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ORIGINAL ARTICLE

Localized Laryngeal Amyloidosis

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ABSTRACT

Amyloidosis in head and neck is rare but a benign disease. Among the sites in head and neck, larynx is the most commonly involved. A 40-year-old female had presented with hoarseness of voice and dyspnea with a mass in false vocal cord, which was biopsied and diagnosed as amyloidosis. After excluding systemic diseases, final diagnosis of localized laryngeal amyloidosis was given.

Keywords: Amyloidosis, Congo red, Laryngeal

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INTRODUCTION

Amyloidosis forms a group of disorders characterized by extracellular tissue accumulation of amorphous hyaline material. Amyloidosis is categorized into two main forms: Systemic and localized. The distinction between localized and systemic disease is important because localized amyloidosis can be managed conservatively with an excellent prognosis, whereas systemic amyloidosis is associated with significant morbidity and mortality.[1]

Localized forms often involve head and neck. The aerodigestive tract is a common location.[2] The majority of patients with head and neck amyloidosis have no underlying chronic systemic disease.[1]

Laryngeal involvement in amyloidosis is rare and accounts for less than 1% of all benign laryngeal tumors. It usually occurs in the 40 to 60 years age range with a male-to-female predominance of about 2:1.[1,3] In the larynx, the most commonly involved sites are the vestibular fold, followed by subglotis, ventricle, vocal folds, and aryepiglottic folds. Clinical manifestations are hoarseness, dyspnea, cough, stridor, and rarely, hemoptysis. The lesion is usually a firm; nonulcerated yellow, orange, or gray submucosal nocule; mass or pedunculated polypoid lesion.[1] The diagnosis is made by a high degree of suspicion based on the history and a characteristic appearance on direct laryngoscopic examination. When such lesions are seen, an adequate deep punch biopsy should be obtained.[4]

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CASE REPORT

A 40-year-old female had presented with hoarseness of voice and mild dyspnea. Laryngoscopic examination showed gray-white lesion in false vocal cord extending up to larynx and clinically represented vocal cord polyp. Punch biopsy was taken from the lesion.

The tissue was received in multiple bits in histopathology section. Sections from the soft tissue bits stained with hematoxylin and eosin showed fragments of tissue lined occasionally by pseudostratified columnar ciliated epithelium. One fragment showed an amorphous, extracellular eosinophilic, and homogeneous deposit which raised suspicion of amyloidosis (Fig. 1). Consequently, Congo red staining was done, which stained the deposit orange with apple-green birefringence on polarization (Fig. 2).

A provisional diagnosis of amyloidosis was given and further investigations to ascertain the cause of amyloidosis was asked for. All routine investigations were within normal range, and systemic diseases were ruled out. Follow-up of the patient also did not show manifestation of any systemic pathology. Consequently, diagnosis of localized laryngeal amyloidosis was given.

