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

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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, Aesthesic, Strain (GRBAS) score of 33.333. 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 anaesthetics 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.

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.

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.

Figure 1. (A) Ulceration, bleeding and crusting lesions of the subglottis and trachea. (B) Muco-purulent and blood-stained ulceration and crusting of the left nasal cavity.

Video 1. Video demonstrating left supraglottic bulge, followed by a short segment highlighting severity of ulcerative lesions in the glottis and subglottis.
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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