

GRANULOMATOSIS WITH POLYANGIITIS: IMPLICATIONS ON ANESTHETIC MANAGEMENT

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Abstract

This is the case of a 44-year-old lady, known to have GPA since 9 years, who presented with symptoms of shortness of breath at rest. On presentation, laryngoscopic examination revealed bilateral mobile cords with significant subglottic stenosis. Computerized tomography of the neck revealed a subglottic stenosis extending over a length of 2.8cm with the airway measuring 9x6mm at the narrowest level. Anesthesiologists must bear in mind the multi-organ system implications of GPA when managing the airway. Delicate anesthetic, pharmacologic, and surgical regimens must be utilized in order to deal with the difficult airway and avoid inadvertent airway risks and unnecessary tracheostomies.

Keywords: Granulomatosis with polyangiitis, Wegner's disease, stenosis, anesthesia, intubation.

Introduction

Granulomatosis with polyangiitis (GPA), also known as Wegner's granulomatosis, is a necrotizing non-caseating granulomatous inflammatory disease of the respiratory tract accompanied by vasculitis of small and medium sized vessels¹. With a prevalence of three cases per 100,000 individuals, GPA is a rare disorder with a peak incidence in the fifth decade of life². The involvement of the laryngotracheal complex in GPA is not rare and ranges from mild narrowing to severe circumferential stenosis that can be life threatening. The active phase of GPA is characterized by inflammation of tracheal mucosa which, within a variable period of time, can cause fibrotic scar tissue formation in an already compromised lumen eventually leading to symptoms of dyspnea, cough, dysphonia, and stridor^{3,4}.

Daijo et al reported the first case of unexpectedly difficult intubation leading to a diagnosis of GPA⁵. As tracheal inflammation and subglottic stenosis in GPA can cause inadvertent risk for endotracheal intubation, the anesthetic management in these patients requires consideration of the pathophysiological alterations in this disease. Reports on GPA and anesthesia are in fact limited^{5,6,7}. The purpose of this report is to describe a rare case of GPA affecting the upper airway and shed more light on the anesthetic management in these patients.

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Case Report

This is the case of a 44-year-old lady, known to have GPA since 9 years, who presented with progressive shortness of breath at rest. The patient was a nonsmoker and had an otherwise negative past medical and surgical history. Her disease had been under control for several years on cycles of intravenous dexamethasone, ranitidine, and rituximab. Examination of nasal cavity revealed excessive crusting and irregular granular thickening of the mucosa (See figure 1). Laryngoscopic examination revealed bilateral mobile cords with significant subglottic stenosis (See figure 2). Patient was prescribed prednisone 20mg orally daily and computerized tomography of the neck without intravenous contrast was requested to further delineate the extent of her airway disease. Radiologic imaging revealed a subglottic stenosis extending over a length of 2.8cm with the airway measuring 9x6 mm at the narrowest level (See figure 3). On follow-up 10 days later, patient reported marked improvement in her shortness of breath.

Discussion

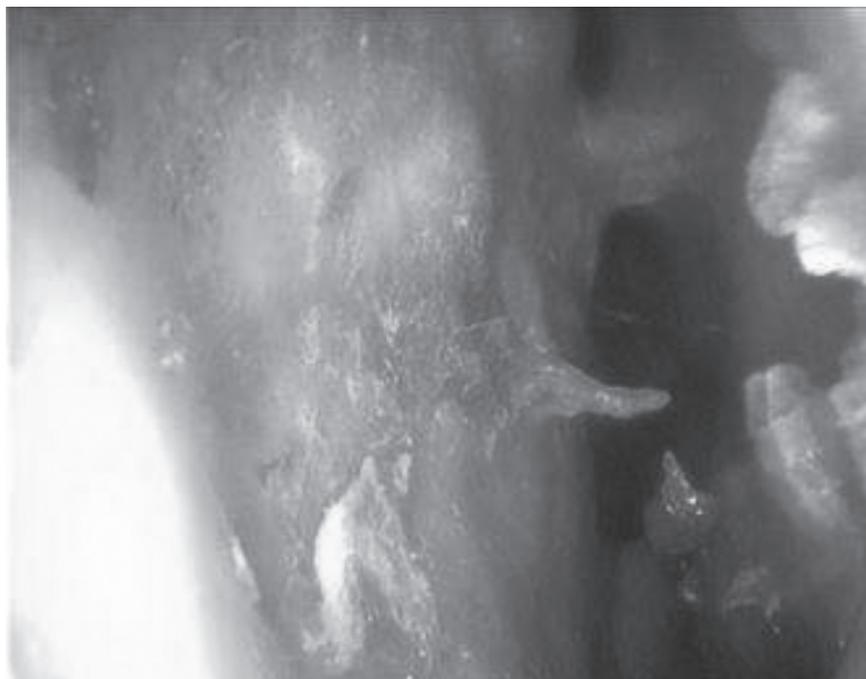
GPA is a rare autoimmune disease of unknown etiology that can affect multiple organ systems. When

