CASE REPORT

Nasopharyngeal Amyloidosis Extending to Larynx: A Rare Clinical Presentation

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Abstract

Aim: Nasopharyngeal amyloidosis is rare, with very few reports in the literature. We report a case of amyloidosis of the nasopharynx extending into the larynx mimicking a tumoral process and compromising the airway, a unique clinical presentation.

Background: Amyloidosis is a condition characterized by the deposition of extracellular proteinaceous material, resulting in damage to the tissues. A total of 19% of cases involve the head and neck with the oropharynx and larynx being the most affected organs. Localized amyloidosis is a rare and slow-growing lesion and is considered benign tumor, but it may be aggressive locally.

Case description: A 49-year-old female presented with voice change and breathlessness. On clinical evaluation, multiple swellings involving the nasopharynx and larynx compromising the airway were noticed and a biopsy was done, which confirmed our primary clinical suspicion of Amyloidosis and was planned for staged coblation-assisted excision of the lesions to secure adequate airway.

Conclusion: A complete clinical and paraclinical work-up is required for the positive diagnosis of primary or secondary amyloidosis. Morbidity is high so postsurgical follow-up is essential.

Clinical significance: The localized form usually follows a slow and benign clinical course with a favourable prognosis of 90.6%. The principal differential diagnosis is a malignant tumor, hence the importance of histopathologic diagnosis. The recurrence rate approximates zero, but the extremely rare presentation and slow progression of this pathology make surveillance mandatory.

Keywords: Amyloidosis, Case report, Coblation, Congo red stain, Flexible video laryngoscopy.

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Introduction

Amyloidosis is a pathology characterized by the deposition of extracellular proteinaceous material, resulting in damage to the tissues. Amyloidosis can be classified as primary (70%), and secondary (3%) and further into localized and systemic. Amyloidosis in head and neck region is rare and is most frequently localized, with only few systemic forms.[1–3] A total of 19% of the cases involving the head and neck area mainly affect the oropharynx and the larynx.[4] The presenting symptoms include nasal obstruction, recurrent epistaxis, posterior rhinorrhea or otologic complaints involving eustachian tube dysfunction.[1] Proteinuria, which is often the first symptom in the systemic type, and other manifestations like peripheral neuropathy, dementia, cognitive dysfunction, and organ dysfunction of the liver, kidney, heart, or spleen were absent, ruling out systemic involvement.[1] Amyloidosis of the nasopharynx is rare, with very few reports in the literature. We report a case of nasopharyngeal amyloidosis extending into the larynx mimicking a tumoral process.

Case Description

A 49-year-old female presented with a history of progressive voice change for 1 year, worsened in the last 6 months, with no associated nasal or laryngeal symptoms.

On clinical examination, the nasal cavity and the oral cavity were normal.

On further evaluation with flexible video laryngoscopy, showed multiple swellings with a yellowish hue seen in the nasopharynx, oropharynx, and supraglottis extending to the right side of the glottis, right vocal cord appeared bulky with normal mobility. Left cord normal (Fig. 1).

The patient was planned for a biopsy of the swelling under the laryngeal mask airway; a biopsy was taken from the nasopharynx, the mass was firm in consistency, with profuse bleeding.

Histopathology revealed focal respiratory epithelium with the presence of prominent eosinophilic amorphous acellular material with few plasma cells.

The extracellular deposits were congophilic and showed apple–green birefringence under polarized light, confirming the diagnosis (Fig. 2).

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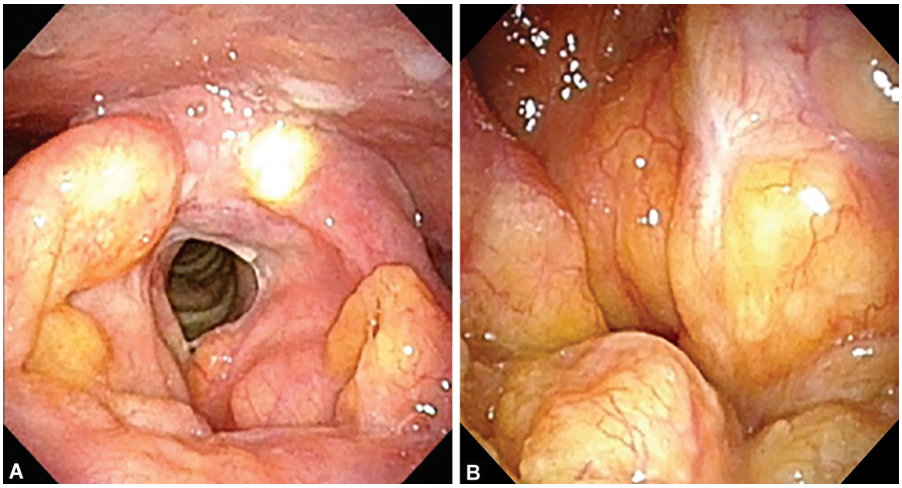
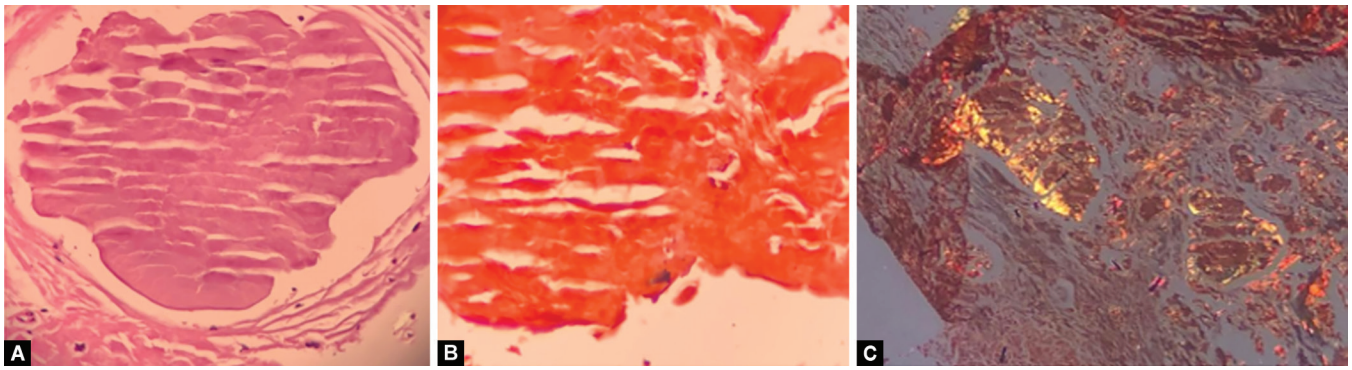
Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.



