J Anesth (2010) 24:284–286
DOI 10.1007/s00540-009-0842-4

CLINICAL REPORT

Unexpectedly difficult intubation caused by subglottic stenosis in Wegener's granulomatosis

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Received: 16 May 2008 / Accepted: 4 September 2009 / Published online: 23 January 2010

Abstract A 76-year-old woman was scheduled to undergo abdominal aortic repair for progressive abdominal aortic aneurysm. After inducing general anesthesia, the 7.5-mm internal diameter (ID) tracheal tube could not be advanced below the level of the vocal cords because of resistance, and intubation was re-attempted several times using smaller tubes. An otolaryngologist was consulted and subglottic stenosis of unknown origin was suggested. The aortic repair was cancelled and tracheostomy was performed instead. She was diagnosed with Wegener's granulomatosis 46 days after the operation because she developed symptoms of renal dysfunction, hemoptysis, gastrointestinal bleeding, and presence of anti-neutrophil cytoplasmic autoantibodies (c-ANCA). The patient was treated with steroids but died 89 days after the operation because of pulmonary bleeding and renal dysfunction. Tracheal stenosis is a rare presenting feature of Wegener's granulomatosis that usually occurs late in the disease; however, anesthesiologists around the world need to bear in mind that the disease can present airway symptoms and can be the cause of airway obstruction.

Keywords Wegener's granulomatosis · Subglottic stenosis · Difficult intubation

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Introduction

Wegener's granulomatosis is a necrotizing systemic vasculitis of unknown etiology, characterized by granulomatous lesions of the respiratory tract and variable renal involvement [1, 2]. Tracheal stenosis is a rare presenting feature of Wegener's granulomatosis that usually occurs late in the disease.

We report a case of unexpectedly difficult intubation in an adult caused by subglottic stenosis. The patient was diagnosed with Wegener's granulomatosis 46 days after the operation because she developed symptoms of renal dysfunction, hemoptysis, gastrointestinal bleeding, and the presence of anti-neutrophil cytoplasmic autoantibodies (c-ANCA).

Case report

A 76-year-old woman (height 149 cm, weight 49 kg) was scheduled to undergo abdominal aortic repair for progressive abdominal aortic aneurysm. She had a history of hypertension, hyperlipidemia, and asthma.

She was medicated with enalapril maleate and theophylline. She presented the symptoms of inspiratory stridor and dysphonia. She underwent paranasal sinusitis surgery 15 years ago.

An epidural catheter was placed without difficulty on the day before the surgery. On arrival at the operating theater, her blood pressure was 155/82 mmHg, pulse rate was 83 beats/min, and saturation of percutaneous oxygen (SpO_2) in room air was 96%. Cannulation on the radial artery was attempted under local anesthesia, but it was unsuccessful. General anesthesia was induced with thiopental 300 mg and vecuronium 10 mg.

The vocal cords were seen clearly at laryngoscopy, but a 7.5 mm internal diameter (ID) tracheal tube could not be advanced below the level of the vocal cords because of resistance. Intubation was re-attempted several times using smaller tubes with ventilation by face mask. Sevoflurane was added to maintain anesthesia. Finally, a 5.0 mm ID tube was passed successfully through the vocal cords and secured in place. Because of the unexpected difficulties with intubation, an otolaryngologist was consulted. Emergency fiber-optic laryngoscopy suggested subglottic stenosis of unknown origin. The aortic repair was cancelled and tracheostomy was performed instead. After the tracheostomy, fiber-optic laryngoscopy was again performed to confirm the subglottic stenosis (Fig. 1). The patient expressed that she could breathe more easily after tracheostomy.

The operation was rescheduled for a week later, and the anesthesia and abdominal aortic repair were performed uneventfully. The patient showed signs of renal dysfunction postoperatively and Wegener's granulomatosis was suspected. She was diagnosed with Wegener's granulomatosis 46 days after the operation because of the developing symptoms of renal dysfunction, hemoptysis, gastrointestinal bleeding, and presence of anti-neutrophil cytoplasmic autoantibodies (c-ANCA). Rapidly progressive glomerulonephritis was present at renal biopsy. The patient was treated with steroids but died 89 days after the operation because of pulmonary bleeding and renal dysfunction.

