

Granulomatosis with polyangiitis in supraglottic larynx, a case report

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Introduction

Granulomatosis with polyangiitis formerly known as Wegener granulomatosis is serious uncommon multisystem autoimmune disease of unknown etiology described by Friedrich Wegener in 1936. It is characterized by a triad of granulomatous lesions of the upper and lower respiratory tract, focal segment glomerulonephritis and disseminated necrotizing vasculitis. The severe untreated Granulomatosis with polyangiitis in the airway is associated with very high mortality rate.

Case report

A 21-year old man consulted for a 1-year history of shortness of breath on exertion and on lying down, choking to solids and fluids, and 8 kilograms of weight loss. Examination revealed fixed left vocal fold, with bilaterally ventricular smooth surface mass mimicked true vocal folds (Figure 1). The histological diagnosis was hyperkeratosis with inflammation. A CT scan of the larynx demonstrated a bilateral supraglottic mass, which was excised with CO2 laser, and the histopathological diagnosis was active chronic inflammation, with no evidence of atypia or malignancy.

Examinations for rheumatoid factor, ANA and VDRL/RPR were negative, as was a PPD test, and the chest X-ray was normal. A month later the patient presented with a nasal obstruction and discharge, with small ulcers all over the oral cavity, and a firm 2*3cm mass was seen on the soft palate. Signs of recurrence in the supraglottic larynx seen (Figure 1). A third CO2 laser microlaryngoscopy was performed, the airway was opened, and multiple biopsies were taken. A tracheostomy was avoided. c-ANCA, and p-ANCA were negative. The histological diagnosis was necrotizing granuloma no vasculitis was observed. The case was shared with a rheumatologist and the patient was diagnosed as having granulomatosis polyangiitis, and a systemic steroid and immunosuppressive therapy were started.¹⁻⁸

Discussion

Friedrich Wegener described Granulomatosis with polyangiitis in 1936. It's characterized by triad of granulomatous lesion of the upper and lower respiratory tract, focal segment glomerulonephritis and disseminated necrotizing vasculitis. Although Granulomatosis with polyangiitis may occur at any age, the mean age of occurrence is 40 to 55 years old. The M/F ratio is equal.

The four diagnostic criteria defined by the American College of Rheumatology (ACR) for WG in 1990 are as follows:

- a. Oral or nasal ulcers or purulent bloody flux
- b. Abnormal lung X-ray revealing nodules and cavities

- c. Abnormal urinary sediment
- d. Granulomatous inflammation in the extra vascular region at biopsy



Figure 1 Signs of recurrence in the supraglottic larynx seen.

The presence of two or more of these criteria has a sensitivity of 88% and a specificity of 99%.

The European Vasculitis Study Group recommends grading disease severity of antineutrophil cytoplasmic antibody (ANCA) – associated vasculitis (AAV) into the following 5 categories:

1. Localized - Upper and/or lower respiratory tract disease without any other systemic involvement or constitutional symptoms.
2. Early systemic - Any, without organ-threatening or life-threatening disease.
3. Generalized - Renal or other organ-threatening disease, serum creatinine level less than 5.6 mg/dL.

4. Severe-Renal or other vital-organ failure, serum creatinine level exceeding 5.6 mg/dL.
5. Refractory - Progressive disease unresponsive to glucocorticoids and cyclophosphamide.

Although over 90% of patient with Granulomatosis with polyangiitis will have positive ENT finding at some stage in their disease, Subglottic stenosis occurs in 10-20%, where supraglottic involvement is extremely rare. To our knowledge, Belloso A has reported one patient in 2008, and our patient is the second to be reported. Vocal fold paralysis can occur as a result of arytenoid cartilage fixation, as described by Eckel HE in 2003, which explains our patient's occurrence of vocal paralysis.

The differential diagnosis should include carcinoma, tuberculosis, syphilis, sarcoidosis, amyloidosis and blastomycosis. The definitive diagnosis requires a histopathological examination. The antineutrophil cytoplasmic antibody (specifically PR3-ANCA) is positive in 85% of active cases, which is reduced to 30-40% in remission. In contrast, in cases of localized granulomatosis with polyangiitis such as the present case the utility of ANCA is markedly decreased relative to generalized cases. Multiple studies have investigated the usefulness of c-ANCA in the diagnosis of localized granulomatosis with polyangiitis Wolfgang L Gross reported only 50% of those patients were ANCA positive. In our patient ANCA analysis was negative. A biopsy might demonstrate the full pathognomonic triad of granulomatous inflammation, vasculitis, and necrosis in 15% of cases. This report was approved by the local ethics committee of King Fahad Hospital.⁹⁻¹⁵

Conclusion

We found unpredictable and atypical supraglottic granulomatosis with polyangiitis. Repeated biopsy was required for the final diagnosis, and surgical intervention to maintain a patent airway is the main course of treatment. Early detection of granulomatosis with polyangiitis is important to induce remission and improve the survival rate.

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Conflicts of interest

Author declares there are no conflicts of interest.

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