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Case Report

Partial Glossectomy for Treatment of Macroglossia Secondary to Myeloma-Associated Amyloidosis: A Case Report

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Abstract

Around 10-15% of patients with multiple myeloma develop amyloidosis due to the extracellular deposition of light-chain amyloid molecules. When this deposition occurs in the tongue, patients may exhibit macroglossia. The sequelae of this macroglossia can include dysphagia and gagging, tooth displacement, and airway compromise. This article presents the case of a 43-year-old African American female who was treated with a partial glossectomy secondary to amyloidosis related multiple myeloma.

Introduction

Macroglossia is the abnormal enlargement of the tongue with significant disproportionate appearance to other structures in the mouth. Macroglossia may be categorized as either primary (i.e., muscular enlargement) or secondary. Secondary macroglossia is related to, or associated with, neoplastic processes, metabolic or endocrine disorders, or syndromic conditions [1-4]. Examples of underlying disease processes causing secondary macroglossia are vascular malformations (such as hemangioma or lymphangioma) and myxedema due to hypothyroidism [1,3]. Macroglossia can also occur secondary to amyloidosis [5]. The intent of this article is to report on the case of a 43-year-old female who presented with enlarged, firm, and scalloped tongue secondary to amyloidosis related to multiple myeloma.

Case Description

A 43-year-old African American female presented to the Augusta University Medical Center due to dysphagia and gagging. She had been diagnosed with multiple myeloma one month earlier at an outside hospital. At that time, she noted weight loss of 40 pounds over the previous four months. Computed tomography (CT) scan showed lytic lesions of the spine, ribs, and sternum. Myeloma workup was performed, and patient was diagnosed with Revised International Staging System (R-ISS) stage 2 multiple myeloma. She was discharged to home in stable condition. The patient's treatment plan was to receive Velcade, Revlimid, and dexamethasone (VRD) therapy, which she started shortly thereafter. Upon presentation at the Augusta University Medical Center, the Otolaryngology service was consulted regarding her complaint of dysphagia and gagging, which she had been experiencing in the months leading up to her diagnosis of multiple myeloma. Otolaryngology performed a transnasal flexible laryngoscopy that showed no structural abnormalities of the base of tongue or supraglottic structures.

Gastroenterology was consulted and performed an esophagogastroduodenoscopy (EGD), which only showed moderate to severe antral gastritis. Additionally, a percutaneous endoscopic gastrostomy (PEG) was performed during that procedure to ensure the patient received proper nutritional intake, as her ability to tolerate food by mouth was impaired secondary to the dysphagia and gagging. The patient also had a modified barium swallow study, and no aspiration or penetration was observed. The only potential etiology of the dysphagia and gagging noted was an enlarged tongue. The oral and maxillofacial surgery (OMS) service was

consulted to determine if surgical management of the macroglossia was appropriate. Intraoral examination showed macroglossia with scalloping in the anterior and lateral borders of the tongue (Figure 1). Amyloidosis of the tongue secondary to multiple myeloma was suspected. The patient had previously had bone marrow aspirate and abdominal fat pad biopsy that were both negative for amyloidosis. OMS performed a partial glossectomy via keyhole technique with submission of excised tissue in 10% buffered formalin for pathological examination, which confirmed amyloidosis. Following the procedure, the patient remained intubated in the surgical intensive care unit (SICU). She was extubated on post-operative day #2 without the need for tracheostomy.

