CLINICAL VIGNETTE

Anesthetic Management for Video-Assisted Thoracoscopic Surgery in a Patient with VACTERL Association

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Introduction

VACTERL association is a sporadic multisystem disorder affecting between 1/10,000 – 1/40,000 live-born infants.[1] It is usually diagnosed clinically by the concurrence of at least three of the following features: vertebral anomalies (V), anal atresia (A), cardiac malformations (C), tracheoesophageal fistula (TE), renal dysplasia (R) and limb abnormalities (L). The exact cause of the VACTERL association is currently unknown, but thought to be multifactorial.[2] In this case report, we present the anesthetic management of a male with previously undiagnosed VACTERL association who underwent a left video-assisted thoracoscopic surgery for spontaneous pneumothorax.

Case

A 19-year-old male presented to the emergency department with a one day history of left-sided chest pain and acute dyspnea following methamphetamine use. Upon arrival to the emergency department, his dyspnea was noted to be severe and he had a witnessed syncopal episode. A chest x-ray revealed a large left-sided tension pneumothorax and needle decompression was performed with improvement in symptoms. A left-sided chest tube was subsequently placed.

On the second day of admission, the thoracic surgery team planned to take the patient to the operating room for a left video-assisted thoracoscopic surgery and the anesthesiology service was consulted. Upon interview, the patient reported a history of tracheo-esophageal fistula that was repaired at 24 hours of life. There were no obvious complications from the surgery although details regarding the surgery and follow-up care were scant. He denied any history of cardiac, renal or anal involvement. He reported a ten month history of daily methamphetamine use prior to presentation as well as longstanding tobacco use. Other than immediately prior to admission, he typically had excellent exercise tolerance without symptoms of dyspnea, dizziness or chest pain. He weighed 52 kg and was 170 cm tall. On examination, he had a webbed neck however the mandible appeared normal. He had normal cervical range of motion and thyromental distance and the Mallampati score was two. There were no murmurs detectable. Examination of the back revealed mild scoliosis but otherwise normal appearing spinous processes. He had bilateral radial hypoplasia with non-palpable bilateral radial pulses. Bilateral radial arteries were not detectable on ultrasound. Head CT was negative for hemorrhage and Chest CT demonstrated normal appearing heart, trachea and esophagus. There was congenital fusion of C6, C7, T1 vertebral bodies and bilateral cervical ribs were noted at C7. EKG was unremarkable.

A thoracic epidural catheter for postoperative pain control was placed uneventfully at the T5-T6 interspace immediately prior to taking the patient to the operating room. Following a negative aspiration test, 3 ml of 1.5% lidocaine with 1:200,000 epinephrine was injected into the catheter and a T4-T9 and T4-T11 block to cold was demonstrated on the left and right, respectively. Upon arrival to the operating room, a ClearSight (Edwards Lifesciences Corporation, Irvine, California) continuous, noninvasive blood pressure monitor was placed on the patient's left fourth finger. Anesthesia was induced intravenously and direct laryngoscopy revealed a grade one view. A 39 F left-sided double lumen endotracheal tube was placed into the trachea and positioned using a bronchoscope uneventfully. Intravenous access proved difficult and a triple-lumen central venous catheter was placed using ultrasound guidance in the right internal jugular vein without complication. The patient was positioned in the right lateral decubitus position. The surgeons proceeded with mechanical and chemical pleurodesis and wedge resection of the apical segment of left upper lobe and superior segment of left lower lobe. Two 28 Fr tube thoracostomies were placed. The surgery was well tolerated by the patient with stable hemodynamics and absence of significant hypoxemia during one lung ventilation. At the conclusion of surgery, the patient was successfully extubated in the operative room. The patient's post-operative pain was well-controlled using patient-controlled epidural anesthesia. On post-operative day three the patient's chest tubes were discontinued and the patient was discharged from the hospital.

