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Granulomatosis with polyangiitis involving the epiglottis

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Abstract

Granulomatosis with polyangiitis (GPA) frequently involves the upper respiratory tracts, but involvement of the epiglottis is extremely rare. This report describes a patient initially presenting with dysphagia and increasing stridor due to epiglottitis. Bronchoscopy showed swelling of the epiglottis with partly whitish nodular lesions, with biopsy specimens showing neutrophil infiltration and necrosis. Chest computed tomography showed multiple nodular consolidations in the bilateral lung parenchyma, and histological findings were consistent with vasculitis. The patient was diagnosed with GPA and responded well to treatment with prednisolone and cyclophosphamide. Although an uncommon manifestation, GPA should be included in the differential diagnosis of epiglottitis, especially in patients with lung parenchymal lesions suggestive of GPA.

Introduction

Granulomatosis with polyangiitis (GPA) frequently involves the upper respiratory tract, including the nose, ear, and larynx, but involvement of the epiglottis is extremely rare [1]. This report describes a patient initially presenting with dysphagia and increasing stridor due to epiglottitis associated with GPA.

Case Report

A 45-year-old man was referred to our hospital because of increasing stridor and dysphagia of 4 weeks. He denied haemoptysis, paraesthesia, and numbness. Physical examination showed a body temperature of 36.6°C, but there were no crackles on auscultation. There were no cutaneous eruptions and peripheral neuropathy was not observed. Chest computed tomography (CT) showed swelling of the epiglottis (Fig. 1C) and bilateral multiple nodular consolidations in the lung parenchyma (Fig. 2C, D). Laboratory findings revealed a white blood cell count of 8500/μL (85.4% neutrophils, 10.2% lymphocytes, 0.1% eosinophils, and 3.9% monocytes) and elevation of C-reactive protein (CRP) at 12.1 mg/dL. The serum creatinine level was normal at 0.5 mg/dL, but routine urinalysis showed microscopic haematulia. He was negative for proteinase 3 (PR3)-anti-neutrophil cytoplasmic autoantibodies (ANCA), myeloperoxidase (MPO)-ANCA, and other autoantibodies. Bronchoscopy showed swelling of the epiglottis with partly whitish nodular lesions (Fig. 1A). Biopsy specimens of these lesions showed neutrophil infiltration and necrosis, but there was no evidence of vasculitis or granuloma (Fig. 2A). We did not perform tissue culture, but special stains for bacteria, acid-fast, and fungal organisms were negative. There were no abnormal findings in the larynx and subglottic trachea. Because 3-day treatment with sulbactam/ampicillin did not improve his conditions, and swelling of the epiglottis partially blocked the airway, he was started on steroid therapy prior to obtaining biopsy samples of the lungs. Histologically, the right lower lobe showed neutrophilic vasculitis (Fig. 2B), but this patient was negative for bacteria, fungi, and acid-fast bacilli. Microscopic haematuria continued to be observed, but his serum creatinine concentration was within the normal range. The patient was diagnosed with GPA and started on treatment with prednisolone and cyclophosphamide. Dysphagia and stridor improved gradually. Follow-up CT and bronchoscopy showed marked improvement (Fig. 1B, D). The dose of prednisone was tapered to 5 mg/day and he has remained stable for over a year.

