CASE REPORT

Primary localised laryngeal amiloidosis

Ferhat Bozkus,[1] Turgay Ulas,[2] Ismail Iynen,[3] Ilyas Ozardali,[4] Imran San[5]

Abstract

Localised amyloidosis of the larynx is uncommon and poorly understood. The precise etiology and pathogenesis are not entirely clear. It usually presents with hoarseness, pain and/or difficulty in breathing. We present the case of a 42-year-old woman with primary localised laryngeal amyloidosis who presented with hoarseness and dyspnoea. Biopsy of the specimen revealed amyloid. There were no signs of any systemic disease in the patient and diagnosis was established histopathologically. She was treated surgically by microlaryngoscopy under general anaesthesia. At 6 months, the patient's voice and breathing had both improved substantially.

Keywords: Dyspnoea, Hoarseness, Amyloidosis, Larynx, Localised.

Introduction

Amyloidosis is a group of disorders in which an extracellular deposition of an abnormal amount of proteins occurs in a variety of organs. Localised deposition of amyloid protein is regarded to be the result of local synthesis rather than the deposition of light chains produced elsewhere in the human body.[1,2] Unlike with other sites in the head and neck region, amyloidosis of the larynx is unlikely to be related to a systemic illness. It is a rare condition and accounts for less than 1% of all benign laryngeal tumours. It usually occurs in the 40-60 years age range, with a male-to-female predominance of about 2:1.4.[3,4]

Patients typically present with hoarseness, although they may present with cough, globus, haemoptysis, stridor or dyspnoea. The diagnosis is made after looking for evidence of systemic amyloidosis and systemic diseases associated with secondary amyloidosis. Immunohistochemical stains, and Congo red staining viewed under polarised light microscopy, or electron microscopic findings of a laryngeal biopsy specimen can confirm the presence of amyloid in the larynx. If symptoms are present, in isolated laryngeal amyloidosis, treatment is primarily endoscopic surgical removal of masses that interfere with laryngeal or airway function. Unfortunately, there is a high likelihood of recurrence after resection, and long-term follow-up is required.[2-4]

In this report, we present the case of a 42-year-old woman with primary localised laryngeal amyloidosis who presented with hoarseness and dyspnoea. The purpose to present this case is to contribute to the literature and promote the detection of these overlooked diseases.

Case Report

A 42-year-old woman presented with complaints of hoarseness and dyspnoea during the preceding 6 months. She had no history of tobacco and alcohol consumption, and reported no loss of weight. Her medical history was unremarkable. Physical examination of the neck was normal. Indirect laryngoscopy and endoscopy with a flexible endoscope revealed a smooth, nodular formation localised in the left laringeal surface of the epiglottis. Magnetic resonance imaging (MRI) of the neck revealed diffuse thickness of vocal cords, especially in the left, and airway narrowing was seen at this level. Defined area revealed a significant contrast enhancement after intravenous contrast media administration (Figure-1). Chest X-ray examination and ultrasonographic scan of abdomen were normal. The mass was removed by microlaryngoscopy under general anaesthesia. The lesion was biopsied and the pathology report was Congo red stained positive for amyloid. Hematoxylin and Eosin stain of biopsy specimen revealed a cellular stromal proliferation of hyalinise homogeneous material, consistent with amyloid deposits. Also, amyloid demonstrating apple-green birefringence with polarised light

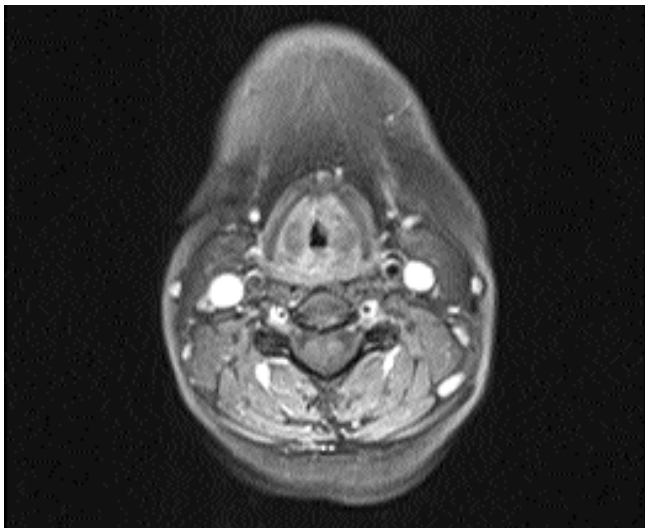

Figure-1: Magnetic resonance image of the neck revealing diffuse thickness of vocal cords, especially in the left. Airway narrowing is seen at this level.

