Undiagnosed Scleroderma in a Patient with a Difficult Airway

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INTRODUCTION

Connective tissue disorders occur because of autoimmune-influenced destruction or synthesis of any of the components that are found in these tissues. Systemic involvement of multiple organs is common. Patients are often chronically ill and treated with corticosteroids. Scleroderma or progressive systemic sclerosis (PSS) is a generalized disorder of connective tissue characterized by degenerative and inflammatory changes that subsequently leads to intense fibrosis [1]. Due to systemic involvement, patients with scleroderma can pose a challenge to the anesthesiologist. In this case report, we describe a patient with an unrecognized difficult airway associated with undiagnosed scleroderma. This disease process not only contributed to our difficulty in visualization of the larynx but also impeded other maneuvers to secure the airway.

CASE REPORT

A 56-year-old, 110 kg, 75 inch Caucasian male was scheduled to undergo an elective coronary artery bypass operation for severe three-vessel coronary artery disease and crescendo angina over a one-month period. He had a normal left-ventricular ejection fraction. Other past medical history included: well-controlled insulin-dependent diabetes mellitus (50 years), hypertension and hypercholesterolemia. The patient also gave a history of sleep apnea, although he was under no treatment and had no signs nor symptoms other than snoring while sleeping. He gave no history of prior surgery. His medications consisted of insulin NPH insulin, 26 units in the morning; diltiazem, 180 mg daily; and simvastatin, 20 mg at bedtime. Vital signs: blood pressure, 130/70 mm Hg; pulse rate, 77 bpm; respiratory rate, 12 bpm; and temperature, 37°C. Inspection of the airway revealed a Mallampati airway class I. The thyromental distance measured 7 cm, and the distance between upper and lower central incisors was 6 cm. Neck extension was limited to 50 degrees. This was not felt to be a contraindication to direct laryngoscopy to secure the airway. The remainder of his physical exam and laboratory investigations was normal. The patient was premedicated with morphine sulfate 10 mg IM and scopolamine 400 g IM one hour before surgery. In the operating room, arterial and intravenous cannulation were accomplished without difficulty, although two attempts were needed to thread the arterial catheter. Prior to induction, the patient was monitored with intra-arterial blood pressure, ECG (leads II and V5) and SPO2. Induction of anesthesia was accomplished with 40g/kg fentanyl and succinylcholine continuous infusion (150 mg). Two persons were required for satisfactory mask ventilation. Neither glottis nor epiglottis was visualized during direct

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b Abbreviations: PSS, progressive systemic sclerosis; FOB, fiberoptic bronchoscope.
laryngoscopy using Macintosh 3,4, Miller 2,3 blades and/or changing head position. Fiberoptic intubation was unsuccessful because of the inability to visualize the larynx due to redundant oropharyngeal soft tissue. At this point, it was decided to perform a retrograde wire intubation. Multiple attempts were required to pass a 14G Jelco needle into the trachea due to obstruction of the needle by skin plugs. Upon further scrutiny, the patient’s skin was found to be stiff and thickened. Consequently, a tentative diagnosis of scleroderma was made. A retrograde wire was ultimately passed and placed into the suction port of a fiberoptic bronchoscope (FOB). The FOB was advanced through the oropharynx but was unable to be advanced into the trachea. The FOB was removed, and an endotracheal tube changer was placed over the wire (Cook Airway Exchange Catheter C-CAE-19, Cook Critical Care, Bloomington, IN). A 7.5 endotracheal tube was passed over the tube changer into the trachea, and the wire and tube changer were removed. Breath sounds were confirmed by auscultation and end tidal CO2. SP02 was maintained at greater than 90 percent throughout this process. A transosophageal echocardiography probe was not placed because of potential involvement of the esophagus. The remainder of the operation was uneventful, and the patient was extubated the following morning without difficulty with the anesthesia care team in attendance. The patient sustained no sequelae of the intubation. Because the patient had signs consistent with scleroderma, multiple biopsies were obtained. Microscopic analysis of the skin biopsy was consistent with PSS while the other biopsies (cardiac, pleural and aortic) were negative.

