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Case Report

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Decade-Long Progression of Localized Amyloidosis from the Larynx to the Nasal Cavity-A Case Report

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Abstract

Localized amyloidosis of the upper aerodigestive tract is an uncommon and benign condition characterized by extracellular amyloid deposition. While the larynx is the most frequently involved site, progressive, multifocal mucosal involvement remains exceedingly rare. We report the case of a 58-year-old male initially diagnosed with localized laryngeal amyloidosis confirmed via Congo red staining. The initial laryngeal lesions were excised using conservative endoscopic surgery to both confirm the diagnosis and alleviate symptoms. However, no additional treatment was recommended by the rheumatology department since the patient's further evaluations did not indicate a diagnosis of systemic amyloidosis. Over an 11-year follow-up period, sequential progression to the nasopharynx was observed after five years and to the nasal cavity after 11 years. The patient remained free of systemic involvement throughout, confirmed by comprehensive evaluations including serum and urine immunofixation, renal and hepatic panels, and 24-hour urine analysis. Symptomatic relief was achieved with a series of conservative surgical interventions addressing airway and auditory symptoms. The patient remains under active surveillance. This case highlights a rare, progressive pattern of localized amyloidosis involving contiguous mucosal sites in the upper aerodigestive tract. It underscores the importance of long-term follow-up, systematic evaluation to exclude systemic disease, and the efficacy of conservative surgical management. To our knowledge, this is the first reported case of progressive, segmental spread of localized amyloidosis involving the larynx, the nasopharynx, and the nasal cavity over more than a decade.

Keywords: Amyloidosis, larynx, nasopharynx, nasal cavity, upper aerodigestive tract, disease progression, case reports

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Introduction

Amyloidosis represents a disorder in which misfolded proteins accumulate within the extracellular matrix, leading to progressive tissue dysfunction. The condition is broadly divided into two clinical entities: systemic and localized variants (1,2). When limited to the upper airway region, the larynx is the site most frequently involved, comprising approximately 0.2-1.2% of benign laryngeal lesions (3). Diagnosis relies on histopathological demonstration of amyloid material that exhibits a distinctive apple-green birefringence under polarized light after Congo red staining (4). These deposits exhibit structural diversity, and several amyloid subtypes have been identified: (i) AL or light-chain amyloid, often linked with plasma-cell dyscrasias and appearing in either systemic or localized forms; (ii) AA or amyloid-associated type, a reactive form secondary to chronic inflammatory processes; and (iii) Aβ or beta-amyloid type, predominantly observed within the central nervous system (4,5). A definitive diagnosis requires histopathological examination, and determining the type of amyloidosis is essential for evaluating systemic

or secondary involvement. Localized forms are managed with conservative surgeries, while systemic forms may require chemotherapy. Thus, confirming amyloid presence alone is insufficient; subtype identification is equally critical, as it directly guides treatment and prognosis.

In this case report, we present the long-term follow-up of a patient with segmental progression of amyloid involvement in the larynx, the nasopharynx, and the nasal cavity.

Case Presentation

A 58-year-old male patient presented to our clinic with a complaint of dysphonia. He had no known comorbidities, no history of malignancy, or previous radiotherapy. Endoscopic examination of the nasal cavity and nasopharynx was unremarkable; however, endoscopic laryngeal evaluation revealed bilateral, orange-colored mass lesions on the ventricular bands, raising suspicion of malignancy and prompting a biopsy as shown in Figure 1a. Thus, biopsy specimens were obtained via conservative surgical excision. Excision was performed for diagnostic purposes; therefore, surgical margins were not evaluated.

The laryngeal biopsy showed amorphous and homogenous, pale pink-colored deposits in the extracellular spaces. The biopsy specimen exhibited classic apple-green birefringence under polarized light with Congo red staining, confirming a diagnosis of laryngeal amyloidosis. Positive staining was detected in immunohistochemical staining with Amyloid AA, as shown in Figure 2. To exclude systemic involvement, serum and urine immunofixation electrophoresis revealed no monoclonal components. Kidney and liver function tests were within normal limits, and 24-hour urine analysis demonstrated no significant proteinuria. In the absence of systemic amyloidosis findings, the patient was classified as localized laryngeal amyloidosis and placed under active surveillance.

