

Case Report

Images in Clinical Otolaryngology: Nasal and Subglottic Manifestations of Granulomatosis with Polyangiitis

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Abstract

Introduction: Granulomatosis with Polyangiitis (GPA) is an antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis. Manifestations of this systemic vasculitis can be seen in otolaryngologic, pulmonary, renal, cutaneous and ophthalmologic organ systems. Otolaryngologic manifestations include nasal discharge or crusting, nasal septum perforation, scarring, subglottic inflammation, conductive hearing loss and sensorineural hearing loss. Herein, we present representative radiographic and endoscopic images of the nasal and subglottic manifestations of GPA.

Methods: Retrospective review of patient electronic medical record.

Case Presentation: A 66-year-old female presented to our institution with several years of nasal obstruction bilaterally. She had a 10-year history of GPA that included previous pulmonary involvement without renal manifestations. For the 18 months preceding her presentation, disease control was excellent.

At her first clinical evaluation, physical exam with rigid nasal endoscopy demonstrated significant scarring between the nasal septum and the lateral wall of the nose extending to the middle turbinate bilaterally. The patient was taken to the operating room for limited endoscopic sinus surgery and bilateral lysis of extensive intranasal synechiae. To date, the patient has had an excellent surgical result and is pleased with her functional result.

Conclusion: It is common for initial presenting symptoms of GPA to occur in the head and neck. Given the broad array of head and neck manifestations seen in this disease process, it is crucial for the otolaryngologist to be cognizant of possible relevant physical exam findings. This case reported here illustrates

representative endoscopic and radiographic findings. Treatment of otolaryngologic manifestations of GPA can include surgical treatment and subsequent improvement in patient symptoms, as in the case presented here.

Keywords

Subglottic; Granulomatosis with Polyangiitis; GPA

Abbreviations

ANCA: Antineutrophil Cytoplasmic Autoantibody

GPA: Granulomatosis with Polyangiitis

Introduction

Granulomatosis with polyangiitis (GPA) is a small and medium vessel vasculitis, that affects both male and female patients that are most often between 45 and 60 years of age. It is associated with cytoplasmic-ANCA (c-ANCA) and can include renal, pulmonary and otolaryngologic manifestations. In the kidneys, rapidly progressing glomerulonephritis can be seen while pulmonary nodules can be found in the lung. Limited forms of GPA tend to only affect the upper airway. Due to the variety of head and neck manifestations that GPA can present with, an understanding of this disease is essential for otolaryngologists. The purpose of this report is to illustrate common imaging and endoscopic exam findings found in GPA.

Methods

Retrospective review of patient electronic medical record was performed.

Case

A 66-year-old female with a 10 year history of GPA presented with several years of bilateral nasal obstruction. Her disease had previous pulmonary involvement without renal manifestations; she had no history of stridor or voice changes. She had excellent disease-control for the 18 months preceding her presentation.

At presentation, rigid nasal endoscopy demonstrated significant scarring between the nasal septum and the lateral wall of the nose extending to the middle turbinate bilaterally (Figure 1a). This scarring impeded thorough visualization of the remainder of the middle turbinate, inferior turbinate and obstructs a path toward examination of the nasopharynx. Computed Tomography showed bilateral dense opacification of the anterior nasal cavities consistent with scar seen on physical examination (Figure 1b), as well as mucosal thickening in the maxillary and ethmoid sinuses. Flexible fiberoptic laryngoscopy demonstrates a narrowing in the subglottis to roughly 10 mm in diameter without obvious active inflammation (Figure 1c). The patient was taken to the operating room for bilateral lysis of extensive intranasal synechiae and limited endoscopic sinus surgery. The patient was

seen at 1 week for nasal debridement and 10 weeks for subsequent follow up and was pleased with her nasal breathing. Examination at this time demonstrating a patent nasal cavity, with the full extent of the nasal septum and lateral nasal wall being easily visualized and noted to be free of scar (Figure 1d). To date, she has been followed for 6 months with excellent symptomatic and endoscopic results.

Discussion

GPA is an c-ANCA-associated vasculitis characterized by otolaryngologic, pulmonary, renal, cutaneous and ophthalmologic manifestations [1,2]. The first symptoms of GPA may be in the head and neck in 80-90% of patients [3]. Therefore, it is crucial for the otolaryngologist – head and neck surgeon to be cognizant of the variety of head and neck manifestations of this disease process. Radiographic and endoscopic images of our case of nasal and subglottic manifestations of GPA are presented here. Because of the likelihood of otolaryngology involvement, it is also essential to be familiar of the natural history of the disease process.

