Mucous membrane plasmacytosis (MMP) of the upper aerodigestive tract is a rare, benign disorder of unknown etiology in which the mucous membranes are infiltrated by plasma cells. Only 22 cases have been reported in the English-language literature. The nomenclature has evolved over time and includes the names plasma cell mucositis, plasma cell orificial mucositis, and idiopathic plasmacytosis. There are reported cases of MMP involving the lips, gingiva, buccal mucosa, tongue, palate, nasal aperture, epiglottis, subglottic and glottic larynx. The classic clinical presentation of MMP is an intensely erythematous mucosa with surface changes described variously as verrucous, cobblestone, papillomatous, or velvety. Common symptoms are oral pain, dysphagia, persistent hoarseness, shortness of breath, and sore throat. Histologically, the majority of lesions are described as having an acanthotic epidermis and a dense subepithelial cellular infiltrate composed of mature plasma cells. Immunoperoxidase staining shows a mixed population of kappa and lambda light chains, demonstrating the benign nature of the condition.

A 49 year old male, non-smoker, presented with a two month history of persistent hoarseness and worsening shortness of breath. He did not have dysphagia of odynophagia. In office flexible laryngoscopy revealed bilateral edema and erythema of the false vocal cords and true vocal cords. The laryngeal surface of the epiglottis was also abnormal with an erythematous and verrucous appearance. Microlaryngoscopy under anesthesia was performed and biopsies of the concerning areas were obtained.

Histologically all biopsies including those of the true vocal cords, false vocal cords and laryngeal surface of the epiglottis showed hyperplastic squamous mucosa with severe chronic inflammation composed mainly of plasma cells, GMS, PAS-D, and Fite’s stains were negative for fungal organisms and acid fast bacilli. Serum protein electrophoresis and free light chain electrophoresis tests were normal (polyclonal). ACE and C-ANCA levels were also within normal limits and syphils testing was negative.

The diagnosis of mucous membrane plasmacytosis was given. Several systemic injections of decadron provided 2-3 weeks of symptomatic improvement each. In an attempt to avoid additional systemic steroids, inhaled flunisolide was prescribed to the patient but gave no relief. The patient then underwent intralesional injection of Depo-Medrol. This provided the most substantial improvement of symptoms which lasted for 3 months. To date, the patient has undergone intralesional injection three times, each time having symptomatic recurrence after 2-3 months.

Mucous membrane plasmacytosis is an idiopathic disorder that can mimic other similar conditions of the upper aerodigestive tract. Diagnosis is made on the basis of exclusion using histology, serology and cultures to narrow the differential diagnosis. Management of MMP is mainly targeted at symptomatic relief. Several treatment modalities have been tried, but none stand out as consistently effective.

Corticosteroids (topical, intralesional, and systemic) have been used most frequently with inconsistent results. Several cases have shown no benefit to topical and intralesional steroids while some have demonstrated marked regression or total eradication of the disease. In our patient, systemic and intralesional steroids provided temporary symptomatic relief while inhaled steroids were not beneficial. Khan et al. reported complete resolution of palatal MMP with inhaled beclamethasone spray. Antibiotics and oral nystatin have not been shown to be an effective treatment for MMP. Tamaki et al., however, described two patients with plasma cell cheilitis who responded to oral griseofulvin with complete remission of lesions. Fogarty reported one case with laryngeal involvement in which systemic chemotherapy provided temporary symptomatic relief. The patient eventually was treated with low dose radiation which caused symptomatic improvement maintained at the 12 month follow up.

Several authors report of resorting to debulking procedures, either surgical excision, electrocoagulation, CO₂ laser, or cryotherapy to reduce the mass effect of MMP involving the pharynx and larynx. These procedures are effective but have a high rate of recurrence. Long-term prognosis for patients with MMP is good. Occasionally, tracheostomy is required for lesions causing airway obstruction. No cases to date have shown progression of MMP to a malignancy of any type.

REFERENCES