

A life-threatening manifestation of granulomatosis with polyangiitis: Subglottic stenosis

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A 37-year-old female patient presented with symptoms of eye swelling, redness, protrusion, and shortness of breath. During examination, she had biphasic stridor, no edema in the nasal mucosa, and no saddle nose deformity. Elevated acute phase reactants and normocytic anemia were observed. Urine microscopy findings were normal, proteinuria was not detected, and immunological tests showed positive p-ANCA (perinuclear antineutrophil cytoplasmic antibody). Thorax computed tomography (CT) revealed stenosis below the cricoid cartilage and millimetrically small lymph nodes at this level, while no pathology was observed in the lung parenchyma. The biopsy results indicate the presence of inflammatory infiltration and edema in the submucosal layer, where a significant number of lymphocytes and neutrophils were observed. However, there was no evidence of granulomatous reaction. The patient was diagnosed with limited granulomatosis with polyangiitis (GPA), formerly called Wegener's granulomatosis, based on clinical and laboratory findings and was treated with methylprednisolone and azathioprine. When dyspnea complaints

increased, rituximab was started. The control neck CT revealed progression of stenosis in the subglottic region and increased concentric wall thickness, with the narrowest air column measured at 6 mm (Figure 1). During rigid bronchoscopy, the patient experienced respiratory arrest and required an emergency

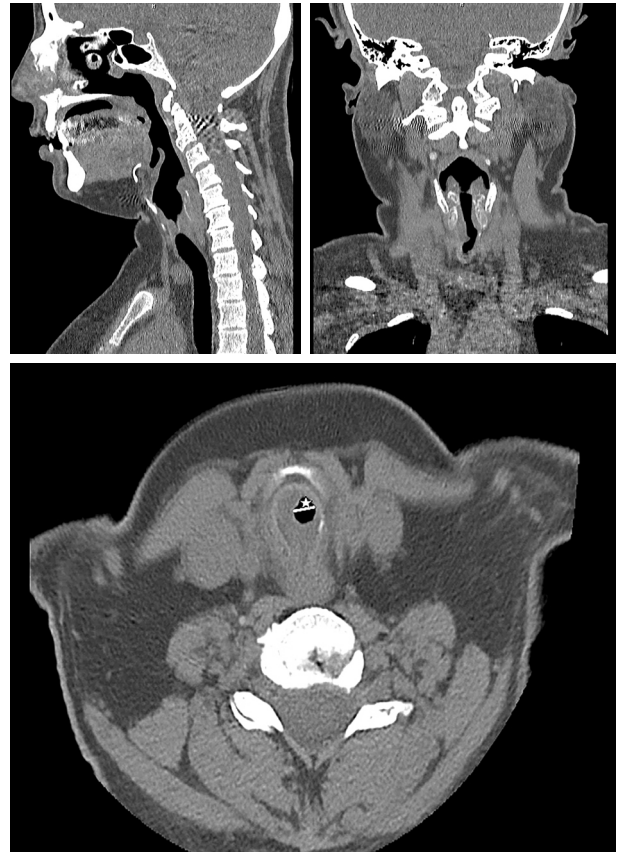


Figure 1. On computed tomography, the tracheal air column is markedly narrowed below the level of the vocal cords with asymmetric wall thickening.

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tracheostomy. A written informed consent was obtained from the patient.

Approximately 10 to 15% of patients with GPA develop subglottic stenosis (SGS), which should be considered in young patients with sinonasal involvement.¹⁻³ Patients often report hoarseness, difficulty breathing during physical activity, and increasing stridor. In the subglottic region, fragile mucosa, ulcers, or granulation tissue may cause narrowing. Biopsies taken from patients with disease limited to the ear, nose, and throat show nonspecific inflammation rather than a granulomatous reaction.⁴ GPA-SGS is generally independent of the disease activity. Interventional procedures such as steroid injection, endoscopic balloon dilatation, carbon dioxide laser, stent, and resection-anastomosis and immunosuppressive agents such as steroid, methotrexate, azathioprine, mycophenolate mofetil, cyclophosphamide, and rituximab are among the treatment options.³ Approximately 40% of patients with GPA-SGS require tracheotomy as part of disease treatment.⁵ It is necessary to diagnose and treat SGS at an early stage in patients with GPA as it is associated with a high risk of morbidity.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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