Pediatric Anesthesia and Critical Care Journal 2016;4(1):43-47
doi:10.14587/paccj.2016.9

PACCJ

Pneumonectomy in Scimitar Syndrome: anaesthesia management

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Key points

Pneumonectomy in a patient with Scimitar syndrome poses unique challenges for anaesthesiologist throughout the perioperative period. Associated congenital cardiac anomalies, difficulty in selection of type of endotracheal tube for one lung ventilation in paediatric patients, provision of lung protective ventilation strategies and adequate analgesia, prevention of Pulmonary hypertension should be taken in to consideration.

Abstract

Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies consisting of a partial anomalous pulmonary venous connection of the right lung (part or the entire right lung) to the inferior vena cava, right lung hypoplasia, dextroposition of the heart, and anomalous systemic arterial supply to the right lung. Surgical treatment includes either ligation of anomalous arteries and the scimitar vein implanted into left atrium or resection of sequestrated or chronically infected lung. Anaesthesia management of patients with Scimitar syndrome is challenging due to coexisting congenital heart disease. The literature on anaesthesia management of patients with Scimitar syndrome undergoing Pneumonectomy is rare and hence authors wish to share their experience of anaesthesia management of a case of Scimitar syndrome posted for Right Pneumonectomy in view of infected and non-functioning lung.

Keywords: Scimitar Syndrome, Pneumonectomy

Introduction

The term scimitar syndrome was coined by NEILL et al. in 1960, describing a syndrome of partial anomalous pulmonary venous drainage of the right lung into the inferior vena cava, partial systemic arterial blood supply, and hypoplasia of the affected lung, with bronchial abnormalities and abnormal lobation. This is a rare anomaly with an incidence of approximately 1 to 3 per 100,000 live births[1,2,3,4,5,6,7]

The term scimitar syndrome was coined because of the radiographic appearance of the anomalous vein, which appears as a tubular opacity paralleling the right cardiac border resembling a curved Turkish sword or scimitar (scimitar sign)[3, 8]. It is also called mirror image lung syndrome, hypo genetic lung syndrome, Halasz syndrome.Clinical presentation is of two types. Adult form and Infantile form with the infantile form presenting as severe pulmonary hypertension, cardiac failure, and a high mortality rate. Surgical intervention is required if there is a large left/right shunt exceeding 50%, resulting in pulmonary hypertension, heart failure or when there is lung sequestration and/or recurrent right-sided chest infections.

