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Case Report

Primary Localized Laryngeal Amyloidosis: an Exceptional Clinical Entity

Sinha R[1], Singh RK[1]* and Kumar R[2]

[1]Department of Otorhinolaryngology, Indira Gandhi Institute of Medical Sciences University, India

[2]Department of Medicine, Katihar Medical College, India

***Corresponding author:** Rakesh Kumar Singh, Department of Otorhinolaryngology, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Zip code: 800014, India

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Abstract

Laryngeal amyloidosis is a rare condition encountered by otolaryngologists, though it is the most common site for amyloidosis in head and neck region. It often presents with dysphonia as it involves the vocal cords. This again makes its diagnosis and management challenging. Precise surgical excision is mandatory for functional preservation of voice along with close follow-up for quick detection of any recurrence. We present a case of laryngeal amyloidosis managed with microlaryngeal surgery resulting in complete resolution of symptoms and no recurrence.

Keywords: Amyloidosis; Larynx; Localized

Introduction

Amyloidosis is a rare benign disease of head and neck region. It is a disorder that results from the extracellular deposition of an abnormal fibrinous protein called amyloid in various organs of the body. It can be of two types, systemic and localized. Although the localized form of amyloidosis is extremely rare in larynx, it is the commonest site of involvement in head and neck region [1]. The condition involves the true vocal cords and laryngeal ventricle, which is primary in nature; nevertheless in some incidence, it may be a part of generalized systemic form [2]. The presenting complaints are hoarseness, fullness in throat and sometimes cough. Diagnosis is based on histopathological evidence of amyloid confirmed by Congo red staining under polarized light or by electron microscopy. Surgery is the mainstay of treatment directed at improving the airway and restoration of quality of voice. We present one such case managed with microlaryngeal surgery resulting in complete resolution of symptoms and no recurrence.

Case Presentation

A 45 years old female presented with progressive hoarseness of voice for past 6 months. There was no associated breathing difficulty, cough or sense of fullness in throat. There were no signs and symptoms of any systemic disease. She was a non-smoker with no other associated debilitating disease. Endolaryngeal examination revealed granular polypoidal mass in right ventricle. Both the vocal cords were mobile with no airway compromise. The remaining head and neck examination and systemic examination was unremarkable. Her routine blood investigations were within normal limits. The pulmonary, cardiac, abdominal and urinary evaluations were normal. Microlaryngeal surgery with excision biopsy under general anesthesia was planned. Microlaryngoscopy showed multiple smooth polypoidal tissue in entire right ventricle and extending through anterior commissure to partly involve the left ventricle also (Figure 1). The mass was completely excised with the help of microlaryngeal forceps and scissors without causing any damage to the vocal cords or vestibular folds. Histopathological examination by haematoxylin and eosin revealed polypoidal lesion lined by squamous to respiratory lining with an acellular amorphous eosinophilic infiltrate in stroma and mild chronic inflammatory infiltrate. Congo red stain was positive along with apple green birefringence with polarized light (Figure 2). The immediate post-operative period was uneventful and patient showed gradual improvement in voice quality. On one year follow up visit, the patient had reasonable speech with no hoarseness. The vocal cords were mobile with no edema or scarring. No residual or recurrent disease was seen.

Discussion

Amyloidosis is a group of diseases defined by extracellular deposition of amyloid fibrils with features such as 'twisted beta-pleated sheet' protein configuration, affinity for Congo red dye and resistance to proteolytic digestion. Initially the deposition of such protein was described by Rokitansky in 1842; however, in 1851, Virchow was the first to introduce the term amyloidosis for such proteinaceous deposition [1]. As being classified into systemic and localized type on the basis of involvement of organs, only 9% to 15% are of localized type [1].

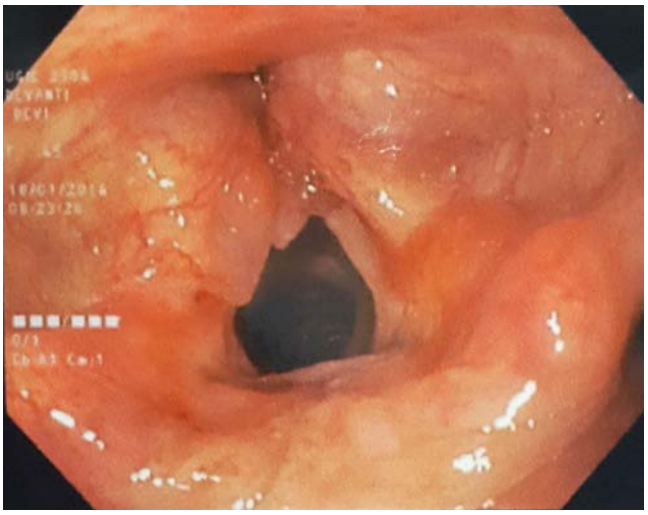

Figure 1: Microlaryngoscopic picture showing smooth polypoidal tissue in right ventricle and extending through anterior commissure to left ventricle.