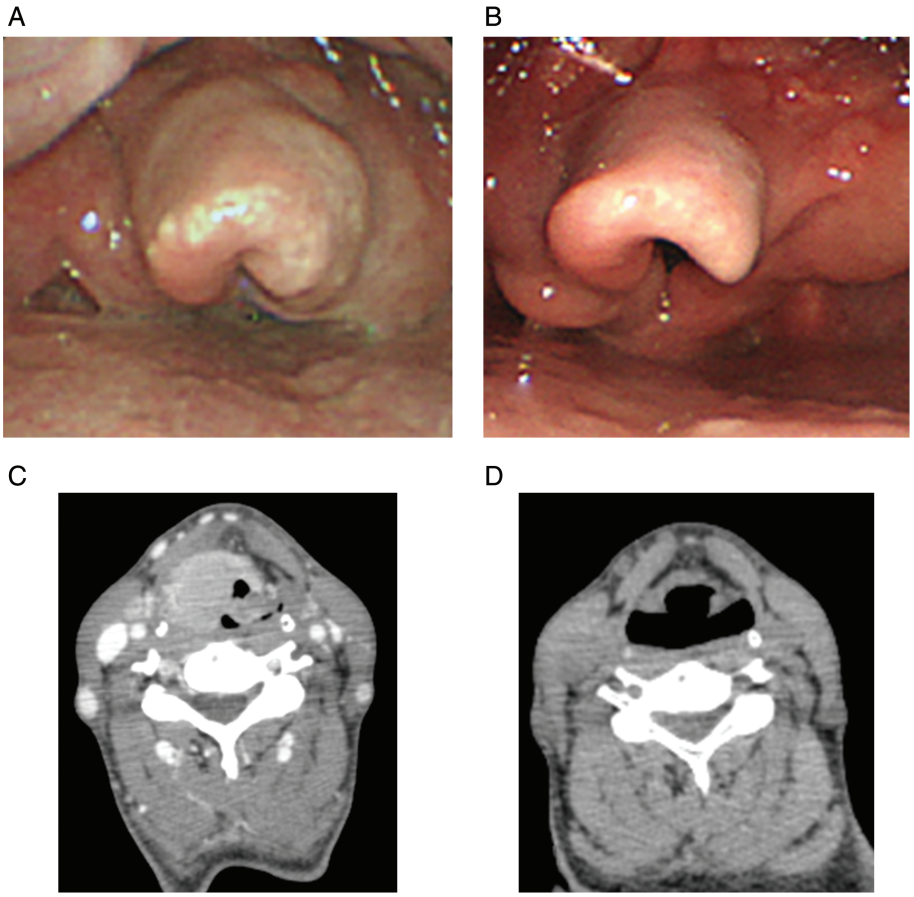

Figure 1. (A) Initial bronchoscopy image, showing swelling of the epiglottis with partly whitish nodular lesions and an irregular surface. Soft tissues beneath the lesions were oedematous. (B) Bronchoscopy image after steroid and cyclophosphamide therapy. Swelling of the epiglottis has decreased and whitish nodular lesions have disappeared. (C) Chest computed tomography (CT) on admission. The epiglottis was severely swollen and the airway partially obstructed. (D) Chest CT after steroid and cyclophosphamide therapy. Swelling of the epiglottis was decreased.

Discussion

GPA is a distinct clinicopathological entity characterized by necrotizing vasculitis of small vessels, along with granulomatous areas of inflammation and necrosis in the upper and lower respiratory tracts and glomerulonephritis. The most commonly affected organs in the upper respiratory tract are the paranasal sinuses, followed by the nose, nasopharynx, and larynx [2]. The epiglottis, however, is rarely affected, with few reports to date [1]. Macroscopic findings of epiglottitis associated with GPA include ulceration of the surface, oedematous changes, and partially granular lesions [1], with the latter two manifestations similar to those observed in our patient.

Biopsy specimens of the upper respiratory tracts of patients with GPA often reveal non-specific features of inflammation and necrosis, with or without granuloma formation; vasculitis is rarely observed [1,2]. Pathologically, the epiglottis of our patient showed only tissue necrosis and neutrophilic inflammation. These pathological findings are similar to bacterial infections. However, negative results with special staining for micro-organisms, ineffectiveness of treatment with antibiotics, and improvement with prednisolone and cyclophosphamide, these clinical findings are against the bacterial infections. Lung biopsy specimens, obtained during video-assisted thoracic surgery (VATS), showed vasculitis, but granulomas were not found. This may have been due to steroid therapy [3], which was started before the lung biopsy samples were obtained, to avoid upper respiratory obstruction by the epiglottitis.

