LETTER TO THE EDITOR

An Unusual Case of Hoarseness of Voice and Dysphagia in Multiple Myeloma with Amyloidosis

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Dear Editor,

Hoarseness of voice is commonly caused by acute and chronic laryngitis, functional vocal disturbances, benign and malignant tumors, and neurogenic disturbances. Dysphagia occurs most commonly as a result of obstruction of the esophageal lumen or non-obstructive causes like neuromuscular and infiltrative disease. Rarely hoarseness of voice and dysphagia are manifestations of systemic diseases. Here we report a patient who presented with hoarseness of voice and dysphagia and was finally diagnosed to be multiple myeloma with secondary amyloidosis (AL).

A 52-years-old male presented with hoarseness of voice along with dysphagia to solids and liquids for 10 months. He also noticed thickening of his tongue subsequently for the last 7–8 months. No history of any fever, weight loss, or any constitutional symptom. Clinical examination showed macroglossia (Fig. 1) with multiple aphthous ulcerations otherwise rest was general and systemic examination was normal.

Hematological and biochemical investigations were normal. Laryngoscopy revealed ulcerative growth involving the posterior 1/3rd of the right vocal cord and extending into the arytenoid. Biopsy from the anterior part of the tongue and buccal mucosa showed stratified squamous epithelium and mild acanthosis with no evidence of dysplasia or malignancy. The UGI endoscopy revealed leucoplakia in the esophagus with a mosaic pattern stomach. To rule out underlying malignancy he underwent a CECT chest/abdomen and PET CT scan of the whole body and which revealed multiple lytic lesions in the skeleton with FDG avid bone marrow deposits in the left humerus along with periarticular (muscular) lesions around both hip joints (SUV max 5.7) and non FDG avid right vocal cord growth (Fig. 2).

Further workup was done for multiple myeloma. Urine for Bence Jones protein was negative. M spike was not seen in serum protein electrophoresis and immune fixation. Serum immunoglobulin levels were normal. The β2 microglobulin was 2480 ng/mL (normal range 609–2366 ng/mL). Free light chain (LC) assay revealed a kappa/lambda ratio >300. Bone marrow biopsy revealed marrow infiltration of more than 30% by plasma blasts, atypical and matured plasma cells. A typical plasma cells are CD38 positive and showed kappa restriction on immuno-histochemistry. Apple green birefringence was noted under polarized light with a Congo red stain. Endoscopic esophageal biopsy and tongue biopsy were studied using special stains and it revealed AL which showed apple-green birefringence under polarized light (Fig. 3). The 2D echo revealed no features of cardiac AL.

The patient was diagnosed as a case of multiple myeloma with light chain AL. He is being managed with bortezomib, cyclophosphamide, dexamethasone, and zoledronic acid. The patient is responding well to treatment.

When a single clone of plasma cells produces monoclonal immunoglobulin which later turns into neoplastic proliferation causes multiple myeloma. One-fifth of multiple myeloma produces only light chains. Amyloidosis is the deposition of fibrillar aggregates of monoclonal immunoglobulin light chain in various organs like the liver, heart, kidney, spleen, and others. Amyloidosis is classified

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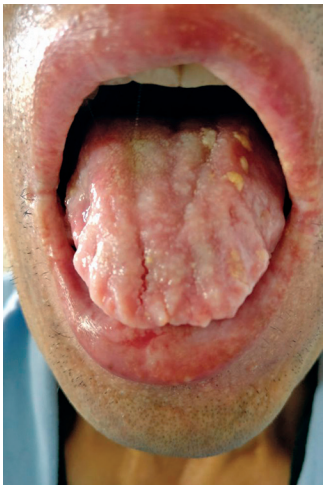

Fig. 1: Tongue showing amyloid deposit in form of macroglossia and leathery texture

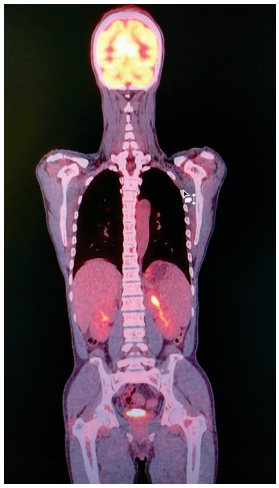

Fig. 2: PET scan showing multiple skeletal lytic lesions

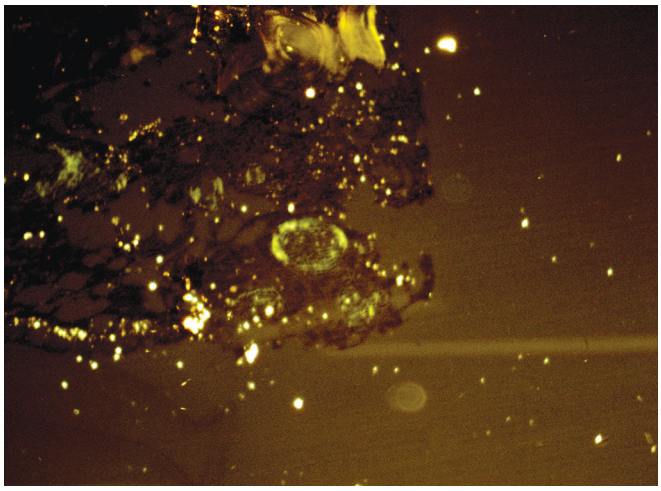

Fig. 3: Endoscopic esophageal biopsy showing apple-green birefringence under polarized light using Congo red stain

into primary AL, secondary AL, hereditary AL, and localized AL. Secondary AL is a serious complication of primary diseases such as multiple myeloma, rheumatic arthritis, Hodgkin's lymphoma, tuberculosis, syphilis, rickets, and others. Light chain AL is the most common form of systemic AL and it complicates about 10% of cases of myeloma, with the most being asymptomatic.[1] Presentation of multiple myeloma is varied and non-specific however bone pain and fatigue are the most common presentations.[2] Amyloidosis of the oral cavity is uncommon however tongue is the most frequent site.[3] Macroglossia is a very rarely reported presenting complaint in multiple myeloma.[4] As compared to other sites in the head and neck region, AL of the larynx is rare.[5] Esophageal involvement in AL ranges from 13% in a radiology study to 22% in an autopsy series.[6] Our patient is a rare case of multiple myeloma with light chain AL who had involvement of the tongue, larynx, and esophagus which are not very common sites for the deposition of amyloids.

Primary or secondary amyloidosis involving the larynx, oral cavity, and esophagus is rare but patients may present with manifestations due to these rare sites like dysphagia, hoarseness of voice, macroglossia, and others. The emphasis should be given to high levels of clinical suspicion and clinically directed lab evaluation to diagnose such kind of cases.

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