Case report

2.5 years old female child weighing 10kg, diagnosed

case of scimitar syndrome presented with episodes of recurrent cough and cold since birth. Last episode of LRTI was one month prior to admission. Her birth history was uneventful. Clinical examination revealed a playful, afebrile child having peripheral cyanosis with on air saturation of 98%.There was no evidence of any other external congenital anomaly. Respiratory system examination revealed markedly reduced breath sounds over the right lung. Heart sounds were heard on right side. An ejection systolic murmur in pulmonary area was heard on cardiac examination. Routine laboratory investigations revealed normal haemogram and coagulation profile. Chest X-ray showed cardiac silhouette shifted to right with right upper lobe opacity and the characteristic saber shape of the lung i.e. ′Scimitar sign′. On further workup, echocardiography revealed hypo plastic right lung, SDS, dextrocardia, PAPVC, 2mm atrial septal defect, right pulmonary vein draining into suprahepatic IVC with a gradient of 20mm of Hg, hypo plastic RPA (4.4mm) and on air saturation of 94% with mild pulmonary hypertension (PHT). CT Pulmonary Angiography revealed hypo plastic right lung with severe hypoplasia of RML, RML bronchus, hemi thoracic volume loss, dextroposition of the heart and compensatory hyperinflation of left lung, an arterial collateral arising from descending aorta was seen supplying the posterior basal segment of lower lobe of the right lung Ventilation/Perfusion scan of the lung showed no tracer uptake in right lung suggestive of hypo plastic/congenital absent lung with normally perfused left lung. In the preoperative holding area fentanyl 20 µg and midazolam 0.2 mg was administered intravenously. She was then induced in the Operation theatre with Propofol 20mg and vecuronium 2mg as muscle relaxant after confirming mask ventilation. The trachea was intubated with 4.5mm endotracheal tube and fixed at mark 12 after confirming adequate air entry. Left Femoral arterial (22g) and right internal jugular vein (5.5F) with triple lumen catheter were cannulated and the patient was positioned in left lateral position for epidural catheter insertion. 19G epidural catheter was inserted at T8-9 level and catheter was fixed at mark 7 with loss of resistance to saline technique. After negative aspiration for blood and CSF and test dose of 1.5ml adrenalized lignocaine, 3ml of 0.25% bupivacaine with 30mcg Buprenorphine (0.04ml/kg x number of segments to be blocked) was given and repeated every two hourly for intraoperative analgesia. Anaesthesia maintenance was carried out using mixture of air and oxygen (50:50) %, infusion of Propofol (4-6mg/kg/hr) and vecuronium (0.08mg/kg/hr). The post induction hemodynamic parameters were: systolic blood pressure ranging from 80-100 mmHg, heart rate 120-140/min and PaO2 >100 mm Hg and PaCO2 < 40 mm Hg on FiO2 0.5. We decided to ventilate the child using Pressure control mode with peak airway pressures of 8 cm H_2O delivering a tidal volume of 50 to 60 ml with PEEP of 4cm of H_2O to prevent barotrauma to left lung. Monitoring included electrocardiogram, blood pressure (Invasive, and non-invasive), pulse-oximetry (SpO2) and $ETCO_2$. The surgery lasted for about six hours and throughout the procedure there was no episode of hypercapnia or hypoxia. Ringer lactate was infused (4 ml/kg/hr) to maintain CVP 6 – 8 cm of H_2O and urine output 0.5-1ml/kg/hr. 120ml blood was given to replace blood loss. Right pneumonectomy was performed through a right 5th intercostal posterolateral thoracotomy incision. During the surgery scimitar vein and the additional right pulmonary vein were each ligated and divided. The right main stem bronchus was stapled and divided. At the end of surgery, residual neuromuscular blockade was reversed, patient was extubated comfortably. Post-operative analgesia was maintained using 3ml of 0.125% Bupivacaine with 30mcg Buprenorphine 8 hourly for 72hrs. The patient had a smooth postoperative course. She remained free of pain, bronchospasm and was discharged home on the 14th postoperative day (see Figures 1, 2, 3, 4)