Although testing for ANCA is very important in diagnosing GPA, approximately 10–20% of patients with GPA are negative for ANCA [4]. Classical pathological findings of GPA include granulomatous inflammation, necrosis, and small vessel vasculitis, but these findings are not observed in all patients with GPA. Our patient was negative for ANCA and only two of the three classical pathological findings of GPA were observed. Therefore, differential diagnosis should include microscopic polyangiitis (MPA), with a multidisciplinary approach necessary in making a diagnosis. Multiple nodular lesions on chest CT [5] and involvement of the upper respiratory tract are more indicative of GPA than MPA, but careful follow-up is required to confirm this diagnosis. Thus, a diagnosis of GPA in our patient should be considered provisional.

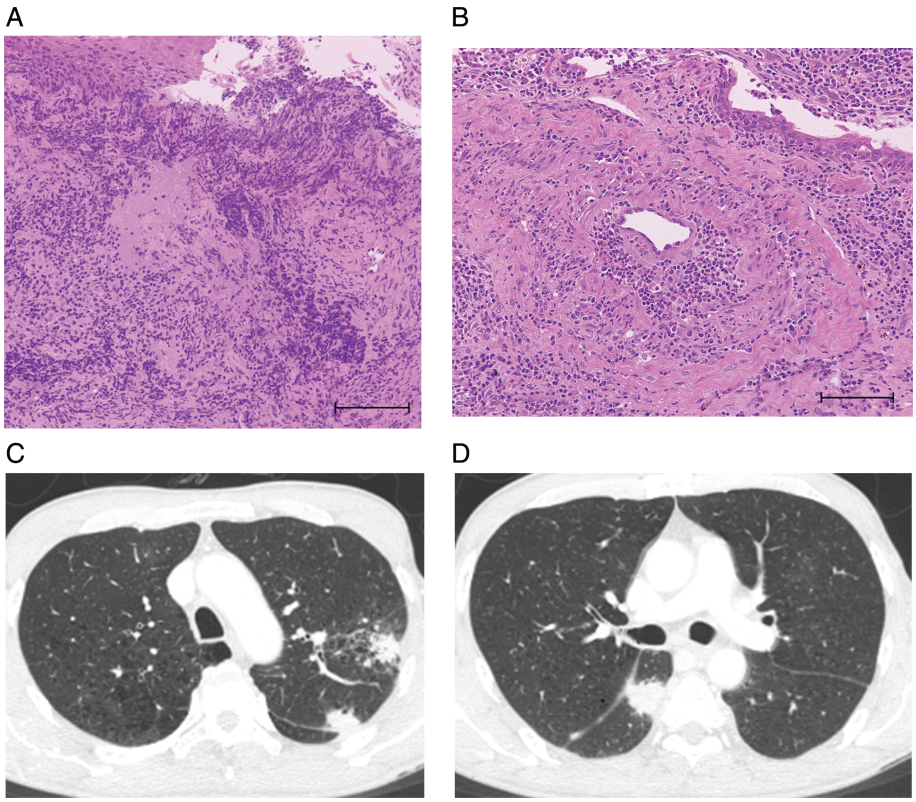

Figure 2. (A) Pathological findings of the epiglottis showed infiltrations of neutrophils and necrosis. Vasculitis, giant cells, and granulomas were not detected. Bar: 100 μm. (B) Pathological findings of the right lower lobe, showing neutrophilic vasculitis, with no evidence of geographic necrosis, giant cells, or granuloma formation. Bar: 200 μm. (C, D) Chest computed tomography on admission. Bilateral multiple nodular consolidations were also observed.

GPA should be included in the differential diagnosis of epiglottitis, especially in patients with lung parenchymal lesions suggestive of GPA. Detailed clinical, radiological, pathological, and microbiological examinations are needed to exclude other diseases.

Disclosure Statements

No conflict of interest declared.
Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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