Scimitar syndrome in pregnancy

INTRODUCTION

Scimitar syndrome is a rare complex congenital cardiopulmonary anomaly characterised by a partial anomalous pulmonary venous return (PAPVR) into the inferior vena cava (IVC), associated with right lung hypoplasia, cardiac dextroposition and a systemic right pulmonary arterial supply. Other congenital cardio-pulmonary abnormalities may coexist. It may present in early infancy with marked pulmonary hypertension (PHT) and right heart failure, or in adult life, as an incidental finding in asymptomatic patients.

We present the case of a young primigravida with Scimitar syndrome. Our literature search could identify only two previously reported cases, neither commenting on the anaesthetic management.[1,2]

CASE REPORT

The present case is about a 19-year-old primigravida with Scimitar syndrome who presented for antenatal counselling at 8-week gestation. At the time of diagnosis of Scimitar syndrome, she had mild shortness of breath. Auscultation revealed markedly reduced breath sounds over the right lung and her chest radiograph showed the 'Scimitar sign'. Echocardiography was unremarkable, with normal pulmonary arterial pressures (PAP). Magnetic resonance imaging (MRI) and contrast enhanced computerised tomography (CT) confirmed the scimitar syndrome.

A multidisciplinary team consisting of an obstetrician, a cardiologist and an anaesthesiologist closely monitored her during the course of her pregnancy. Cardiac function was assessed by echocardiograms every month. At 29-week gestation, increased breathlessness on exertion was attributed to reduced pulmonary reserve from pulmonary hypoplasia and increasing gestation, since PAP was normal, with no signs of right heart failure. A decision to give a trial of vaginal birth was agreed upon, as a planned caesarean section at that stage was not deemed necessary.

She presented in spontaneous labour at 39-weeks gestation and was slightly more breathless than before, but PAP and cardiac function remained unchanged. A lumbar epidural was sited at 3 cm cervical dilatation for labour analgesia. A patient-controlled epidural analgesia regime using a solution of 0.1% levobupivacaine and 2 mcg/ml of fentanyl was administered, providing 7 ml by continuous background infusion, with 5 ml bolus and a lock out interval of 20-min, allowing a maximum dose of 22 ml/h. Saline 0.9% was cautiously infused intravenously for maintenance of preload. Frequent clinical examination for signs of heart failure, with vital signs and SpO_2 monitoring were performed during labour. Invasive monitoring was not required. She laboured uneventfully to full cervical dilation within 5 h. Forceps were applied to reduce cardiovascular stress and cut short the second stage of labour and a live, male infant was delivered. The third stage was managed with 10 units intramuscular oxytocin. No signs of cardiac failure were noted in the peripartum period. Her post-operative recovery was uneventful and she was discharged from hospital after 3 days.

DISCUSSION

PAPVR is an uncommon anomaly accounting for 0.5-1% of all congenital heart diseases.[3] The characteristic abnormality is PAPVR (partial or complete) of the right lung to the IVC, either at the junction of right

atrium or below the level of diaphragm. Drainage into the right or left atria, superior vena cava, azygous, hepatic or portal vein can also occur.[3,4] Atrial septal defects are the most common anomaly with PAPVR, but other congenital heart defects have also been described.[5] Scimitar syndrome is a rare complex variant of PAPVR (3-5%), occurring in 1-3/100000 live births and consists of PAPVR, hypoplasia of the right lung and pulmonary artery, dextroposition of the heart, with right pulmonary arterial supply from the systemic circulation.[6-8] Mostly, the pulmonary vein from the right lower lobe and occasionally the right middle lobe forms an abnormal 'Scimitar' vein that drains into the upper IVC below the level of the diaphragm.[7] Consequently, a left to right shunt is established, with risk of right heart failure due to right ventricular volume overload.[3]

Three forms of Scimitar syndrome are described. In the infantile form, patients become symptomatic early in life, developing severe PHT and cardiac failure, with cyanosis and failure to thrive at presentation. In the adult form, most patients are asymptomatic but the extent of symptoms depends upon the degree of shunting and the number of anomalous veins, associated valvular abnormalities and the presence of other concomitant cardiac or pulmonary disease.[3] Recurrent pneumonia, mild dyspnoea on exertion and occasional haemoptysis are the most commonly reported symptoms. PHT is rare or mild and not associated with right heart failure when present.[6] Scimitar syndrome in these patients is often discovered incidentally following a chest radiograph. The third form is associated with complex cardiac malformations like hypoplastic left ventricle or aortic arch obstruction, which are far more significant clinically. Some previously fit adults can present with infant form complications of PHT and right heart failure later in life.[7] Previously asymptomatic pregnant patients may present with symptoms of volume overload due to the physiological increase of around 40% in blood volume by term; auto transfusion of 300-500 ml blood during each contraction in labour or iatrogenic fluid overload in the peripartum period.