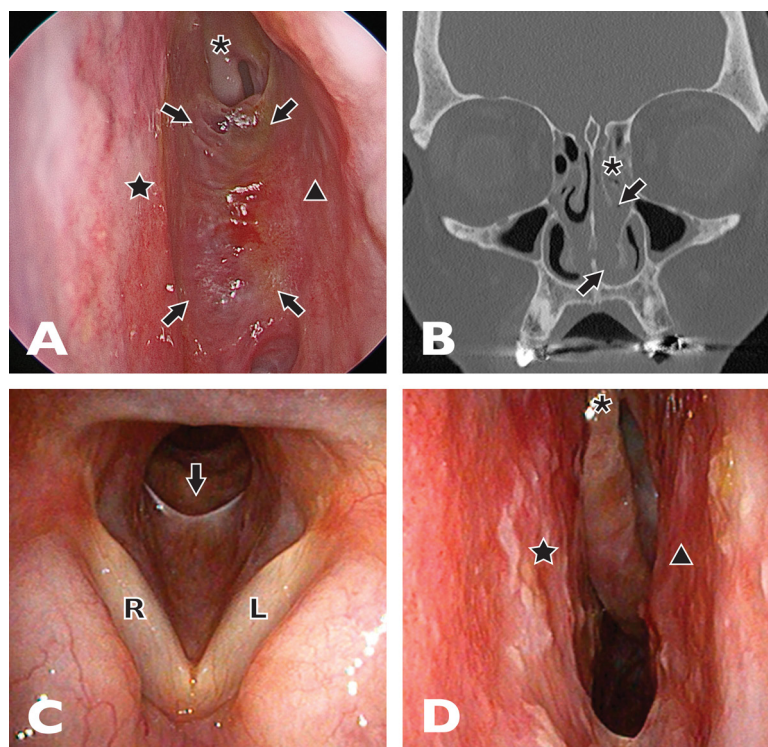


Figure 1:

- (a) Endoscopic view of left nasal cavity demonstrating extensive scarring (arrows) between the nasal septum (star) and lateral nasal wall (triangle). The root of the middle turbinate attachment is also shown (asterisk)
- (b) CT demonstrating scarring (arrows) and root of middle turbinate (asterisk)
- (c) Flexible fiberoptic laryngoscopy demonstrating subglottic stenosis (arrow) inferior to the left (L) and right (R) vocal folds
- (d) Endoscopic view of left nasal cavity at 10 post-operative weeks demonstrating patent nasal cavity, free of scar. Shown are nasal septum (star), lateral nasal wall (triangle), root of middle turbinate attachment (asterisk)

Head and neck manifestations in the sinonasal cavity include nasal discharge or crusting, nasal septum perforation, scarring, in the upper airway include subglottic inflammation, and in the ear include conductive hearing loss and sensorineural hearing loss [4-6]. In some scenarios, the disease may initially present at a single site (i.e. nasal septal perforation). Frequently, serum analysis will not yet be positive for c-ANCA, making for a diagnostic challenge when considering potential underlying etiologies such as ANCA-negative vasculitis, neoplasm drug-induced or idiopathic. Over the course of the disease, the incidence of these nasal, otologic and subglottic manifestations increase. In one study, disease involvement over time spread to additional sites in the head and neck in more than half the patients studied [6].

In patients with GPA, approximately 16% to 23% of patients are found to have subglottic stenosis [7]. Multiple observations have shown subglottic stenosis to occur more frequently in patients who have the onset of the disease at a younger age. Our patient did have evidence of subglottic narrowing requiring placement of a 5.0 cuffed endotracheal tube at the time of surgery. Approximately 16% to 19% of patients have otologic complaints [8]. Our patient did not present with otologic complaints.

Medical and surgical therapies are mainstays of treatment. Medical therapy is typically immunomodulating and most commonly includes methotrexate and cyclophosphamide. Surgical therapy is typically targeted at symptom improvement. In one study, 79% the patients in the study received surgical intervention of head and neck disease as part of disease treatment [6]. The most common intervention is airway intervention at the level of the subglottis. While our patient did not require subglottic intervention, the nasal cavity did require lysis of synechiae for relief from nasal obstruction.

Conclusion

GPA is a small and medium vessel vasculitis that affects the renal, pulmonary and otolaryngologic systems. Given the potential for head and neck manifestations to be the presenting symptoms of this disease, it is imperative for the otolaryngologist to be famil-

iar with the presentation and management of this disease. This report highlights a representative case of GPA with otolaryngologic manifestations and presents relevant imaging and physical exam findings.

Conflict of Interest

No conflicts of interest to declare

Patient Consent

Exempt from IRB

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