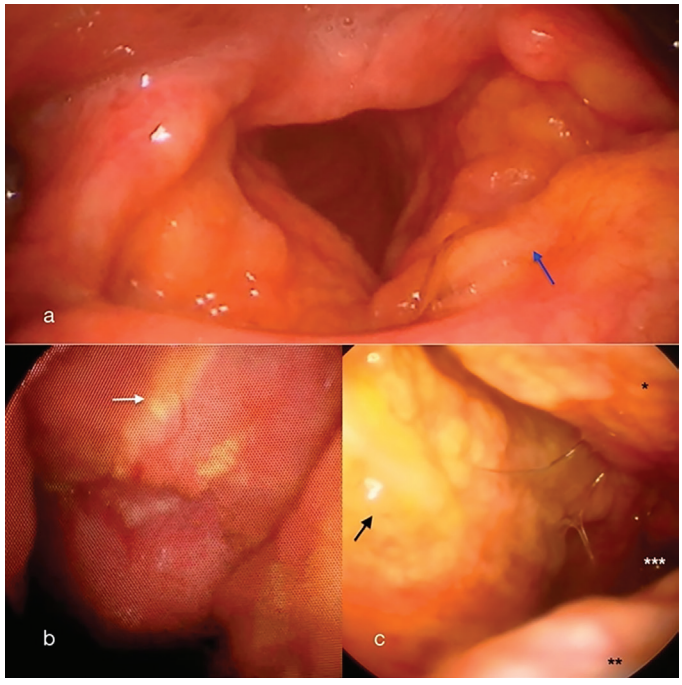

Figure 1. a) Localized amyloid appearance in the larynx at the initial diagnosis. Blue arrow: amyloid deposits, **b)** Amyloid deposits observed in nasopharynx. Five years after initial diagnosis. White arrow: Amyloid deposits in the nasopharynx; **c)** Amyloid deposits observed in the left nasal cavity 11 years after initial diagnosis.
*: Middle turbinate, **: Inferior turbinate, ***: Choana. Black arrow: amyloid deposits on the posterior part of the nasal septum

After 5 years, the patient returned with right-sided hearing loss and a sensation of a foreign body in the pharynx. Examination revealed right-sided otitis media with effusion and a clustered, orange-colored mass around the right torus tubarius and midline in the nasopharynx as shown in Figure 1b, resembling the mucosal features of the previous laryngeal lesion. Amyloid deposits were noted in the ventricular bands and epiglottis with slight progression; though there was no evidence of airway obstruction, as also demonstrated on magnetic resonance imaging (MRI), as shown in Figure 3a. Contrast-enhanced MRI demonstrated soft tissue hypertrophy narrowing the right posterior nasopharynx, a finding that becomes more evident when compared with the patient's initial MRI obtained at presentation (Figure 4a vs. Figure 4b), along with loss of aeration in the right middle ear and mastoid cells. A tympanostomy tube was placed in the right ear. A conservative surgical excision of the mass obstructing the torus tubarius was performed. Similarly, Congo red staining was positive, confirming amyloidosis in the nasopharynx. The patient experienced relief of hearing loss postoperatively. No sign of systemic amyloidosis was detected upon re-evaluation. The patient subsequently missed follow-up due to the coronavirus disease-2019 pandemic and absence of symptoms.

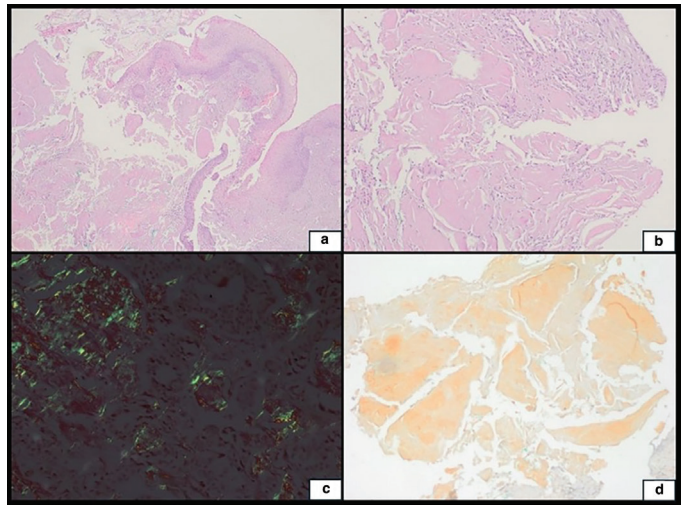

Figure 2. Amyloid deposition in larynx biopsy at the patient's initial presentation. **a)** H&E staining X100, **b)** H&E staining X200, **c)** Congo-red staining x200, **d)** Amyloid AA staining X200
H&E: Hematoxylin & eosin

