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A Unique Case of Localized Laryngeal Amyloidosis

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CASE REPORT

ABSTRACT

Laryngeal amyloidosis, a rare pathological condition characterized by the abnormal deposition of amyloid proteins within the laryngeal tissues, presents a unique diagnostic and therapeutic challenge. We report the case of a 50-year-old woman with no significant medical history, presenting with an 8-month history of worsening hoarseness. Clinical examination revealed submucosal thickening in the left supraglottic area, while imaging confirmed irregular thickening on the anterior aspect of the left vocal cord, involving adjacent structures. Subsequent direct laryngoscopy and biopsy confirmed amyloidosis. A systemic workup for systemic amyloidosis was negative. Conservative management post-biopsy resulted in marked improvement, with only occasional mild hoarseness reported. This case underscores the importance of thorough evaluation, interdisciplinary collaboration, and long-term monitoring in the diagnosis and management of laryngeal amyloidosis, offering hope for improved outcomes in the future.

KEYWORDS

Laryngeal amyloidosis, Amyloid deposition, Systemic involvement

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INTRODUCTION

Amyloidosis encompasses a rare and diverse group of diseases characterized by the abnormal extracellular deposition of amyloid proteins, which disrupt normal cellular and tissue structure, ultimately impairing their functions [1]. These amyloid deposits can manifest either as localized, confined to a single organ or site of the body, or systemic, affecting multiple organs and tissues. Localized amyloidosis constitutes approximately 15% of all amyloidosis cases [1, 2]. Due to its non-specific clinical and radiological presentation, diagnosing localized amyloidosis can be challenging, often requiring the exclusion of systemic involvement [2].

Within the realm of localized amyloidosis, the head and neck region stands out as a significant site of affliction, affecting up to 20% of cases, with the majority presenting as the localized form [2]. Remarkably, among head, neck, and respiratory tract amyloid depositions, laryngeal amyloidosis, though rare, claims the distinction of being the most frequent site of amyloid accumulation [2, 3].

The diagnosis of laryngeal amyloidosis hinges on histological examination, where the presence of pathognomonic Congo red stain positivity, displaying an apple-green birefringence under polarized light, serves as a crucial diagnostic marker [2].

CASE REPORT

A 50-year-old woman with no significant medical history, no smoking, no alcohol consumption, and no history of gastroesophageal reflux disease, presented to our ENT Head and Neck Surgery Department with an 8-month history of worsening hoarseness. There were no other complaints, no

stridor, dyspnea, dysphagia or painful swallowing. On examination, she appeared well with no sign of respiratory distress, or general condition deterioration.

Flexible laryngoscopy showed a left supraglottic submucosal thickening involving the left vestibular fold. True vocal cords were otherwise normal in appearance and mobility, with no signs of paresis or paralysis. Clinical examination of the neck showed no palpable lymph nodes.

Computed Tomography (CT) scan of the neck with contrast showed an irregular thickening on the anterior aspect of the left vocal cord, involving the anterior commissure, the paraglottic space, the left aryepiglottic fold and vallecula, with no signs of cartilage or adjacent soft tissue invasion (Figure 1). Direct laryngoscopy was performed under general anesthesia and enabled us to conduct multiples biopsies on the suspected area (Figure 2). Pathology report showed submucosal inflammation, and deposition of eosinophilic amorphous material. Congo Red staining results were consistent with amyloidosis (Figure 3).

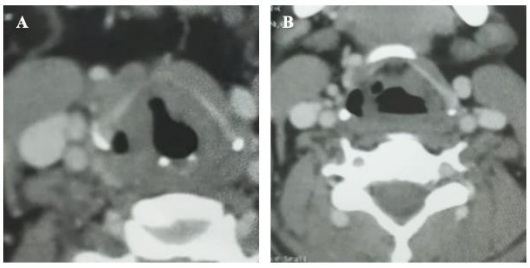

Figure 1: Axial CT scan of the neck showing an irregular thickening on the anterior aspect of the left vocal cord (A), involving the anterior commissure, the paraglottic space, the left aryepiglottic fold and vallecula (B), with no signs of cartilage or adjacent soft tissue invasion.

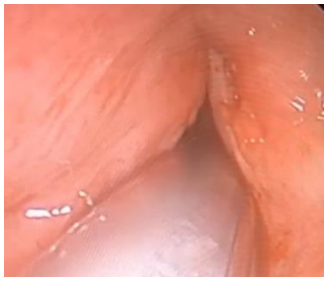

Figure 2: Direct laryngoscopy showing a left supraglottic submucosal thickening involving the left vestibular fold.

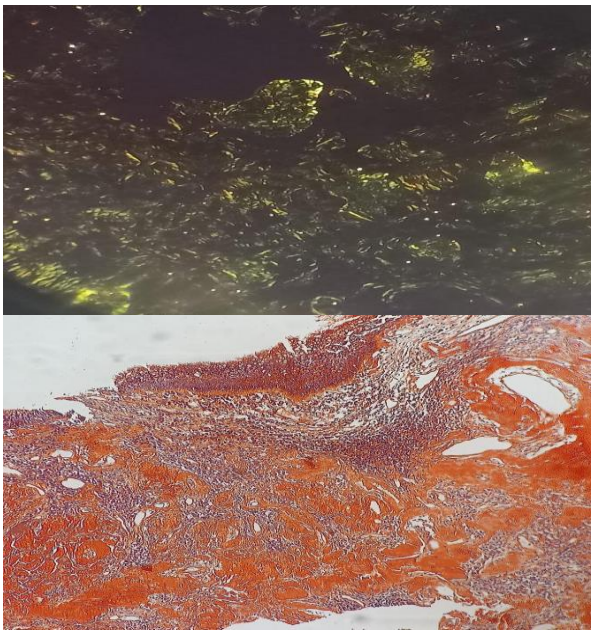

Figure 3: Eosinophilic amorphous deposits stained with congo red showing characteristic apple-green birefringence under polarized light.