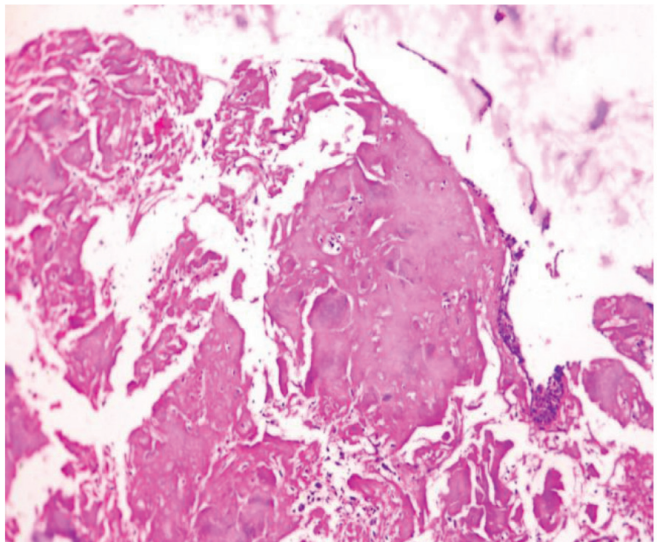

Fig. 1: Hematoxylin and Eosin section showing eosinophilic amorphous substance

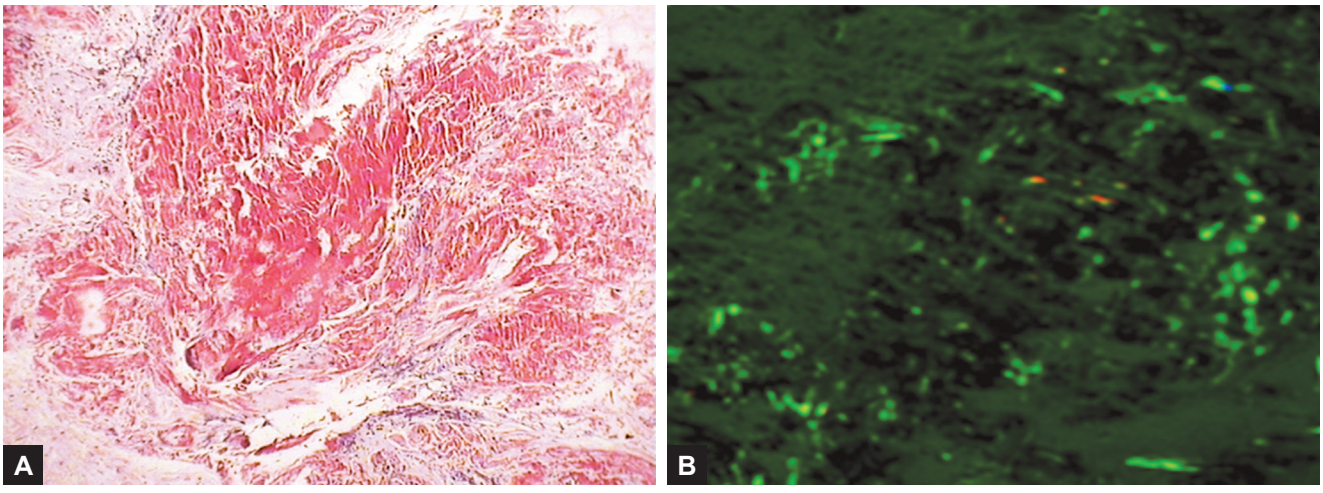

Fig. 2: Congo red staining showing orange areas, which showed apple-green birefringence under polarized microscopy

DISCUSSION

Amyloid consists of relatively insoluble fibrils of 8 to 12 nm, composed of polypeptide chains arranged in a twisted β-pleated sheet configuration. This particular protein configuration accounts for its characteristic staining properties and permits identification by light microscopy. The proteins found in the amyloid deposits are immunoglobulin light and heavy chains, secondary reactive serum amyloid A protein, leukocyte cell-derived chemotaxin-2, fibrinogen-a chain, transthyretin, apolipoprotein A-I and A-IV, gelsolin, and b-2 microglobulin. It imparts unique chemical properties like resistance to protease digestion and insolubility, which promote continued deposition within organs. While amyloid can be suspected on routine hematoxyin–eosin sections, special stains are important for definitive diagnosis. With the polarized microscope, amyloid is seen to have a green birefringence when stained with Congo red.[1]

There are currently three known forms of amyloidosis. The first, primary systemic amyloidosis, with no known underlying cause. It is different from the secondary systemic amyloidosis, which occurs with other underlying medical problems, such as tuberculosis, rheumatoid arthritis, and multiple myeloma. The third form of amyloidosis is localized and which occurs without any evidence of systemic or underlying disease. Localized amyloidosis is a clinical entity with variable presentation, depending on the organs involved.[1]

The localized amyloidosis in the head and neck is a rare and benign disease and may involve the orbit, paranasal sinuses, nasopharynx, oral cavity, salivary glands, and larynx.[5-7] Among the sites in head and neck region, larynx is affected most frequently (61%), followed by oropharynx (23%), trachea (9%), and orbit (4%). Only 3% of cases occur in nasopharynx.[1,8] Within the larynx, a number of sites can be involved, including ventricular folds (55%), laryngeal ventricle (36%), subglotic space (36%), vocal folds (27%), aryepiglottic folds (23%), and anterior commissure (14%).[1]

Laryngeal amyloidosis may present clinically in a variety of ways like hoarseness of voice, dyspnea, cough, stridor, and rarely hemoptysis. Hoarseness of voice for a long period and dyspnea are seen in our patient, as has also been observed by Taimur et al[9] and Chow et al.[10]

In the larynx, amyloid deposits are submucosal and homogeneous and are not associated with cartilage changes. The differential diagnosis includes other submucosal diseases, such as laryngeal sarcoidosis, lymphoma, and pseudotumor.[1]

The diagnosis of amyloidosis requires tissue biopsy, with Congo red and immunohistochemical staining (for AA or AL amyloid), showing an orange or red color under normal light and the classical apple-green birefringence under polarized light.

The computed tomography (CT) provides excellent information on the anatomic location and topography of different laryngeal benign lesions but cannot be used to differentiate inflammatory masses from benign neoplasms.[1]

Once localized amyloidosis has been established, it is important to diagnose or exclude systemic involvement. The natural history of systemic amyloidosis is to progress much more quickly. The present case did not show any signs of systemic involvement even on follow-up, based on which it was diagnosed as a case of localized laryngeal amyloidosis.

The localized amyloidosis has excellent prognosis and never evolves toward systemic forms. Hence, it may be treated with surgical excision only.

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