Eleven years after the initial presentation, the patient returned with nasal obstruction and right-sided hearing loss. Endoscopic examination revealed a nasopharyngeal mass originating from the posterior wall that caused significant anterior narrowing, along with non-bulging mucosal amyloid deposits on the posterior one-third of the nasal septum as shown in Figure 1c. Laryngeal examination showed amyloid deposits extending over the left arytenoid and the laryngeal surface of the epiglottis without airway obstruction. These findings were also visualized on MRI, as demonstrated in Figure 3b. Laryngeal deposits were stable and non-obstructive; therefore, no surgical intervention was performed for the laryngeal component. Surgical excision of the nasopharyngeal and nasal cavity lesions performed to relieve the nasal obstruction. Amyloid deposits were detected via Congo red and amyloid AA staining in the biopsy, as shown in Figure 5. Systemic amyloidosis evaluation was again negative. The patient had an uneventful postoperative recovery and remains under surveillance. The patient reported being satisfied with the surgical results, noting significant improvements in both breathing and hearing. Written informed consent for publication was obtained from the patient for the data included in this case report.

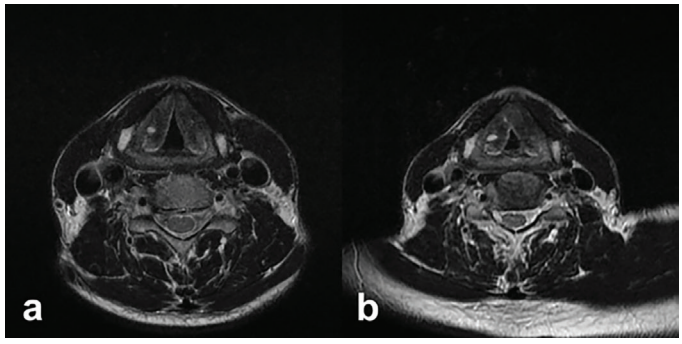

Figure 3. Axial T2-weighted MRI sections of the patient. Although an increase in amyloid deposits was observed during follow-up, no significant airway narrowing was noted. **a)** MRI obtained in initial diagnosis, **b)** MRI obtained in 11 years after initial diagnosis
MRI: Magnetic resonance imaging

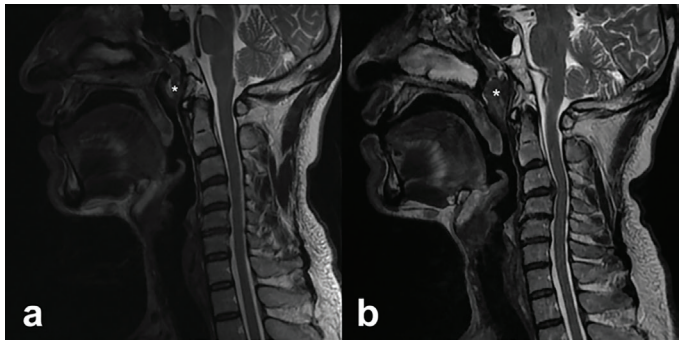

Figure 4. Sagittal T2-weighted MRI images of the patient show soft tissue enlargement in the nasopharynx. **a)** MRI obtained in initial diagnosis, **(b)** MRI obtained in 5 years after initial diagnosis.
*: Soft tissue in the nasopharynx, MRI: Magnetic resonance imaging

Discussion

Localized amyloidosis is a benign disease that presents with tumor-like amyloid deposits that may cause site-specific symptoms. Although amyloid deposition in the upper aerodigestive system is rare, the most commonly affected sites include the larynx (60%; the most frequently involved site), the oropharynx (23%), the trachea (9%), and the orbit (4%) (1). Nasal cavity and nasopharyngeal involvement (approximately 3%) represent uncommon manifestations. Sakagiannis et al. (2) reported only 32 patients with nasopharyngeal amyloidosis between 1935 and 2017, while Naidoo et al. (3) found merely 15 patients with sinonasal amyloidosis from 1946 to 2012. In recent decades, case presentations of laryngeal and nasopharyngeal amyloidosis have increased, possibly related to the improvement in diagnostic capabilities through routine endoscopic head and neck examinations and an increase in clinical awareness (4-6).