Figs 1A and B: Preoperative yellowish hue seen in the nasopharynx, oropharynx, and supraglottis extending to right side of the glottis, left cord normal



Figs 2A to C: (A) Histopathology—focal respiratory epithelium with presence of prominent eosinophilic amorphous acellular material with few plasma cells; (B and C) Congo red stain—the extracellular deposits were congophilic and demonstrated apple–green birefringence under polarized light, confirming the diagnosis

Staged excision planned even though the nasopharyngeal lesion was extensive, the laryngeal lesion was considered first to relieve the airway obstruction.

The tissues were immunoassayed with CD138 test; no substantial plasma cells were seen.

Considering, the histopathological diagnosis patient underwent a tracheostomy to secure the airway followed by staged microlaryngoscopic debulking of the swellings done with coblator after taking an incision over the lesion in the right false vocal cord pushing the true vocal cord inferiorly to preserve the voice.

The patient was decannulated after 48 hours after confirming adequate airway with flexible video laryngoscopy.

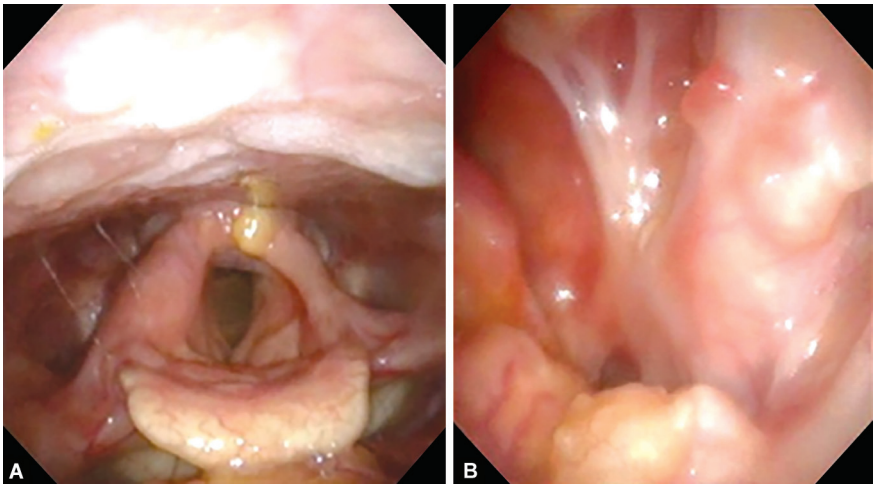
She was referred to a medical oncologist for further evaluation to rule out Systemic amyloidosis. She was advised for a two-dimensional echocardiogram (2D echo), and renal, and liver function tests showed normal values. She was also evaluated with Serum free light chain (kappa and lambda) assay and showed elevated kappa free chain—39.2 (6.7–22.4 mg/L) levels, indicating primary amyloidosis [amyloid light-chain (AL) type] and serum protein electrophoresis, in this myeloma band was not detected and total protein was normal—7.2 (6–8 gm/dL). The elevated kappa light chain indicates primary systemic amyloidosis AL type, but since the patient had no systemic complaints and all other biochemical parameters including total proteins were normal, treatment of systemic amyloidosis was not indicated.

The patient was followed up at regular intervals for 1 year with a flexible video laryngoscopy examination, patient was symptomatically better initially, she then presented with difficulty breathing post 1-year follow-up. Flexible video laryngoscopy showed multiple amyloidosis swellings over the true and false vocal cords and in the nasopharynx and underwent coblation-assisted debulking of the swellings until adequate airway was achieved (Fig. 3).

Discussion

Amyloid was first described by Rokitansky in 1842. It was not until 1853 that Virshow termed these lesions "amyloid" in the belief that carbohydrate was the main constituent.[4] Even though the localized amyloidosis is a rare and slow-growing pathology and considered benign tumors, they may be aggressive locally leading to osteolysis.

The main differential diagnosis is a malignant tumor, hence the importance of histopathological confirmation. The recurrence rate approximates zero, but the rare presentation and slow progression of this condition make the surveillance mandatory.[4]



Figs 3A and B: Postoperative post debulking of the swellings until adequate airway was achieved

The onset of amyloidosis is usually in the fourth to eighth decades. It mainly affects males, with a sex ratio of 3–1.5 but the etiology and exact pathogenesis remain unknown.[4]

The localized form usually follows a slow and benign clinical course with a favorable prognosis of 90.6%.[3]

Conclusion

A complete clinical and paraclinical work-up is required for the positive diagnosis of primary or secondary amyloidosis. It is a benign but locally aggressive pathology. Since the disease involves the airway, morbidity is high; so, postsurgical follow-up is essential to look for localized complications and even to rule out systemic involvement.

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