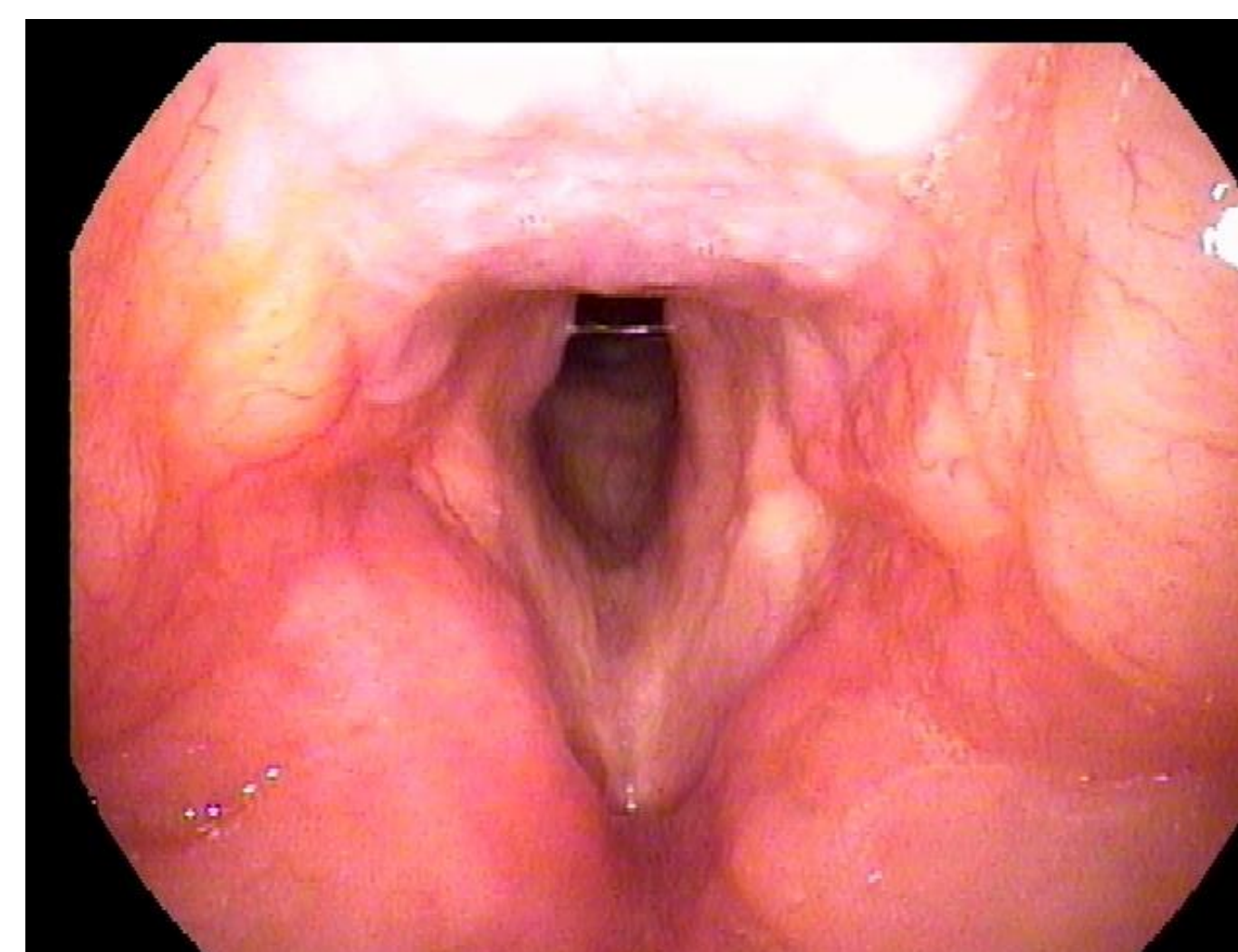


Figure 3. Markedly improved airway patency in patient PT one month after initial intralesional steroid injection.

Case 1

A 35 year old female (PT) presented with a history of progressively worsening hoarseness over the past 7 years. She noted increased strain and effort with phonation. In addition she complained of an abnormal laugh. She denied cough and shortness of breath.

Case 2

Patient JB is a 68 year old male initially diagnosed with laryngeal amyloidosis 19 years ago. Since that time he's undergone multiple treatment approaches including CO₂ and KTP laser as well as cold resection of lesions. Each time the patient has noted improvement of his voice immediately after the procedure but eventual worsening as lesions recur. The patient complains of a raspy, weak voice that is effortful and low in volume. In addition he reports a loss of low range.

The patient was initially injected with intralesional dexamethasone in September 2014. Since that time he has noted significant improvement in his voice. He now returns to clinic annually for examination and steroid injection of any new lesions.