the sinonasal tract is affected, patients complain of recurrent epistaxis, nasal crusting and nasal obstruction with facial heaviness, pain, and tenderness. These symptoms are invariably secondary to irregularly thickened nasal mucosa with granular formation, crusting, ulceration, and necrosis often times leading to septal perforation and nasal saddle deformity⁸⁻¹⁰. On the other hand, laryngotracheal involvement may remain silent with subtle or non-existent symptoms in view of the rarity of vocal cords involvement. A retrospective chart review of 35 GPA patients by Gardiani et al revealed multilevel airway involvement in one third of patients and cricoarytenoid involvement with fixation of the vocal folds in 20% of the cases¹¹. Since airway involvement may precede systemic manifestations of the disease, physicians must have a high degree of suspicion in order to ensure a timely diagnosis and prevent inadvertent risks during surgical and anesthetic management.

To that end, anesthetic management in these patients should start with the assumption of a difficult airway. Although the implications of the disease on anesthetic management are ample, reports on the anesthetic management of GPA are limited^{5,6,7}. Anesthesiologists must take into consideration the abnormalities in the various organ systems involved, namely respiratory, cardiovascular, nervous and renal

Fig. 1

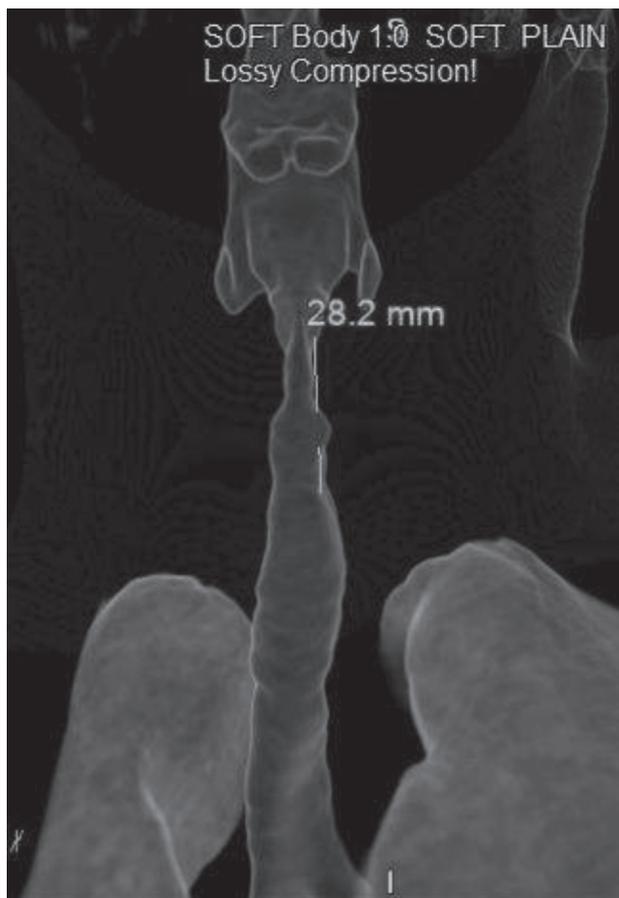
Nasal cavity with excessive crusting and irregular granular thickening of the mucosa



*Fig.2
Laryngoscopic examination revealing
normal vocal cords with significant
subglottic stenosis*



*Fig. 3
Computerized tomography revealing subglottic stenosis over a
length of 2.8cm and measuring 9x6mm at narrowest level*



systems⁶. When a diagnosis of GPA is suspected, anesthesiologists are encouraged to perform a thorough nasal and laryngoscopic examination to note the presence of any crusts, ulcers or perforations in the nose, palate or pharynx^{6,12}. Careful planning and extreme gentleness during intubation are necessary to avoid bleeding from granulomas or dislodgement of crusts or tissue into an already narrowed airway as a case of mortality due to impaction of nasal crusts has been reported¹². Subsequently, endotracheal tubes of different sizes, especially smaller sizes, must be prepared and available at hand.

GPA may also involve the coronaries and peripheral arteries and veins¹³. Anesthesiologists must avoid any increase in preload, afterload, or heart rate that may cause myocardial ischemia or arrhythmias⁶. Due to the risk of infarction of the digits, caution must be taken with arterial punctures and indwelling arterial lines as well⁶. In addition, careful neurologic examination must be done in order to rule out relative anesthesia contraindications. At the same time, anesthetics and medications that are renally excreted must be used with caution^{6,13}. Therefore, special treatment employing delicate anesthetic, pharmacologic and surgical regimens must be used to avoid inadvertent airway risks and otherwise unnecessary permanent tracheostomies¹⁴.

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