This case report obviously illustrates the segmental progression of localized amyloidosis in the upper aerodigestive system in the long-term. Initially, the amyloid lesions manifested in the larynx subsequently developed in the nasopharynx after a 5-year interval and ultimately involved the nasal cavity after 11 years of follow-up. This stepwise anatomical progression challenges the conventional view of localized amyloidosis as a static condition, demonstrating its potential for clinically significant advancement along contiguous upper aerodigestive tract mucosa. Although most reported cases generally remain confined to a single anatomical site, emerging evidence of multifocal mucosal involvement corroborates this progressive disease pattern (5,6). In addition, it is important to remember that localized

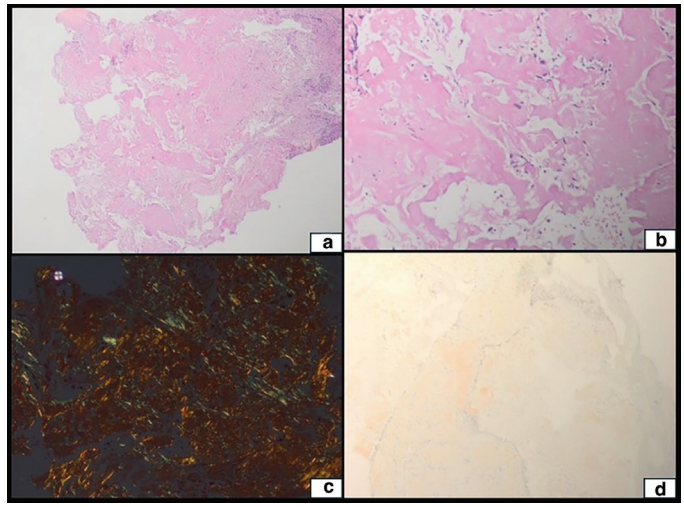

Figure 5. Amyloid deposits in nasopharynx biopsy. **a)** H&E staining X100, **b)** H&E staining X400, **c)** Congo-red staining x200, **d)** Amyloid AA staining X200
H&E: Hematoxylin & eosin

amyloidosis in the upper airway may mimic malignancy, and histopathological confirmation is essential for diagnosis. Amyloid deposits may lead to a variety of symptoms such as hearing loss, epistaxis, dysphagia, globus, and nasal and airway obstruction according to the involved region/s.

While most patients with amyloid deposits involving the upper aerodigestive system have localized disease, systemic or secondary amyloidosis must be excluded both at initial diagnosis and during follow-up particularly when disease progression occurs. Comprehensive evaluation should include serum and urine protein electrophoresis, serum free light chain analysis, renal and liver function tests, and 24-hour urine protein quantification, essential investigations for excluding AL-type amyloidosis (4,7). In our case, systemic evaluation was conducted regularly, and the patient was diagnosed with localized amyloidosis of the larynx, the nasopharynx, and the nasal cavity without evidence of systemic involvement. Systemic surveillance included annual serum and urine immunofixation, renal and hepatic function tests, and 24-hour urine protein quantification to exclude systemic AL amyloidosis.

The mainstay of treatment for systemic amyloidosis is chemotherapy (8). For localized disease, surgical intervention remains the mainstay of therapy (9). The surgical approach should be individualized, ranging from conservative surgeries to more extensive radical resections. Conservative surgeries are generally preferred to relieve symptoms while preserving organ functions. When addressing airway patency, voice quality, or Eustachian tube dysfunction, meticulous tissue preservation is paramount. Radical procedures may be considered for selected patients with recurrent disease following conservative surgeries (1).

Study Limitations

A strength of this case is the extensive long-term follow-up that demonstrates the natural history of progressive localized amyloidosis. The main limitation, however, is the inability to perform serial molecular subtyping of the amyloid deposits, which could have provided additional insights into the evolution of the disease. In the presented case, conservative surgical approaches have effectively achieved symptomatic relief while maintaining anatomical integrity.

Conclusion

To the best of our knowledge, this case represents the first documented example of localized amyloidosis showing a decade-long, stepwise progression from the larynx to the nasopharynx and, subsequently, to the nasal cavity. This rare pattern emphasizes three key clinical points: (i) localized amyloidosis may exhibit progressive, multifocal mucosal spread, (ii) thorough and repeated systemic evaluation is essential to confidently exclude systemic disease throughout follow-up, (iii) conservative surgical approaches can effectively provide durable symptomatic relief while preserving anatomical and functional integrity. Long-term surveillance remains fundamental in managing this uncommon condition.

Ethics

Informed Consent: Written informed consent for publication was obtained from the patient for the data included in this case report.

Footnotes

Authorship Contributions

Concept: U.K., G.E., Design: G.E., Data Collection and/or Processing: U.K., S.E.A., A.T., Analysis or Interpretation: A.T., U.U., G.E., Literature Search: U.K., S.E.A., Writing: U.K., G.E.

Conflict of Interest: The authors declare that they have no conflict of interest.

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Main Points

- Localized amyloidosis of the upper aerodigestive tract is rare and may mimic malignancy.
- This case demonstrates sequential progression from larynx to nasopharynx and nasal cavity over 11 years.
- Conservative surgical excision provided effective symptomatic relief while preserving function.
- Long-term surveillance is essential due to the risk of multifocal progression despite localized disease.

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