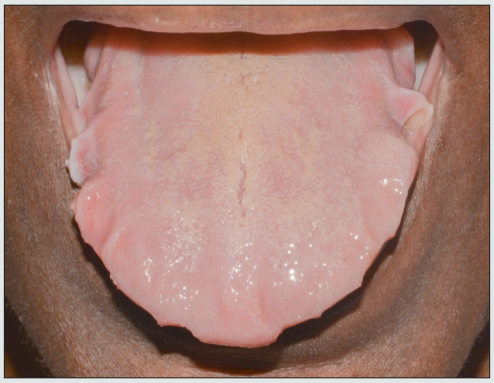

Figure 1: Macroglossia with scalloping of anterior and lateral borders of tongu

The patient had post-operative edema of the tongue, and thus still had dysphagia after surgery; however, this resolved over the next few months, and her dysphagia had significantly improved compared to her pre-operative state. Revlimid was discontinued prior to surgery to mitigate risks of complications with healing. In place of VRD therapy, she was switched to cyclophosphamide, bortezomib and dexamethasone (CyBorD) therapy shortly after surgery. A cardiac MRI was subsequently performed and did not show signs of cardiac involvement of amyloidosis. The patient's PEG tube was removed five months after partial glossectomy after she demonstrated the ability to swallow solid foods by mouth. One year after surgery, she stated that her swallowing had significantly improved prior to the partial glossectomy. She did note some alterations in taste and intermittent lingual paresthesia, but these were not major complaints. Her oncologist noted very good response to CyBorD therapy, and the patient underwent an autologous peripheral blood stem cell transplant 13 months after her partial glossectomy.

Pathologic Findings

Microscopic examination of formalin-fixed paraffin-embedded 5µ thick sections revealed tongue mucosa covered by parakeratinized squamous mucosa which supports subepithelial diffuse amorphous eosinophilic deposits. The amorphous deposits were markedly eosinophilic, largely acellular and afibrillar. The material deposition is noted in the superficial and deep lamina propria and in perivascular and intramuscular areas (Figure 2A). Sections stained with Congo red showed positive reddish deposits with the stain confirming the diagnosis of amyloid deposition (Figure 2B). The Congo red-stained deposits demonstrated characteristic apple-green birefringence when viewed under polarized light.

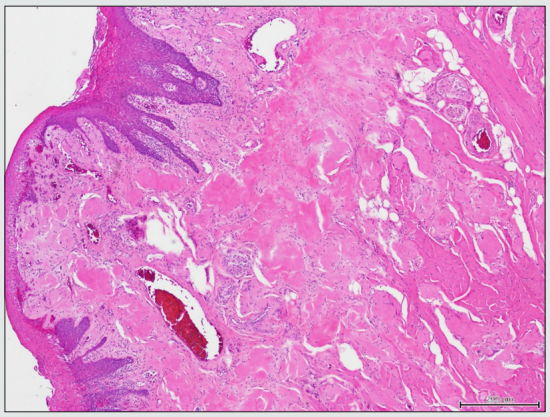

Figure 2A: Photomicrograph of tongue biopsy showing subepithelial stromal exctracellular amorphous eosinophilic amyloid deposition (hematoxylin and eosin-stained; original magnification X100).

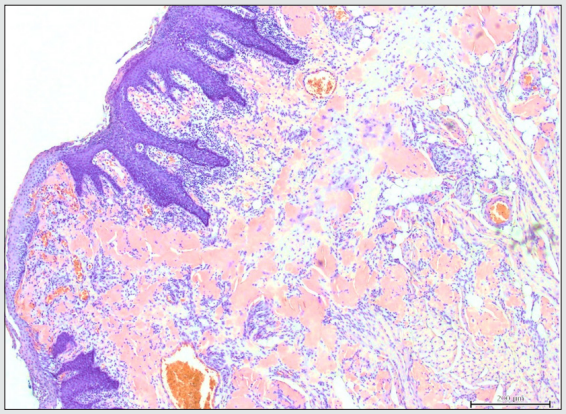

Figure 2B: Photomicrograph of Cong red-stained sections showing red amyloid deposits in the extracellular, perivascular and intramuscular zones (Congo red-stained; original magnification X100).