[1,3,5]Department of Otolaryngology, Head and Neck Surgery, [2]Department of Internal Medicine, [4]Department of Pathology, Harran Unviersity, Sanliurfa, Turkey.
Correspondence: Ferhat Bozkus. Email: drferhat65@hotmail.com

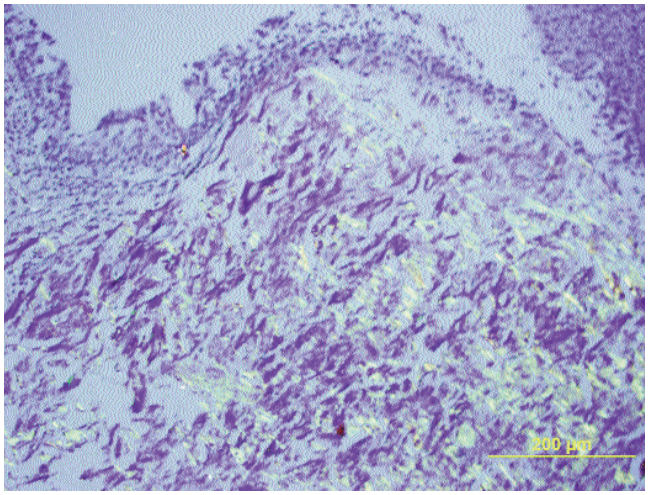

Figure-2: Congo red staining under polarised light revealed apple-green birefringence of amyloid (Congo-red&hematoxylin x200).

with Congo red (Figure-2). Further examination was performed for the exclusion of systemic amyloidosis. The patient's complete blood count (CBC), erythrocyte sedimentation rate (ESR), basic metabolic and biochemical panel and liver function tests were within normal limits. Laboratory investigation of serum protein electrophoresis was normal and did not reveal any monoclonal spike. Serum electrophoresis was normal. The histopathological results of duodenal, rectal and abdominal fat tissue biopsies were not compatible with amyloidosis. Based on these findings, systemic amyloidosis and multiple myeloma were excluded from the differential diagnosis. After negative work-up for systemic disease, the patient was diagnosed with primary laryngeal amyloidosis. For 6 months following the operation, the patient was asymptomatic and her voice was described as normal in quality and strength.

Discussion

Amyloidosis is a metabolic disorder of protein in which extracellular protein fibrils are deposited in various tissues. Amyloidosis is classified as systemic or local.[2] The exact etiology of localised amyloidosis of the larynx remains unclear. Two theories have been asserted to explain localised amyloidosis of the larynx. One blames a plasma cell reaction to inflammatory antigens and is supported by pathologic studies showing mixed polyclonal plasma cells interspersed with the amyloid tissue.[5] Another more likely scenario points to the body's inability to clear light chains produced by plasma cells located in the mucosal-associated lymphoid tissue.[3] Laryngeal amyloidosis is a type of localised amyloidosis that is characterized by monoclonal deposits of the light chain type.[6]

Clinical presentation varies depending on the site of involvement. Fibril accumulation causes progressive disruption of the structure and function of involved tissues and organs.[7] The ventricles and the false and true vocal cords are the most common sites for localised amyloidosis in the larynx.[1-3,7] Hoarseness is the main symptom, but dysphagia and cough may be the presenting symptoms.[7] In our case, localised amyloidosis was detected on laryngeal surface of the epiglottis and primary symptoms of our patient were hoarseness and dyspnoea.

Computed tomography or MRI may be helpful in showing the extent of the disease.[7] The most frequently used method to detect the amyloid protein is the histological staining of biopsy samples excised with Congo red stain.[2,4] Currently, the most popular and highly effective treatment available is microdirect laryngoscopy with a carbon dioxide laser excision. If symptoms are present, endoscopic surgical extirpation of any amyloid tumour with maintenance of functional anatomy is the goal of treatment.[1] In nearly half of the cases reported in literature, the surgery had to be repeated due to localized recurrent or large lesions. Therefore, regular follow-up with laryngoscopy is crucial for early diagnosis of recurrence.[1] In our case, we only performed microlaryngoscopic surgical excision and diagnosis was made by histopathological examination. Also, no further procedures were performed in the subsequent 6 months.

Conclusion

Laryngeal amyloidosis should be kept in mind while making the differential diagnosis of laryngeal masses, as was the case in our patient. Diagnosis is always made histopathologically. Screening of systemic amyloidosis are necessary in these cases. Correct recognition and long-term follow-up after diagnosis are essential and important.

References

1. Ma L, Bandarchi B, Sasaki C, Levine S, Choi Y. Primary localized laryngeal amyloidosis: report of 3 cases with long-term follow-up and review of the literature. Arch Pathol Lab Med 2005; 129: 215-8.
2. Yiotakis I, Georgolios A, Charalabopoulos A, Hatzipantelis P, Golias C, Charalabopoulos K, et al. Primary localized laryngeal amyloidosis presenting with hoarseness and dysphagia: a case report. J Med Case Reports 2009; 3: 9049.
3. Neuner GA, Badros AA, Meyer TK, Nanaji NM, Regine WF. Complete resolution of laryngeal amyloidosis with radiation treatment. Head Neck 2010; 34: 748-52.
4. Gallivan GJ, Gallivan HK. Laryngeal amyloidosis causing hoarseness and airway obstruction. J Voice 2010; 24: 235-9.
5. Benning S, Technau-Ihling K, Fisch P, Fradis M, Schipper J, Maier W. Amyloid tumor of the larynx associated with plasma cell infiltration: differential diagnosis. Ear Nose Throat J 2004; 83: 839-43.
6. Lewis JE, Olsen KD, Kurtin PJ, Kyle RA. Laryngeal amyloidosis: a clinicopathologic and immunohistochemical review. Otolaryngol Head Neck Surg 1992; 106: 372-7.
7. O?uz H, Safak MA, Demirci M, Arslan N. Familial primary localized laryngeal amyloidosis in two sisters. Kulak Burun Bogaz Ihtis Derg 2007; 17: 283-6.