**DISCUSSION**

Our patient presented with some of the skin manifestations of PSS: namely, thickened and inelastic skin. This impeded our ability to secure his airway because of two reasons. First, the patient had limited neck extension making direct laryngoscopy impossible. Furthermore, his thickened skin made trans-tracheal cannulation difficult. Other contributing factors may have led to a difficult intubation in this patient. His history of sleep apnea combined with increased body weight could have contributed to redundant soft tissue in the oropharynx, but his body fat distribution was predominantly below the thorax. Additionally, diabetes mellitus can lead to diminished range of motion in joints like the cervical spine, although this patient had no other sites of joint involvement.

PSS is an autoimmune disease of unknown etiology affecting three females for every male. It usually manifests in the third to fifth decades of life [2]. Progressive inflammation and fibrosis occurs in the skin, heart, lungs kidneys and gastrointestinal tract. Progression of these lesions leads to thickening and tightening of the skin and the destruction of the normal architecture of visceral organs. Pathophysiologically, early infiltration by T-cell lymphocytes of target organs is seen. This stimulates fibroblasts to produce excessive amounts of normal collagen. Later, collagen-laden tissues are seen without lymphocytes [3]. The diagnosis of PSS is made clinically and histopathologically with a biopsy of the involved time. Laboratory investigations are non-diagnostic, but anti-nuclear antibodies are present in most patients. The speckled or nucleolar pattern is seen in 65 percent of PSS patients, while 20 percent of patients show reactivity to ANA scl-70 [3]. Anemia of chronic disease is often present. Treatment options are supportive and symptomatic. Steroids are used occasionally for inflammation. D-penicillamine prevents cross-linking of collagen and has an immunomodulating effect. Progression of PSS leads to death due to hypertensive renal disease, heart failure, pulmonary disease and malabsorption syndromes. The CREST syndrome is a PSS variant. Calcinosis, Raynaud’s syndrome, esophageal dysmotility, sclerodactyly and telangiectasias are seen. A lack of visceral
involvement and a more benign course characterizes this variant. ANA against centromere is seen in 80 percent of patients with CREST syndrome.

During pre-operative evaluation, attention to the skin, musculoskeletal system, heart, lungs, kidneys, gastrointestinal tract and temporomandibular joint should be paid by the anesthesiologist. Skin fibrosis may make vascular access difficult and central venous catheterization may be necessary. Raynaud’s disease is commonly present.

Intraoperatively, patients should be kept warm, administered warm intravenous fluids and medications to prevent Raynaud’s phenomena. If possible, arterial cannulation and vasoconstrictors should be avoided. Cardiac involvement can include cardiomegaly, pericarditis, pericardial effusions and conduction defects. Pulmonary infiltration can lead to abnormal diffusion capacity, fibrosis and pulmonary hypertension. Vitamin K malabsorption may lead to clotting abnormalities. Gastrointestinal involvement is common with the esophagus as the target organ. Symptoms may include dysphagia and gastric reflux. Aspiration prophylaxis should be prescribed. Alternate methods to secure the airway may be needed as TMJ fibrosis will impede the ability to open the mouth. Limited neck extension may be present secondary to thickened and inelastic skin. Furthermore, oral and nasal telangiectasias can bleed extensively when manipulated. Sellick’s maneuver will be ineffective due to a fibroed esophagus. Additionally, a rapid sequence induction may be difficult because of TMJ fibrosis and limited neck extension. Therefore, alternate plans to secure the airway, including awake fiberoptic intubation, should be made. Regional anesthesia is an acceptable alternative, but there are several case reports of prolonged sensory block following conduction anesthesia [5, 6].

We have presented a case of a patient with a difficult airway with previously unrecognized scleroderma that required a retrograde wire intubation. Patients with scleroderma undergoing surgical procedures present a challenge to anesthesiologists because of the disease’s multisystem involvement. Although the etiology of scleroderma is unknown, anesthesiologists can take precautions in the operating room to prevent exacerbation of the condition during surgery.

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