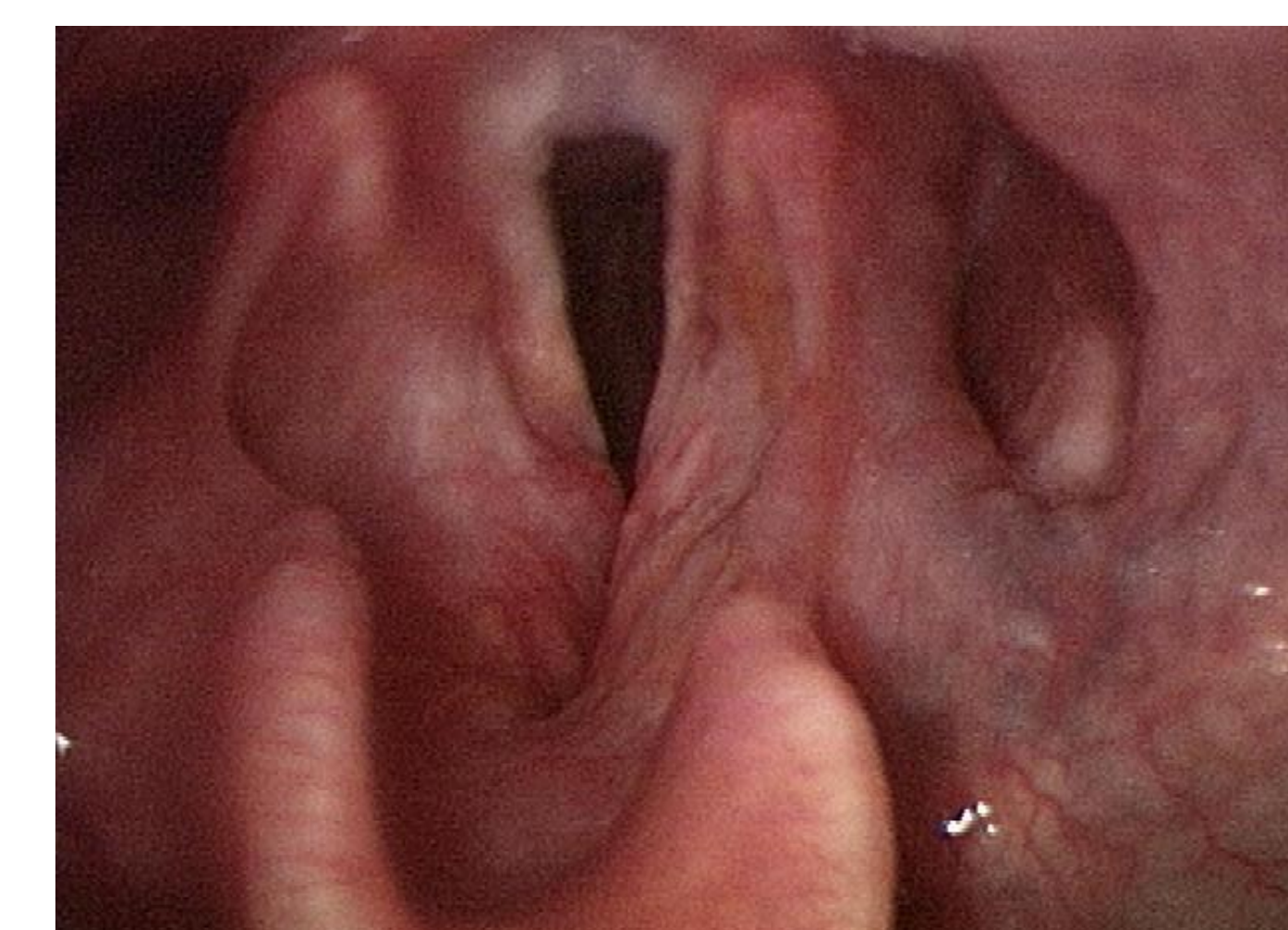


Figure 4. Laryngeal examination of patient JB prior to steroid injection. There is diffuse involvement of the glottis and amyloid deposits within the anterior supraglottis.



Figure 5. Markedly improved Laryngeal examination of patient JB 3 months after steroid injection. The airway is widely patent. Small plaques are seen in the anterior glottis bilaterally.

Discussion

Intralesional steroids represent a successful and safe treatment for laryngeal amyloidosis. The goal of treatment is to maintain an adequately patent airway with near normal voice.

Currently, the most effective treatment available is thought to be microdirect laryngoscopy with CO₂ laser excision. Multiple surgeries are required to remove residual and recurrent amyloid deposits. Cold resection of amyloid lesions can be complicated by significant bleeding. Although CO₂ laser excision may be advantageous in terms of hemostasis it increases the risk of scarring and poor voice outcome.

Intralesional steroid injection provides a viable alternative to repeated surgery. It is best used in conjunction with CO₂ laser removal of large lesions. The injection of steroids after removal of large lesions also prevents their recurrence – resulting in a need for fewer surgical interventions.

Adjuvant therapies including irradiation, chemotherapy and systemic steroids have proven no benefit in the treatment of laryngeal amyloidosis.

Conclusions

These cases demonstrate the efficacy of intralesional steroid injection for the non-surgical management of laryngeal amyloidosis. It is an effective method to avoid the risks of surgery which include hemorrhage, postoperative stenosis and persistent obstruction. This must be added to our armamentarium of treatments for laryngeal amyloidosis.