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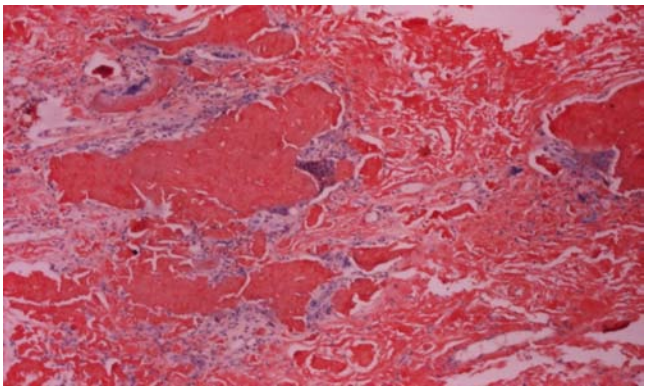

Figure 2: Congo red stain positive along with apple green birefringence with polarized light.

In 1873, Borow documented the first case of localized laryngeal amyloidosis and since then very few such cases have been described in English literature [3]. Nevertheless, the laryngeal involvement by amyloid is the commonest site in head and neck region that accounts for 2% of all benign laryngeal tumors. It occurs most commonly in the fifth to seventh decade of life, perhaps more common amongst males with 3:1 male to female predominance [2]. In a study conducted on 188 new patients with amyloidosis between 1990 and 2003 in Netherlands, only five patients had localized laryngeal amyloidosis [4]. Another study revealed eleven cases of laryngeal amyloidosis retrieved from the files of the Otorhinolaryngic–Head & Neck Tumor Registry, Washington D.C. from 1953 to 1990 [5].

The amyloidosis have been classified in different ways from time to time based upon immunocytochemical nature, pattern of extra cellular deposition, localization, clinical type and the presence or absence of underlying diseases. On the basis of multiple factor consideration, in 1980, Glenner classified amyloidosis in three groups as: (A) Amyloid protein type such as immunoglobulin light chain protein (AL) and secondary amyloidosis (AA); (B) the protein precursor such as κ or λ light chain, trans-thyretin, or β_2-microglobulin; and (C) primary, secondary, myeloma associated, familial, localized, aging and hemodialysis associated on the basis of clinical presentation. The primary and localized amyloidosis is the commonest form of laryngeal amyloidosis and is being grouped as AL/κ or λ/primary type of Glenner classification [1]. However the exact etiopathogenesis of primary amyloidosis is still not defined. Systemic amyloidosis, multiple myeloma, lymphoma and extramedullary plasmacytoma are rarely associated with laryngeal amyloidosis. Hence, the elimination of these diseases by urinary Bence-Jones protein, echocardiogram, electrocardiography, liver function test, renal function test, complete blood count and abdominal ultrasound is needed [1]. These all test were found to be normal in our case, which was a primary localized type of amyloidosis of larynx.

The presenting symptoms are highly influenced by site and size of the amyloid deposition. Laryngeal amyloid may deposit at different sub sites of endolaryngeal structures. Mittrani and Biller concluded the ventricle and false cord as the commonest involved site, while Finn and Farmer have been reported the true cord as the commonest site of deposition [2]. In our case the disease involved the laryngeal ventricle and true cords, which was presented with slowly progressive weak to hoarse voice. The true cord involvement commonly presents as change in voice of weak, high pitched, husky to hoarse nature, which is slowly progressive to stridor in due course of the disease. The involvement of other sites presents as progressive dysphonia and foreign body sensation in throat, dyspnea, cough, odynophagia, and seldom hemoptysis. Endolaryngeal examination shows firm, non-ulcerating, and orange-yellow to gray epithelial nodules [6]. But the diagnosis is based on histopathology with characteristic 'Apple-green birefringence' produced after Congo red stain and viewed under crossed polarized light [5]. These typical features of laryngeal amyloidosis were consistent with the findings in our patient.

Surgery is the gold standard treatment of primary localized laryngeal amyloidosis. It is best done by Endoscopic CO2 laser excision of the mass [2,3]. Pulsed laser, removal of gas to avoid overheating, a properly aligned beam and extreme care with subglottic use is necessary when using laser to avoid permanent voice changes and stenosis. Accuracy, less bleeding and less scarring leads to better preservation of vocal cord function, which is easier to achieve with the use of laser than conventional surgery [2]. But the choice should depend on the experience and expertise of the surgeon with these procedures. In this patient the mass was excised surgically using microlaryngeal forceps and scissors carefully without causing any inadvertent trauma to the vocal cords. There was no residual disease left behind. A case with similar surgical approach has been reported where patient was treated successfully with endoscopic curettage of the mass with no inadvertent ill effects [6]. Medical treatment of laryngeal amyloidosis including use of corticosteroids and radiotherapy has been shown to be ineffective [2,7]. The slowly progressive course of signs and symptoms of the disease necessitates the need for regular follow-up over a number of years to ensure that recurrence is not missed [8]. We followed this case for a period of one year with no signs and symptoms of any recurrence and the patient is in good state of health.

Conclusion

Laryngeal amyloidosis despite being a rare disease should be included in the differential diagnosis of laryngeal masses. Its diagnosis is based on characteristic 'Apple-green birefringence' after Congo red stain under crossed polarized light. The treatment is preferably surgical with regular follow-up to detect early recurrence.

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