Diagnosis is suspected on a chest radiograph showing a curved vascular shadow of the anomalous pulmonary vein descending towards the diaphragm along the right side of the heart (scimitar sign) [Figure 1], along with lung hypoplasia and cardiac dextroposition.[9] A transthoracic echocardiogram, MRI, contrast enhanced CT [Figure 2] or cardiac catheterisation are more diagnostic.

Surgical correction in adults should only be considered in symptomatic patients or in asymptomatic patients with a pulmonary to systemic shunt blood flow ratio exceeding 1.5:1 due to their higher likelihood of developing PHT and right ventricular failure.[7] In our patient, a conservative approach was deemed best, given her good functional capacity.

In the presence of significant PHT, anaesthetic management should focus on prevention of increased pulmonary vascular resistance (PVR) by avoidance of pain, hypoxemia, hypercarbia and acidosis. Fluid management may be difficult, as both hypovolemia and hypervolemia can prove detrimental. Crystalloids or colloids are both acceptable, but unmonitored fluid challenges may worsen right ventricular function and are not recommended.[10] Myocardial depression or

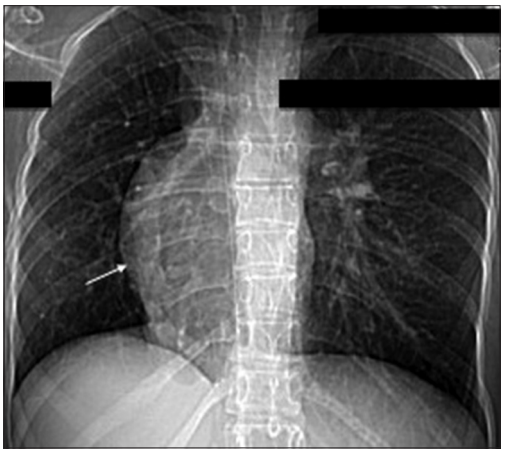

Figure 1: 'Scimitar sign' on the chest radiograph, marked by an arrow, refers to the curvilinear 'scimitar' sword like shadow of the descending anomalous pulmonary vein, the tip pointing inferomedially to the diaphragm/right heart border junction and widening inferiorly

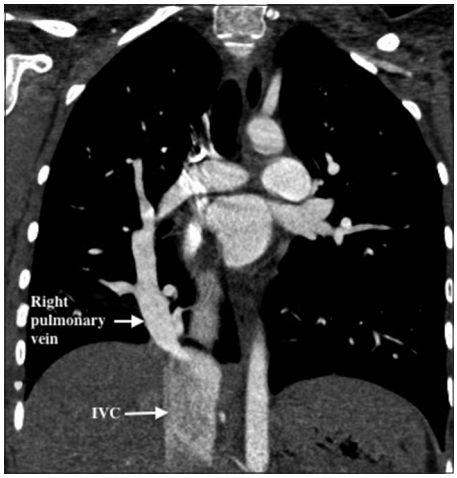

Figure 2: Contrast enhanced computerised tomography of the patient, showing the anomalous right pulmonary 'scimitar' vein draining into the inferior vena cava

marked reduction in systemic vascular resistance (SVR) and venous return should also be avoided and consequently epidural anaesthesia with incremental doses or a low-dose combined spinal-epidural anaesthesia may be preferable over spinal or general anaesthesia. By reducing pain in labour, epidural anaesthesia prevents catecholamine release and a consequent increase in heart rate and SVR, and may be better tolerated than Entonox for labour analgesia. This is because nitrous oxide increases PVR and may worsen pre-existing PHT. Furthermore, use of vasoactive drugs like phenylephrine or ephedrine and uterotonic agents like oxytocin or ergometrine should be cautious in order to prevent acute haemodynamic effects. Since the cardiac status in this patient was well-maintained throughout pregnancy, a cautiously extended epidural or a combined spinal-epidural was planned for any surgical intervention.

CONCLUSION

Anaesthetic management of patients with Scimitar syndrome for obstetric interventions should be guided by the mode of delivery planned; the degree of arterial-venous shunt present; the severity of PHT and adequacy of ventricular function. An epidural or a low dose combined spinal-epidural may be the technique of choice for obstetric surgical interventions or for labour analgesia in these patients.

It would be prudent to manage a parturient with deteriorating or complex pathology in a centre where facilities and expertise in dealing with adult congenital heart disease are available.

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Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/0019-5049.130832