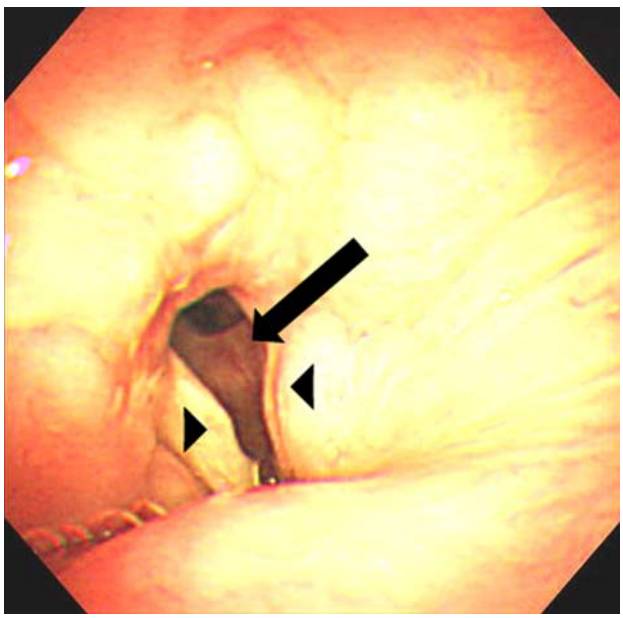

Fig. 1 Fiberscopic image taken during laryngoscopy; image shows focal subglottic stenosis

Discussion

We have reported a case where an unexpectedly difficult intubation lead to the diagnosis of Wegener's granulomatosis. Wegener's granulomatosis is a necrotising systemic vasculitis of unknown etiology [1]. The disease is characterized by pathophysiologic changes due to the formation of necrotising granulomas in inflamed vessels, such as those present in the central nervous system, upper and lower respiratory tract, cardiovascular system, and kidneys [3]. Although the lung is the most commonly affected organ (94% of cases) at histologic examination in many cases, the initial symptoms are typically those of upper respiratory tract involvement, including rhinitis, sinusitis, and otitis media [4]. In our case, subglottic stenosis, the cause of the unexpectedly difficult intubation, was the initial symptom that led us to suspect Wegener's granulomatosis. Discovering it prompted us to perform the c-ANCA test, which is a specific test for Wegener's granulomatosis [5].

Our case is the first reported case of an unexpectedly difficult intubation leading to the diagnosis of Wegener's granulomatosis with the aid of the c-ANCA test.

Reports on this disease and general anesthesia are limited [6, 7]. A recent report described a 10-year-old patient with Wegener's granulomatosis who required tracheostomy due to severe respiratory distress caused by granulations in the subglottic region and both bronchi [7]. The diagnosis was made only after the endoscopy and biopsy following the operation.

As reported for this patient, renal failure is the most common cause of death in untreated patients, but renal disease is rarely evident at presentation. Although the diagnosis of Wegener's granulomatosis is difficult, early recognition of the disease is important. The untreated disease is almost uniformly fatal, with a mean survival time of 5 months [4].

Although cardiovascular involvements were evident and life-threatening in the present case, cardiac involvement is uncommon in this disease. Unfortunately, the airway symptoms presented by the patient, such as rhinitis and sinusitis, which are criteria for the disease [2], were not regarded as important preoperatively. Definitive diagnosis was delayed only after the subglottic stenosis was found at the time of intubation.

Airway management of Wegener's granulomatosis starts with the assumption of difficult intubation due to a narrowed respiratory tract. Endotracheal tubes (ETT) of various—especially smaller—sizes must be prepared. Gentle management of ETT is mandatory in order to avoid unnecessary bleeding in the respiratory tract [3, 8]. In the present case, tracheostomy was performed because the cause of the airway obstruction was not clear. Tracheostomy may be

unavoidable in some other cases too, because early extubation is reported to be related to airway obstruction [7].

Special treatment involving a delicate combination of surgical and pharmacological regimens may be required to avoid permanent tracheostomy [9].

Wegener's granulomatosis is rare and there is a preponderance of cases of it in Caucasians [1]. However, anesthesiologists around the world must bear in mind that this disease can be a cause of unexpected upper airway obstruction.

Early diagnosis of the disease with the help of the c-ANCA test may benefit patients by enabling earlier commencement of treatment.

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