Discussion

Macroglossia can arise from many different etiologies, including amyloidosis. Amyloidosis is a group of diseases that share the feature of the deposition of an extracellular proteinaceous substance referred to as amyloid [6]. Approximately 39% of patients with amyloidosis exhibit oral manifestations, with the tongue being the most common site [7,8]. There are multiple classification systems for amyloidosis. Broadly, amyloidosis can be categorized as either localized or systemic [8]. The systemic type can be classified at least into primary, secondary, and hereditary, with some authors suggesting additional categories, such as for cases associated with multiple myeloma [8]. Other authors group amyloidosis associated with multiple myeloma into the primary or secondary categories [9-11]. Multiple myeloma is a monoclonal, multicentric neoplastic proliferation of bone marrow-based B lymphocytes which differentiate into plasma cells [12]. Myeloma cells produce various proteins, most frequently M protein in the form of IgG (60%), followed by IgA (20%-25%); only rarely are IgM, IgD, or IgE M protein observed. In few other cases, myeloma cells produce only κ or λ light chains. These light chains are small and can be excreted in urine where they are referred to as Bence-Jones proteins.

Males, African Americans, and adults between 60 and 70 years of age are most affected by this condition [9]. The malignancy results in an uncontrolled proliferation of atypical and nonfunctional immunoglobulins [13]. The solitary plasmacytoma is also a monoclonal neoplastic proliferation of plasma cells, but unlike multiple myeloma, only a single mass is present. Plasmacytomas can arise either in bone or in soft tissues. Most patients with plasmacytoma of bone will ultimately develop multiple myeloma when observed on a long-term basis [9,14]. Clinically, patients with multiple myeloma present with bone pain, anemia, thrombocytopenia, neutropenia, metastatic calcifications, renal failure, and Bence-Jones proteins in urine [15]. These patients are more susceptible to infections due to the reduced number of functional immunoglobulins and leukopenia. If there is maxillofacial involvement, osteolytic "punched out" radiolucent lesions may be seen on radiographs affecting the jaw bones. Tumor-like yellowish nodules may be found in the oral mucosa. Salivary glands may also be affected [16]. In about 10-15% of patients with multiple myeloma, the accumulation of the abnormal light chain proteins results in amyloidosis in the oral mucosa, and may present as macroglossia [9,16]. The reported prevalence of amyloidosis-associated macroglossia secondary to multiple myeloma varies, but it has been suggested to be up to 40% [9,17]. Macroglossia from this condition can become severe enough to splay teeth, affect speech and swallowing, and cause airway obstruction, sometime necessitating a tracheostomy [11]. Medical history, clinical findings, biopsy, and histological exam of amyloid lesions are all key components of determining the underlying cause of macroglossia and best treatment plan. While this condition leads to both functional and cosmetic problems for patients, there are no formal guidelines for the treatment of macroglossia secondary to amyloidosis [18,19]. In this case, a partial glossectomy was performed and tissue was submitted to confirm the macroglossia was a complication of amyloidosis associated with multiple myeloma. The patient had improvement of her symptoms starting around 5 months after surgery, and the improvement was sustained through one year after surgery.

This case, as well as others presented in the literature, document successful surgical intervention for macroglossia associated with amyloidosis and its complications [18]. However, it must be noted that macroglossia is likely to recur if the systemic disease process leading to amyloid deposition is not medically controlled. Thus, it is necessary to judge each case based on multiple criteria, including the severity of symptoms (airway compromise, nutritional compromise) and likely of recurrence (i.e., whether medical management can suppress further deposition of amyloid). The major shortcoming of this report is the lack of volumetric data prior to, and after, surgery. This would have given an objective measure of the amount of reduction achieved through the partial glossectomy, as well as the volumetric stability of the tongue over time. Nonetheless, the patient's improvement in

swallowing and the ability to remove her PEG tube are certainly markers of successful treatment. In conclusion, although there is no consensus in the literature regarding the surgical management of macroglossia due to myeloma-associated amyloidosis, the authors of this paper suggest, as have others, that partial glossectomy for the treatment of symptomatic macroglossia caused by myeloma-associated amyloidosis is a reasonable treatment option [20].

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