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Subglottic plasma cell mucositis: a case study highlighting challenge in management

Plasma cell mucositis (PCM) is a rare inflammatory condition of polyclonal plasma cell infiltrate in mucosa. Diagnosis and management are challenging given its variable nature, yet defining appropriate treatment is paramount to improve quality of life. Upper airway PCM can result in life-threatening strictures requiring surgical intervention or tracheostomy. We report a case of isolated subglottic PCM in which symptoms were controlled without need for surgery.

A 40-year-old woman was referred to a multidisciplinary airway clinic for opinion regarding suspected reactive airway disease refractory to lifestyle and medical management. Informed consent was obtained to share deidentified images and test results. The patient reported frequent episodes of heartburn and indigestion, particularly after consumption of tomatoes and caffeinated drinks, and her Belafsky reflux symptom index (RSI) score was elevated at 23.[1] She also reported 10 years of cough with occasional yellow/green sputum and fluctuating side-to-side nasal congestion with sneezing particularly during spring. Family history was significant for her father with sarcoidosis. Comprehensive clinical examination demonstrated mild nasal septum deviation, rhinitic nasal mucosa, interarytenoid pachydermia suggestive of laryngopharyngeal reflux and grade 1 subglottic stenosis (Fig. 1). Serum allergy testing (total immunoglobulin E 11 IU/mL; minimal specific immunoglobulin E reactivity to dust mite, staple food, grass, animal and mould), serum angiotensin converting enzyme (ACE) and spirometry (pre-bronchodilator forced expiratory volume over forced vital capacity (FEV_1/FVC) of 84% and post-bronchodilator FEV_1/FVC of 83%) were unremarkable. Bronchial provocation testing and computed tomography sinuses were normal. Computed tomography of the chest and upper airways confirmed a short segmental (5 mm) subglottic stenosis of 40–50% (upper end of Cotton–Meyer grade I)[2] with pseudodiverticulae and mild circumferential mural thickening (Fig. 2), with no features of mediastinal lymph nodal or pulmonary sarcoidosis.

The patient underwent microlaryngoscopy and biopsy of the stenotic region with histopathology showing mucosal ulceration and plasma cell infiltration (Fig. 3), with no evidence of dysplasia or malignancy. After exclusion of multiple myeloma and Immunoglobulin G (IgG)-related disease on blood testing and film, histology results were discussed at a head and neck multidisciplinary team meeting and the diagnosis of PCM was made. She was commenced on 50 mg oral prednisolone for 10 days, inhaled fluticasone 250μg two puffs twice daily and low-dose esomeprazole.

On follow-up at 3 months, she had excellent symptom control and improved subglottic stenosis, allowing us to reduce the fluticasone to

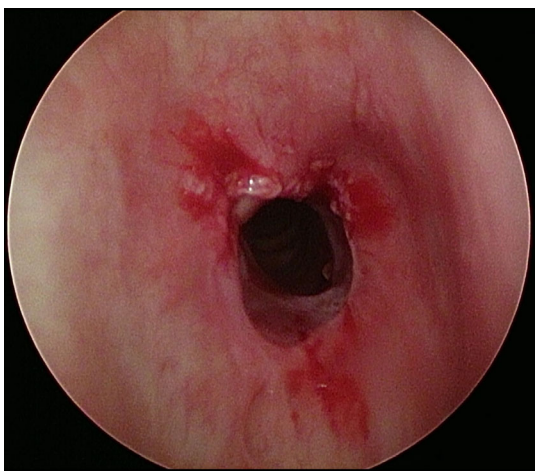

Fig. 1. Pre-treatment photograph showing grade one subglottic stenosis.

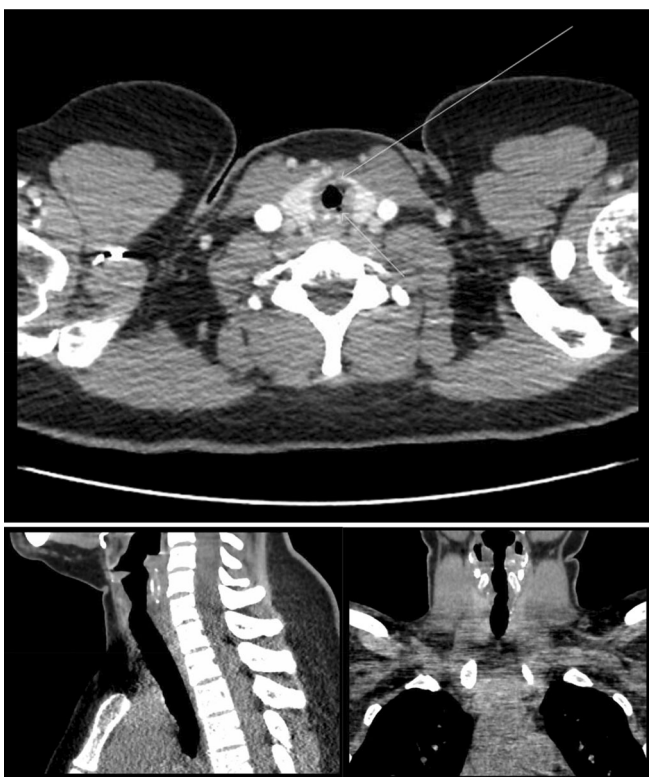

Fig. 2. Computed tomography of the upper airway demonstrating focal segmental narrowing in subglottis with pseudodiverticulae (arrows).

