Laryngeal mucous membrane pemphigoid: transnasal laryngoscopy to highlight a severe case

Carolina Tania Maria Watters, Benjamin Miller, Yakubu Karagama

DESCRIPTION
A 45-year-old man with a background of mucous membrane pemphigoid (MMP) was referred to ENT (Ear, Nose and Throat) clinic with a 7-month history of progressive sore throat and dysphonia. He had been under the care of Dermatology and Ophthalmology for management of his disease which was diagnosed 7 years ago on skin biopsy and despite courses of corticosteroids, dapsone and mycophenolate mofetil his disease remained active with ocular, oral and skin manifestations. His past medical history included sickle cell anaemia and tuberculosis for which he had been treated.

Oral examination revealed extensive bullous disease of both tonsils and posterior pharyngeal wall. He had a Voice Handicap Index (VHI) score of 32 and a Grade, Roughness, Breathiness, Asthenia, Strain (GRBAS) score of 33.33. Fibre-optic nasendoscopy demonstrated dry nasal mucosa with extensive crusting of the false vocal cords but no airway narrowing or stenosis. Following optimisation of his systemic therapy with a course of rituximab, he was reviewed in a specialist laryngology clinic with persistent dysphonia. The blistering of his oropharynx had resolved, however transnasal videolaryngoscopy demonstrated extensive active disease in the nasal cavity, nasopharynx and larynx, with persistent crusting and inflammation of the false cords, more pronounced on the left side (figure 1 and video 1). Administration of topical anaesthesia facilitated detailed inspection of the true vocal cords and subglottis which were also affected by crusting and ulceration down to the third tracheal ring, with no associated stenosis. A contrast-enhanced CT scan of the neck was performed, which demonstrated a left-sided fluid-filled laryngocele. In addition to his existing systemic therapy, he has been commenced on a short course of oral antibiotics and regular Naseptin cream, saline douches and saline and steroid nebulisers. He continues to receive multidisciplinary follow-up for active disease, although most recent video-laryngoscopy has confirmed involution of the laryngocele.

Learning points

- Mucous membrane pemphigoid is a rare autoimmune disease which affects the larynx in 5% to 15% of cases.
- Scar formation can lead to laryngeal strictures, stenosis and ultimately airway obstruction.
- Laryngoceles are a rare but observed sequelae of the disease.
- A multidisciplinary approach is key to ensuring early diagnosis, timely individualised treatment and minimising the need for surgical intervention.

Mucous membrane pemphigoid is a rare autoimmune disease causing blistering and ulcerative lesions of mucous membranes. It most commonly affects the oral and ocular membranes however can also affect the skin, nasopharynx, larynx, oesophagus and anogenital regions.1,2 Laryngeal manifestations of MMP occur in only 5% to 15% of cases, and on a population level are extremely rare with a prevalence of 1 in 10 million.3 Untreated laryngeal MMP can result in airway stenosis and obstruction, with a small proportion of reported cases requiring laryngeal surgical procedures or tracheostomy.4 Patients also have a relatively higher possibility of developing a cancer.5 Laryngoceles represent an extremely rare sequela of laryngeal MMP, with only one case previously reported in the published literature.6 Biopsy is essential for immunohistochemical diagnosis and targeted therapy. The larynx is considered an area of ‘high risk’ involvement requiring prompt use of systemic corticosteroids, immunosuppressive and biological therapies.6,7 Due to immunosuppression, there is high risk of...
additional bacterial and fungal infection sometimes necessitating antibiotic and antifungal cover. Early diagnosis and a multidisciplinary approach are paramount in order to achieve early remission and minimise the need for surgical intervention.

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REFERENCES