After diagnosis, the patient was admitted to the internal medicine department for a comprehensive evaluation to rule out systemic amyloidosis. Extensive investigations were conducted, and fortunately, all findings from these tests were negative for any signs of systemic amyloidosis. Subsequently, the patient was managed conservatively, with marked improvement noted after the initial biopsy procedure. She reported only occasional mild hoarseness as a residual symptom. To ensure ongoing monitoring, the patient is under regular follow-up to watch for any recurrence of symptoms or potential systemic involvement. As of the 6-month follow-up period following the biopsy, there have been no signs of recurrence, indicating a positive response to conservative management.

DISCUSSION

Amyloidosis is characterized by the abnormal accumulation of insoluble fibrils outside of cells within various tissues and organs, and it can manifest as either a systemic or localized condition [1]. In the latter category, a generally favorable prognosis is observed in the majority of the cases [2]. Within the category of localized amyloidosis, the head and neck region is impacted in approximately 20 percent of cases, with documented occurrences in the orbit, nasopharynx, lips, floor of the mouth, tongue, larynx, and tracheobronchial tree [2]. This condition displays a higher incidence among females, particularly within the fifth and sixth decades of life [1, 2].

Among these sites, the larynx stands out as the most frequently affected, with the vestibular folds and ventricle being the primary sites of localization, followed by the vocal folds, aryepiglottic folds, and subglottis in decreasing order of frequency [2, 4, 5]. Notably, instances of simultaneous involvement of multiple sites within the larynx have been documented [2].

In comparison to amyloidosis in other organs, laryngeal amyloidosis tends to be diagnosed earlier due to the onset of symptoms at an earlier stage [1, 2]. Nevertheless, the slow growth of lesions often leads to a protracted course of the disease, making the diagnosis challenging and necessitating a high level of clinical suspicion [6].

The specific symptoms experienced by individuals are influenced by both the size and anatomical location of the amyloid deposits [2]. Among these symptoms, hoarseness and progressive dyspnea emerge as the most frequently reported, while dysphagia, cough, and even episodes of hemoptysis are also commonly encountered [2, 3].

The laryngoscopic findings varied notably across cases, encompassing a spectrum from nodules to soft, cystic, or diffuse swelling, and occasionally forming tissue masses [1, 2]. Vocal cord fixation, although infrequent, typically arises from other underlying predisposing factors [1]. During direct laryngoscopy, amyloid deposits have historically manifested as firm, unulcerated lesions in hues of yellow, red, or white [2, 5].

Imaging techniques are valuable for delineating the extent of lesions, often revealing a more comprehensive involvement than what is apparent during laryngoscopy [1]. Computed tomography (CT) typically reveals a uniform thickening of endolaryngeal soft tissues, which may exhibit spontaneous hyperdensity and only mild enhancement following intravenous contrast administration [1, 7]. Tracheal involvement often results in extensive longitudinal narrowing and may encompass the posterior tracheal membrane, a key distinguishing feature from relapsing polychondritis [7, 8].When examined via magnetic resonance imaging (MRI), these lesions commonly display intermediate T1-weighted signal intensity and low T2-weighted signal intensity [1, 2].

Nevertheless, the definitive diagnosis hinges on biopsy. Under light microscopy, amyloid presents as an unspecific, homogeneous, acellular, amorphous, eosinophilic appearance when stained with H & E. The hallmark diagnostic feature is observed through Congo red staining, which displays a distinctive apple-green birefringence when viewed under polarized light [1-3].

Following diagnosis, all patients should undergo an investigation to ascertain the presence of systemic disease due to the significantly greater morbidity and mortality associated with systemic involvement compared to localized disease [2].

The primary objective in managing localized laryngeal amyloidosis is to maintain a patent airway with minimal interventions, while definitive treatment often entails the surgical excision of the affected tissue [2]. Management approaches vary, ranging from close observation to procedures such as balloon dilatation, endoscopic excision, and in some cases, laryngectomy [2, 9]. While laser excision, including carbon dioxide and potassium titanyl phosphate lasers, has demonstrated effectiveness in numerous studies [10, 11], there is also evidence supporting the use of cold endoscopic excision for

small laryngeal lesions and localized glottic lesions [12]. In a pediatric case reported by Phillips et al. [2], a microdebrider and coblation wands were employed, marking the first instance of such an approach documented in the literature.

Laryngeal amyloidosis can exhibit a protracted course, with reports of recurrence occurring as late as 8 to 14 years after initial diagnosis, and some lesions remaining unchanged for up to 17 years [12]. Recurrence is a frequently documented, with one study indicating that nearly half of its cases required subsequent surgery due to either recurrent localized lesions or the development of large lesions [13]. While there is no consistent pattern regarding the most vulnerable site for recurrence, a limited case series suggests that supraglottic disease may have a higher incidence of recurrence, potentially attributed to surgical techniques [12]. As a result, annual follow-up for a minimum of 10 years is recommended [2, 3].

CONCLUSION

Laryngeal amyloidosis, though uncommon, presents a diagnostic puzzle and therapeutic conundrum. The diverse clinical and radiological manifestations necessitate vigilant investigation and a multidisciplinary approach to care. Management strategies, ranging from conservative observation to surgical excision, must be tailored to each patient's unique presentation. Recurrence underscores the need for long-term monitoring, while the specter of systemic involvement requires thorough evaluation post-diagnosis.

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