A 62-year-old woman presented with a history of dry mouth, recurrent oral ulceration, periodic swelling to the upper right lip, dysphagia, and changes over the last 2 months to her voice. She sang in a choir and had problems vocalizing both while singing and day-to-day voicing. She reported that she had lost some range when singing, and her voice was occasionally fatigable. Her inability to sing had greatly affected her quality of life. She had no skin, genital, gastrointestinal, or ocular complaints and did not smoke or drink alcohol. Her medical history included allergic rhinitis, occasional migraines, and gastroesophageal reflux disease (for which she took omeprazole 20 mg once a day).

Clinical examination revealed a diffuse erythematous and edematous lesion on the soft palate, along with two ulcers in the left buccal sulcus (Figure 1). Various preliminary working diagnoses were given, including a lymphoproliferative disorder, sarcoidosis, granulomatosis with polyangiitis (Wegener granulomatosis), deep mycoses, and syphilis.

Initial investigations comprised the following blood tests: Full blood count, hematinsics, renal and liver profiles, C-reactive protein, immunoglobulins, serum and urine electrophoresis, antinuclear antibodies, antineutrophil cytoplasmatic antibodies, serum angiotensin-converting enzyme, complement, hemoglobin A1c, syphilis serology, HIV, and a celiac screen. Incisional biopsy and microbiology swabs were also performed; the biopsy showed inflammation with evidence of a dense plasma cell–rich infiltrate. Polyclonality of the infiltrate was confirmed with κ and λ free light chain in situ hybridization. A flexible nasendoscopy showed a spongiform appearance along the pharyngeal wall with significant edema; however, the vocal cords themselves appeared unaffected. A diagnosis of plasma cell mucositis (PCM) was made.

Initial management was with systemic prednisolone 30 mg. There was an improvement in symptoms and clinical appearance following the addition of dapsone 50 mg/100 mg on alternative days (Figure 2). Recurrent flare-ups of symptoms resulted in prolonged systemic steroid use so mycophenolate mofetil 1 g twice a day was used in addition to the dapsone. She has been relatively symptom-free for the last year and remains under regular review.
Discussion

Plasma cell mucositis is a benign polyclonal plasma cell proliferative disorder of the mucous membranes with an unknown etiology. The clinical appearances of PCM are varied, but typically it presents as a florid erythematous oral mucosa with cobblestone, nodular, papillomatous, granular, or velvety surface changes. There is a slight male predominance of 1:2.1, and the average age of patients is 56.6 years. Plasma cell mucositis is often associated with a synchronous or metachronous autoimmune or immunological dysfunction such as seronegative rheumatoid arthritis, Sjögren syndrome, autoimmune hepatitis, polymyositis, and diabetes mellitus. There is one case report of invasive squamous cell carcinoma developing from existing PCM of the lip.

Plasma cell mucositis can affect the oral cavity, as well as the nasal mucosa, nasopharynx, larynx, oropharynx, hypopharynx, and esophagus. Patients can present with symptoms of oral pain, dysphonia, chronic cough, persistent hoarseness, dyspnea, stridor, pharyngitis, and dysphagia. Complications of PCM that require surgical intervention include tracheal strictures causing airway obstruction symptoms, along with debulking procedures of glottic and pharyngeal tissue causing dysphonia or dysphagia due to a mass effect.

Clinicians must have a high index of suspicion for multiple myeloma and other plasma cell malignancies when considering a diagnosis of PCM. Evidence of monoclonal proliferation can be identified using serum protein electrophoresis, immunofixation, and serum-free light chains, which together have a sensitivity of 97% to 98%. The principle histological features of PCM include epithelial hyperplasia and spongiosis, along with a dense polyclonal plasmacytic infiltrate in the superficial lamina propria. Failure to perform the above investigations can delay diagnosis of a plasma cell malignancy and have a significant impact on the clinical course of the disease and patients’ quality of life.

Management of PCM is challenging, and there is no consensus on treatment. The use of topical and systemic steroids is beneficial, but adverse side effects limit prolonged use. Immunosuppressive agents have been used with varying success, including methotrexate, tacrolimus, dapsone, mycophenolate mofetil, cyclosporine, colchicine, azathioprine infliximab, golimumab, and adalimumab. Caution should be exercised when using potent immunosuppressive agents for this benign condition particularly where risk factors for malignancy exist.

Improvement in symptoms has been documented with low-dose radiotherapy; however, the follow-up duration was short at 12 months. Surgical interventions are required in cases of subglottic stricture or airway compromise, with two reported cases requiring tracheostomy.

It is important that PCM is recognized by head and neck practitioners, as its diagnosis is dependent on clinical and histopathological correlation.

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