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Case Report

Primary Amyloidosis of Larynx – A Case Report

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Abstract

Amyloidosis is an idiopathic benign disorder in which extracellular proteinaceous material deposits in variety of organs leading to tissue damage and functional problem of that organ. Besides larynx being the commonest site, amyloidosis is found in other head –neck sites such as salivary glands, oral cavity, pharynx, nasopharynx, sinonasal cavity, trachea, bronchi, lungs & lymph node. This is a report of a 40 year old man admitted in the departement of Otolaryngology-Head and Neck Surgery, BSMMU, Dhaka, Bangladesh, in 2014, presented with history of hoarseness of voice and polypoidal swelling involving left vocal cord, ventricle and vestibular fold. Initially he was treated with antitubercular chemotherapy as direct laryngoscopic biopsy reported tuberculosis. As symptoms did not subside repeat direct endoscopic biopsy was done when histopathological examination confirmed the diagnosis of amyloidosis. MRI shows transglottic involvement of lesion which appears more extensive than endoscopic findings. Regular follow-up is necessary to assess accurately the disease progression, using conservative clinical management to preserve laryngeal function.

Key words: Amyloidosis, larynx, tuberculosis, CO2 laserIntroduction

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Amyloidosis is an idiopathic benign disorder in which extracellular proteinaceous material deposits in variety of organs leading to tissue damage and functional problem of that organ. Von Rotinasky first identified amyloid deposits in the liver and spleen in 1842[1]. In 1875 first case of laryngeal amyloidosis was reported by Burrow & Neuman.[2] Nearly 125 articles on laryngeal amyloidosis had been reported till 2002 most of which described only one to three cases of laryngeal amyloidosis[3]. Amyloid deposition may be localized to a single organ or systemic. Besides larynx being the commonest site, amyloidosis is found in other head – neck sites such as salivary glands, oral cavity, pharynx, nasopharynx, sinonasal cavity, trachea, bronchi, lungs & lymph node. Amyloidosis in head-neck region is commonly seen in absence of any systemic involvement or associated diseases[4].Due to rarity of the

disease clinical suspicion of other common conditions such as neoplasm, polyp appears first than amyloidosis. Because of more submucosal involvement deep punch biopsy is required to get representing tissue simple for histopathological confirmation of amyloidosis as well as exclusion of other conditions.

We report a case of primary amyloidosis of larynx which may be the first case reported from Bangladesh.

Case report

A 40 years old man presented with 10 month history of hoarseness of voice and admitted for treatment in the Department of Otolaryngology-Head & Neck Surgery, Bangabandhu Sheikh Mujib Medical University, Dhaka. He is neither a smoker nor a drinker. He had a history of direct laryngoscopic examination and biopsy reported as tuberculosis of larynx for which he had been treated with anti-Tubercular chemotherapy for 6 months without relief of symptoms. He had no history of weight loss or cough. Laryngoscopic examination revealed a polypoidal swelling involving the anterior one third of vestibular fold, ventricle and vocal cord. Biopsy was taken and histopthological examination of the biopsied tissue with special staining (Congo red) confirms the diagnosis of amyloidosis. Both CT scan and MRI were done. MRI shows transglottic involvement of lesion which appears more extensive than endoscopic findings. Further investigations were done to rule out systemic amyloidosis. Complete blood count, liver function tests, renal function tests, urinary test for Bence-Jones protein, chest radiography, ECG, echocardiography & Montaux test reported normal finding.

The patient was kept on regular follow-up without any surgical intervention.

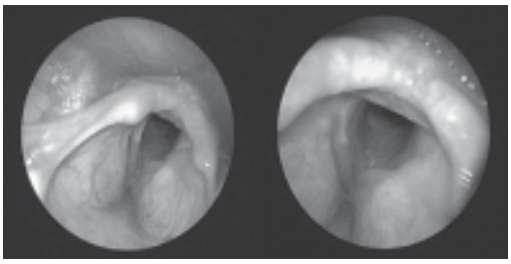

Fig.-1: *Endoscopic picture of Larynx showing polypoidal swelling involving left side.*