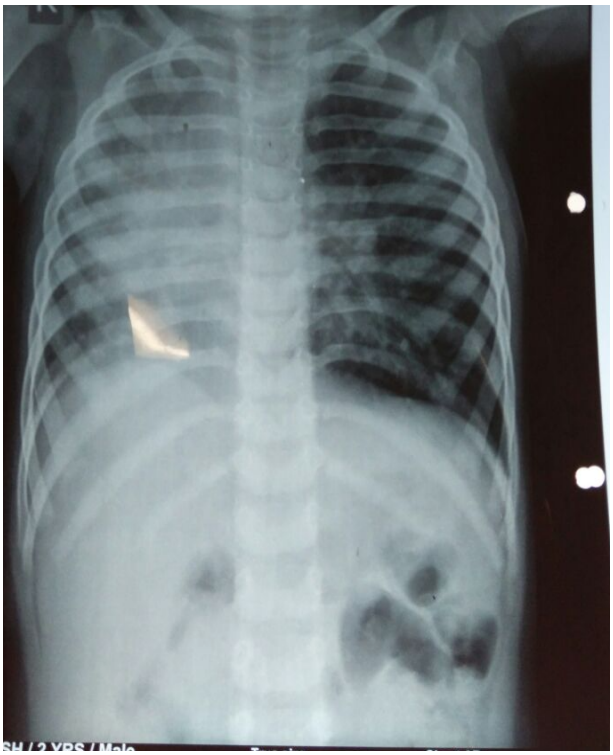

Figure 1. X ray chest PA view

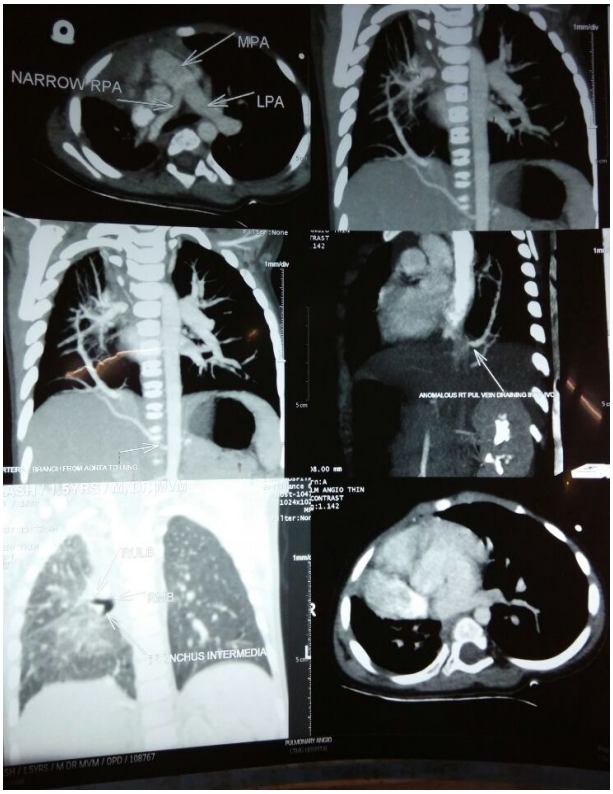

Figure 2. CT Pulmonary angiography

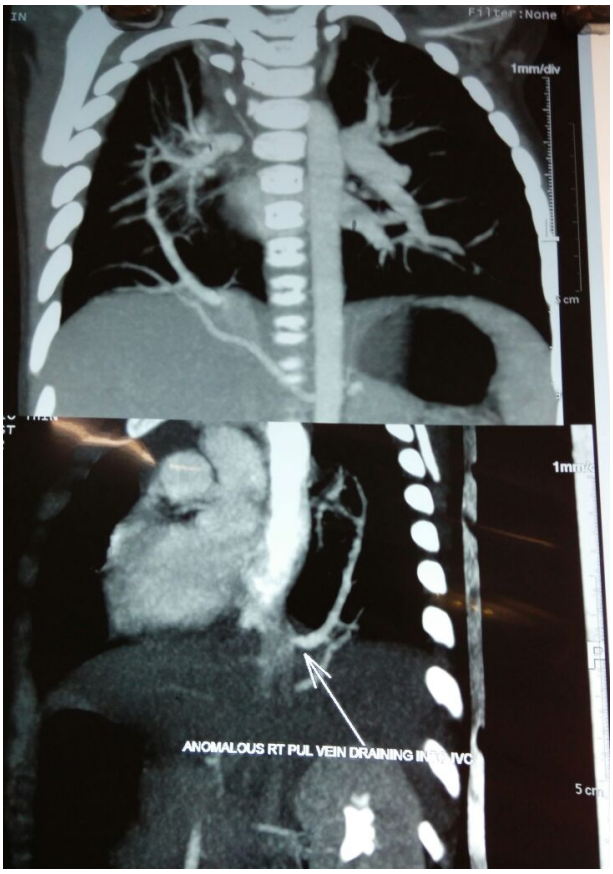

Figure 3. CT Pulmonary angiography

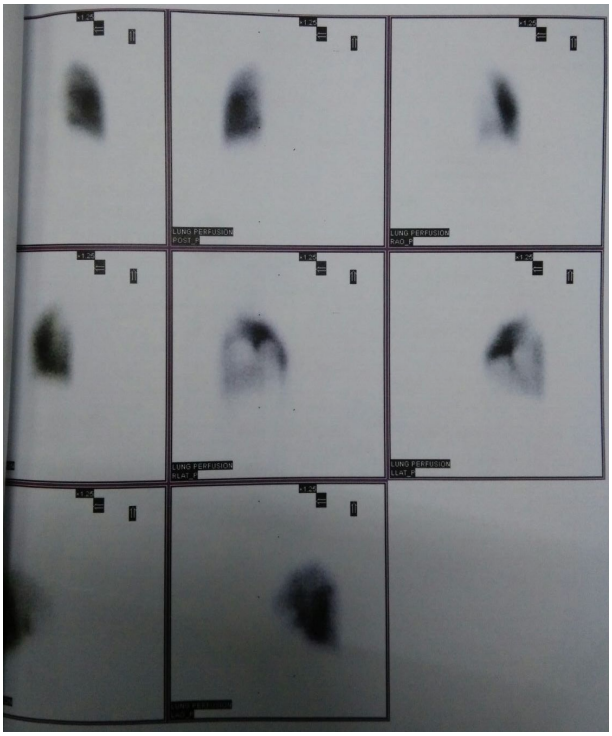

Figure 4. Ventilation-Perfusion scan

Discussion

The scimitar syndrome is part of a congenital disorder called as partial anomalous pulmonary venous connection (3–5% of all PAPVC).[7] Decision to perform a right pneumonectomy was taken in our patient due to presence of a single pulmonary vein draining right lung with aberrant arterial supply, hypo plastic right lung and due to recurrent lower respiratory tract infection to prevent other lung from getting infected. F.M.N.H. Schramel etal[7] in their study of seven patients with scimitar syndrome reported that isolated ligation and reimplantation of the anomalous vein was unsuccessful due to thrombosis and anastomotic stenosis leading to lung infarction and ultimately requiring pneumonectomy. Inhalation induction is less effective due to the pulmonary hypoplasia and may cause myocardial depression. Hence, we induced our patient with Propofol because of its bronchodilator properties. Endotracheal tube of size 4.5 was inserted. We decided against left endobronchial intubation as the right lung was hypo plastic and had no perfusion. The goals of anaesthetic management in PAPVC are maintenance of systemic vascular resistance and reduction in pulmonary vascular resistance. Our patient had mild pulmonary artery hypertension and we were vigilant to avoid any factor leading to increase in Pulmonary artery pressure (PAP) like hypoxemia, hypercarbia, acidosis and we put an