Discussion

Reports of the management of anesthesia in patients with VACTERL association are limited. In our patient, the VACTERL association had not been previously diagnosed prior to our evaluation. The diagnosis was made based on the following three findings: vertebral anomalies, history of tracheo-esophageal fistula and radial limb abnormalities. Given the multisystem involvement of the VACTERL association, these patients can present various challenges for the anesthesiologist.

A literature search revealed only three prior case reports of neuraxial anesthesia in patients with VACTERL association, all of which were lumbar epidural catheters placed in pregnant patients.[3-5] As vertebral anomalies are a hallmark of the VACTERL association, occurring in approximately 60-80% on patients, due caution should be employed when considering neuraxial anesthesia in these patients. Vertebral abnormalities can arise from a failure of formation (hypoplastic vertebrae, hemivertebrae or "butterfly" vertebrae) or a failure of segmentation (fused vertebrae, block vertebrae). In patients with colorectal or urogenital abnormalities, neurologic anomalies have also been reported such as tethered spinal cord and lipomeningomyelocele. A thorough review of the patient's neurologic history and imaging studies, which included chest x-ray and CT, was undertaken before consideration of neuraxial anesthesia in our patient. Other than mild scoliosis, which could potentially make placement technically more difficult, there were no obvious contraindications to the procedure for our patient and placement was uneventful.

Cardiac abnormalities occur in 50-80% of VACTERL patients, most frequently including ventricular septal defects, atrial septal defects and Tetralogy of Fallot. A preoperative echocardiogram is typically indicated, particularly if the patient demonstrates signs of cardiac compromise. In our patient, who had excellent exercise tolerance with an unremarkable cardiac physical exam and electrocardiogram, as well as a normal sized heart on prior imaging studies, an echocardiogram was not pursued preoperatively. Likewise, a renal ultrasound can be considered to rule out kidney involvement as part of the VACTERL association. Review of our patient's chemistry studies did not reveal any signs of renal involvement and anatomy appeared normal on CT and therefore an ultrasound was not taken preoperatively.

Intraoperative monitoring for thoracic surgeries at our institution typically includes a radial arterial catheter for close monitoring of blood pressure and arterial blood gases as well as large bore intravenous access for the possibility of sudden blood loss during surgery. In the case of our patient, both procedures proved difficult. The patient's radial arteries were undetectable by both palpation and ultrasound and were likely absent, as can be seen in up to 86% of cases of VACTERL.[6] The lack of veins appropriate for cannulation was also noted early on during the evaluation of the patient. The use of a continuous, noninvasive blood pressure monitor allowed for close hemodynamic monitoring while avoiding cannulation of the patient's brachial or femoral arteries and a central venous catheter provided adequate access for the surgery.

The management of the airway in VACTERL patients can be particularly challenging given their cranio-facial and vertebral anomalies. In our patient, although he did have a webbed neck, the remainder of the airway exam was reassuring. Features of VACTERL that would make airway management difficult, such as macrosomia or hypoplastic mandible or cervical instability were absent. Despite this, we were sure to have all difficult airway equipment available during the intubation. In this case, the patient's tracheo-esophageal repair was remote without any residual symptoms or complications seen on imaging studies. We therefore proceeded with a typical placement of a left-sided double-lumen endotracheal tube, as well as an early extubation of the trachea in the operating room. In patients with an unrepaired or complicated tracheoesophageal fistula, management of ventilation may require placement of the endotracheal tube distal to the level of the fistula and extubation should be carefully considered in patients that may have an increased risk of aspiration, such as those with unrepaired cleft lip or palate abnormalities.

In this case report, we describe the successful management of anesthesia for thoracic surgery in a patient with VACTERL association, which included single lung ventilation via a double-lumen endotracheal and a thoracic epidural catheter for postoperative pain control. This report highlights the many anesthetic considerations that are required when evaluating a patient with such an association involving varied organ systems.

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