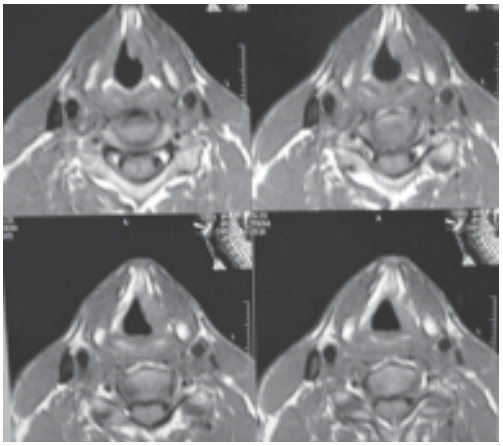

Fig.-2: *MRI of larynx showing isointense shadow involving left side of larynx.*

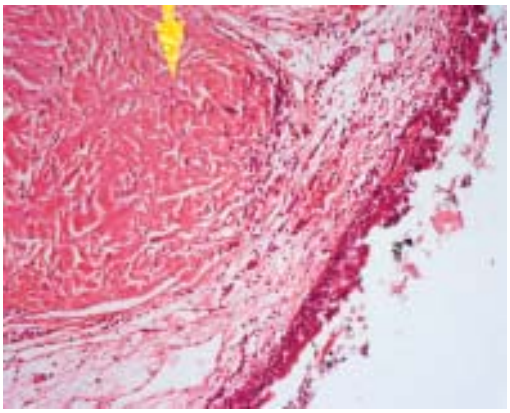

Fig.-3:*Congo red stain of laryngeal biopsy.Amyloid deposit (arrow) is seen as congo red positive material below the epithelium(Congo red x400).*

Discussion

Deposition of amyloidosis in the larynx is rare, accounting for o.2 to 1.5% benign tumors of larynx. Unlike other benign conditions it has no definite established link with smoking, vocal abuse or recurrent infection[5, 6]. Progressive change of voice is the commonest & earlier presenting complaints in almost all reported cases. But patients may rarely present with dysphagia, dyspnea & stridor. Laryngeal amyloidosis usually presents fourth to sixth decade of life with male predominance[6].

In this case a 40 year old male patient presented with progressive change of voice for 10 months without any associated symptoms. Within the larynx amyloidosis may affect all sub sites of the larynx but supraglottic part at the level of false cord is the most frequent involved site[1].There is a shorter duration of symptoms for patients who have amyloidosis of the false cord than for patients who have masses in the true vocal cord alone or for patients who have transglottic lesions[7].

This case has transglottic presentation.

Laryngeal amyloidosis can not be diagnosed by visual inspection alone, because the clinical appearance of amyloid mimics the appearance of other lesions e.g. benign vocal cord polyp, laryngocele[8]. Histopthological examination of the biopsied tissue with special staining (Congo red) confirms the diagnosis of amyloidosis[5,6,7,9]. Congo red staining demonstrates the classic appearance of bright green birefringence under polarizing light microscope. Methyl violet and thioflavine-T stain may also be used to diagnose amyloidosis, revealing a metachromatic pink-violet in the presence of amyloid fibrils[7, 10].

In this case diagnosis was confirmed with Congo red staining technique.

As amyloidosis is mostly a submucosal disease Computed Tomograpy (CT) or Magnetic resonance imaging (MRI) may be helpful in mapping lesions, which may be more extensive than they appear during laryngoscopic examination. MRI is the preferred technique to detect the most specific features, since amyloid deposits show an intermediate T1-weighted signal intensity and low T2 –weighted signal intensity, and MRI is thus considered to be more specific technique than CT scanning[11].

Both CT scan and MRI were done in our case. MRI shows transglottic involvement of lesion which appears more extensive than endoscopic findings.

Although laryngeal amyloidosis usually presents as an isolated localized lesion a number of cases have been demonstrated a relationship between extramedullary plasmacytoma or multiple myeloma and localized laryngeal amyloidosis [12, 13]. Therefore most authors suggest systemic evaluation of the disease because of high morbidity.

Complete blood count ,liver function tests, renal function tests, urinary test for Bence-Jones protein chest radiography, ECG, Echocardiography, biopsies may be warranted. Areas to be biopsied may include the rectum, abdominal fat, lip, kidney, liver, spleen, bone marrow and skin[7].

Most cases of suspected localized amyloidosis may not necessary to investigate for systemic disease with biopsies of the lip, rectum, or abdominal fat, because of the low yield [14].

In this case all relevant investigations are done to exclude systemic involvement of disease except biopsy.

Treatment of laryngeal amyloidosis can be one of observation or surgery. Adjuvant

therapies such as irradiation, chemotherapy, and steroids have no proven benefit in the treatment of this disorder. Surgical options range from microdirect laryngoscopic laser or cold-knife excision to external partial laryngeal resection. Currently the most popular and highly effective treatment available is microdirect laryngoscopy with carbon dioxide laser excision [5, 6, 9].

Laryngeal amyloidosis seems to be an indolent process, with patients living a long time with evidence of disease. Therefore it is important to have sustained and regular follow-up of these patients to assess accurately the disease progression, using conservative clinical management to preserve laryngeal function for as long as possible [7].

In this reported case the patient is on regular follow-up without any surgical intervention except biopsy.

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