epidural for effective analgesia[9]. Right ventricular dysfunction can occurs after pneumonectomy due to an increase in pulmonary artery pressure and pulmonary vascular resistance. Thoracic epidural analgesia helps to decrease PAP, postoperative respiratory complications, arrhythmias and allows early extubation. Hence, considered gold standard method of analgesia in lung surgeries[10]. Lung protective ventilation strategies include low tidal volumes, higher rate and use of pressure mode in paediatric patient[8]. We, also used Pressure control mode with peak airway pressures of 8 cm of H_2O and PEEP of 4 cm of H_2O to prevent barotrauma to the left lung. Fluid management in PAPVC may be difficult due to existing left to right shunt leading to right ventricular volume overload. Also, excessive fluid administration in patients undergoing thoracic surgery has been found to be an independent risk factor for acute lung injury. Crystalloids and colloids are both acceptable, but unmonitored fluid challenges may worsen right ventricular function and hence not recommended[10]. Because of the above strategies and because of the fact that there were no episodes of Hypoxia, hypercarbia, hemodynamic instability, we could extubate this patient on table. ABG post extubation was good and pain relief was adequate.

Conclusion

Scimitar syndrome is a rare but well described disorder which is associated with a range of congenital cardiac and pulmonary anomalies. The anesthetic goals should be focused on the management of pulmonary hypertension, lung protective ventilation, postoperative analgesia and prevention of postoperative pulmonary complications.

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