Push It To The Limit

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Granulomatosis with polyangiitis (GPA) is a potentially life threatening immune-mediated disease with protean manifestations which may follow a relapsing and remitting course. It is important that clinicians recognize the various ways in which the disease may present.
A 32-year-old man presented to the rheumatology clinic with voice hoarseness, exertional dyspnea, dry cough, nasal crusting, and sinus congestion all of which had been present for one month. He was an established patient to our clinic but had been lost to follow-up since 2019. He was previously diagnosed with GPA in 2006 when he was 15. Initial exam upon return to our clinic was significant for nasal crusting, erythematous nasal mucosa and appreciable voice hoarseness. CT imaging disclosed stenotic changes of his larynx and scattered nodular opacities throughout the lungs. Subsequent laryngoscopy revealed marked subglottic inflammation with stenosis and polypoid lesions. He was treated with a combination of local vocal cord corticosteroid injections, systemic corticosteroids, rituximab and avacopan; his condition stabilized and he did not progress to respiratory compromise. He was briskly tapered from systemic corticosteroids with continued stabilization and subsequent improvement of his condition. Of note, his disease course as a teenager, including development of subglottic stenosis, was remarkably similar.
This case both highlights the importance of recognizing an uncommon but potentially life threatening manifestation of GPA, subglottic stenosis, as well as the need to remain vigilant for disease relapses.