CLINICAL VIGNETTE

An Obstetric Patient with Scimitar Syndrome and Its Anesthetic Considerations

Festus Ohan, MS4, Elizabeth Tsai, MD and Fei Zheng-Ward, MD

Background

Scimitar syndrome is a rare congenital heart and lung malformation in which there is anomalous pulmonary venous return to the inferior vena cava from either part or the entire right lung.[1] There have also been rare cases in which left-sided Scimitar syndrome has been described where left pulmonary veins connect to the inferior vena cava. Scimitar syndrome has been reported in 3% to 6% of patients with partial anomalous venous connection and about 70% of patients have an associated atrial septal defect.[2] Other cardiac anomalies that can be present in Scimitar syndrome include, but are not limited to, ventricular septal defect, total anomalous pulmonary venous return, hypoplastic left heart syndrome, subaortic stenosis, patent ductus arteriosus and tetralogy of fallot. A variety of additional cardiac anomalies are seen in 10-30% of patients with Scimitar syndrome. Due to the varying degrees of hypoplasia in the right pulmonary artery and bronchial anomalies, dextrocardia is often present as a result of the hypogenetic right lung with accompanying mediastinal shift.[3]

The clinical presentation of Scimitar syndrome varies ranging from mild symptoms in childhood to severe congestive heart failure early in infancy.[4] Symptoms include dyspnea, cyanosis, palpitations, syncope, respiratory distress, cardiac failure or recurrent pneumonia. Symptoms may present early during infancy (infantile form) or it can also be found incidentally with mild or no symptoms (childhood/adult form).

Case Report

A 29-year-old pregnant G2P1 female with Scimitar syndrome presented to Labor and Delivery for anticipated normal spontaneous vaginal delivery, with possible cesarean section. She was diagnosed with Scimitar syndrome in 2012 after a chest radiograph was obtained for asthma evaluation. She subsequently underwent a CT angiogram and transthoracic echocardiogram revealed a normal right ventricular function and size. The patient denied any cardiopulmonary limitations with activity, palpitations, or presyncope/syncope. She was evaluated by cardiology, who recommended continuated monitoring without any current interventions since that patient was asymptomatic. Review of systems was negative except medical history above. Her first pregnancy was two years prior with successfully uncomplicated vaginal delivery.

Noteworthy physical exam findings included a blood pressure of 89/53, but otherwise normal vital signs. Cardiac exam revealed a normal rate and rhythm without murmurs, rubs, gallops. There was no increase in jugular venous distention or lower extremity edema. Lungs were clear to auscultation bilaterally. Airway exam was reassuring with a Mallampati Classification of II and normal mouth, neck, and dental exam.

Patient had a previous trans-thoracic echocardiography demonstrating a left ventricle ejection fraction of 65% with mild tricuspid regurgitation. However, there was no pulmonary hypertension or regional wall abnormalities. Electrocardiograph (EKG) showed normal sinus rhythm and labs were significant for low hemoglobin of 10.9 but the complete blood count was otherwise normal.

The patient presented to Labor and Delivery at 37 weeks gestational age and 4 days in active labor with regular contractions. Standard monitors consisting of non-invasive blood pressure, EKG, heart rate, pulse oximetry were placed on the patient and supplemental oxygen via facemask was provided. At 5-cm cervical dilation, the patient received a lumbar epidural at L4-5 for labor analgesia, per her request, which was placed smoothly without complications. After confirming a normal test dose response, the epidural was activated with an infusion of bupivacaine 0.125% at 10 ml/hr. Patient remained hemodynamically stable throughout the labor process with stable vitals signs and no symptoms attributable to Scimitar syndrome, such as heart failure or dyspnea. Patient had a normal spontaneous vaginal delivery of a neonate with Apgar score of 8 at 1 minute and 9 at 5 minutes. Her postpartum course was uncomplicated, and she was discharged home on postpartum day 2 with a cardiology appointment follow-up as an outpatient.

Discussion

The incidence of Scimitar syndrome is about 1 to 3 per 100,000 live births.[5] The infantile group of patients with Scimitar syndrome typically develops symptoms soon after birth, frequently with a complicated course including severe dyspnea, respiratory failure and cardiac failure likely secondary to pulmonary hypertension. In contrast, the adult form of Scimitar syndrome is typically either found incidentally on chest radiograph or during work up for fatigue, recurrent respiratory infections or dyspnea. Radiographic findings include a Scimitar sign, a curved, tubular opacity descending alongside the right cardiac border, however, it may be obscured due to dextrocardia.[6] Opacity of the right hemothorax, right pulmonary

hypoplasia with occasional mediastinal shift towards the affected side, and other varying cardiac anomalies can be seen radiographically in patients with Scimitar syndrome. Echocardiography, three-dimensional computed tomography, cardiac gated magnetic resonance imaging, 3-D contrast enhanced MR angiography and cardiac angiography are useful tests that allow better visualization of the vessels to confirm the diagnosis of Scimitar syndrome.

Anesthetic management of patients with Scimitar syndrome is dependent upon the severity of disease and effects on cardiac and respiratory function. Prior to surgery, a comprehensive evaluation of the patient should be performed, including a thorough physical exam of the heart and respiratory function, and ancillary studies such as a complete blood count (CBC), arterial blood gas, blood typing, chest radiograph, and echocardiogram. Our patient was diagnosed with Scimitar syndrome incidentally as an adult, and did not develop significant cardiac or respiratory abnormalities during her pregnancy. Thus, routine follow up without any treatment was recommended.[7]

Patients with Scimitar syndrome may have symptoms or imaging suggestive of pulmonary hypertension. In these patients, the anesthetic management should emphasize prevention of increased pulmonary vascular resistance (PVR) by avoidance of hypoxemia, pain, acidosis and hypercarbia.[8] Consideration of epidural or spinal-epidural anesthesia in delivery patients to reduce labor pain. One of the goals in controlling pain is to prevent catecholamine release which contributes to the elevation in pulmonary vascular resistance. In some cases, pulmonary hypertension is diagnosed postoperatively, and ICU care may also be needed.[9]

Approximately 70% of patients with Scimitar syndrome also have atrial septal defect. In these patients, special caution is needed to avoid air in IV lines, tubings, or stopcocks.

Conclusion

We presented a 29-year-old G2P1 female with Scimitar syndrome who underwent successful lumbar epidural placement for analgesia for vaginal delivery. It is important to recognize that patients with Scimitar syndrome can develop pulmonary hypertension or hypertensive crisis during the pregnancy, labor and delivery. These patients should be monitored appropriately for symptoms including but not limited to cardiac failure, dyspnea, cyanosis, palpitations, syncope, or respiratory distress.

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Submitted June 19, 2019