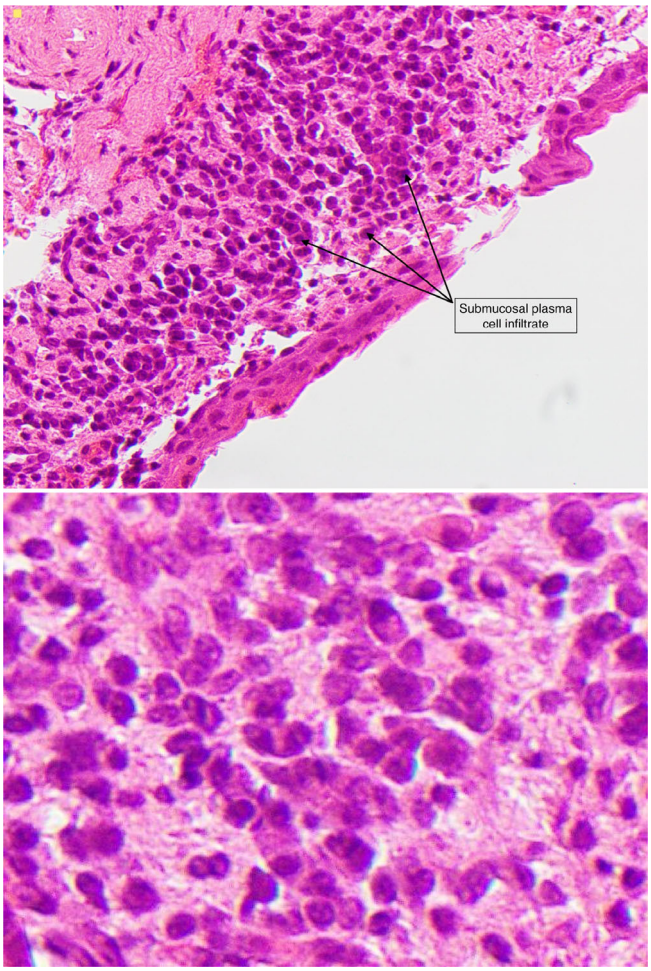

Fig. 3. Biopsy specimens obtained from the subglottis demonstrating mucosal ulceration and underlying dense infiltration of the submucosa with plasma cells (labelled).

one puff at night (Fig. 4). The patient is scheduled for regular surveillance with a view to halving the dose of fluticasone in 6 months.

PCM is a chronic inflammatory condition of polyclonal plasma cell infiltrate in mucosa. Case reports of PCM are few, with roughly 70 in the literature, yet true incidence is unknown given the difficulty of diagnosis. Hypotheses on the aetiology of PCM include hypersensitivity to substances, autoimmune processes[3] or a sequela of periodontitis;[4] ultimately, the cause remains unclear.

PCM has been identified at sites including gingival mucosa,[4] the upper aerodigestive tract[5–7] and distal airway.[8] Most aerodigestive cases occur in the oral cavity.[3] Isolated cases of the upper airway are rare, with few reports of supraglottic PCM[9,10] and subglottic PCM.[10,11] Histologically, dense sheets of subepithelial plasmacytosis with psoriasiform changes of the epithelium characterize PCM. Macroscopically, it produces lesions with an erythematous, oedematous, 'cobblestone' like appearance. This presentation mimics amyloidosis, sarcoidosis and squamous cell carcinoma, hence early biopsy is paramount.

PCM manifests as pain, sensitivity and swelling of the lesions. Aerodigestive signs and symptoms include dysphagia, dysphonia,

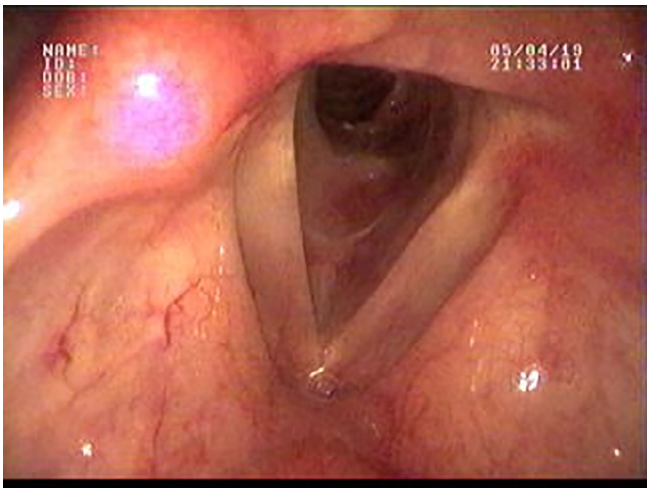

Fig. 4. Post-treatment nasendoscopy of the subglottis at 3 months showing improved subglottic stenosis.

vocal hoarseness, cough, dyspnoea or stridor. In the upper airway, PCM can lead to strictures and obstruction. In the absence of clear pathophysiology, no consensus treatment exists. Immunosuppression with topical steroids,[6] systemic steroids,[3] inhaled corticosteroids and systemic mycophenolate mofetil therapy[9] has been trialled with variable degrees of symptom improvement. Radiotherapy has been used successfully[5] and surgical excision,[7] balloon dilatation[12] or even temporary tracheostomy may be required in severe, intractable airway compromise.[5] Each modality has drawbacks, such as chronic exposure to corticosteroids and inherent surgical risk; however, these must be weighed against the benefit of symptom resolution. A good prognosis is expected upon successful symptom control and prevention of inflammation progression. We demonstrate successful symptom resolution in the medium term with minimally invasive measures.

PCM is rare and challenging to diagnose and manage given its variable nature. We outline successful diagnosis and treatment of subglottic PCM and hope our findings contribute to the body of knowledge on this condition.

Acknowledgements

The authors thank Bryan Knight (pathologist) for histology, Arunima Gupta (radiologist) for imaging, Dr Leon Kitipornchai (ENT fellow) for input on investigation and management and Professor Andrew Jones (Respiratory physician) for input with respiratory assessment.

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doi: 10.1111/ans.15987