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Anaesthesia Considerations in Scimitar Syndrome

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Abstract

Scimitar syndrome is a rare congenital heart disorder characterised by abnormal drainage of pulmonary veins into the inferior vena cava, associated with hypoplasia of the right lung, pulmonary artery, atrial septal defects and other congenital cardiac anomalies. Scimitar syndrome has a very low prevalence in the population but it has a very high perioperative morbidity and mortality due to pulmonary artery hypertension and its sequel. Hence understanding the anaesthetic implications becomes important. This study presents the anaesthetic management of an 11-year-old case of scimitar syndrome planned for adenotonsillectomy under general anaesthesia. After a complete evaluation of the present cardiopulmonary status and obtaining consent from the guardians, the child was taken up for surgery. Perioperatively, all the factors that precipitate pulmonary artery hypertension like hypovolemia, pain, hypoxia, hypothermia, hypercarbia and metabolic acidosis were avoided. Intermittent positive pressure ventilation with low tidal volumes for the management of any underlying lung hypoplasia was maintained. Vasoconstrictors like noradrenaline and inotropes like dobutamine and milrinone were readily available in the event of any untoward cardiac complications. Haemodynamic parameters were stable intraoperatively. Adequate recovery was ensured after which tracheal extubation was performed followed by an uneventful post-operative period.

Keywords: Adenotonsillectomy, General Anaesthesia, Partial Anomalous Pulmonary Venous Connection, Pulmonary Arterial Hypertension, Scimitar Syndrome

1. Introduction

Scimitar syndrome is a rare congenital heart disorder with an incidence of 1 to 3 per 100,000 live births. Cardiac anomalies may also include atrial and ventricular septal defect, patent ductus arteriosus, subaortic stenosis, coarctation of the aorta, pulmonary lobar and respiratory tract anomalies[1].

There are three forms of scimitar syndromes. The 'adult' form, is of a mild variety without pulmonary hypertension, requiring no treatment. The 'Infantile' form presents with severe pulmonary hypertension generally requiring surgery. The third form is associated with complex cardiac malformations like hypoplastic left ventricle or aortic arch obstruction[2]. This report throws light on the major perioperative concerns and their management which includes pulmonary artery hypertension, intraoperative ventilation strategies for managing associated lung hypoplasia and post-operative right ventricular dysfunction.

2. Case Report

An 11-year-old female patient weighing 30kg, diagnosed with adenotonsillitis was admitted for adenotonsillectomy. On pre-anaesthetic evaluation, it was noted that the child was a known case of scimitar syndrome diagnosed at the age of 5 and was advised regular follow-up. The patient had a history of repeated

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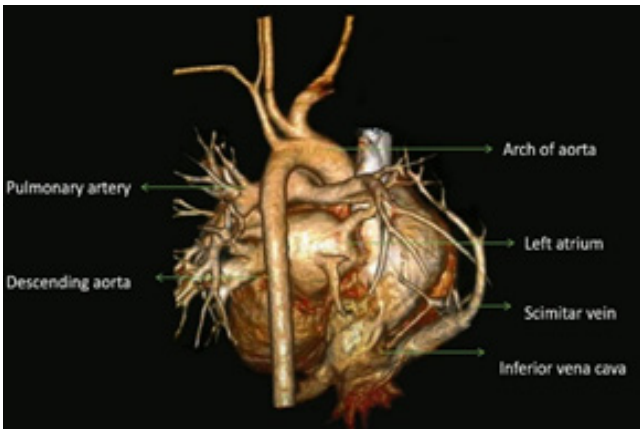

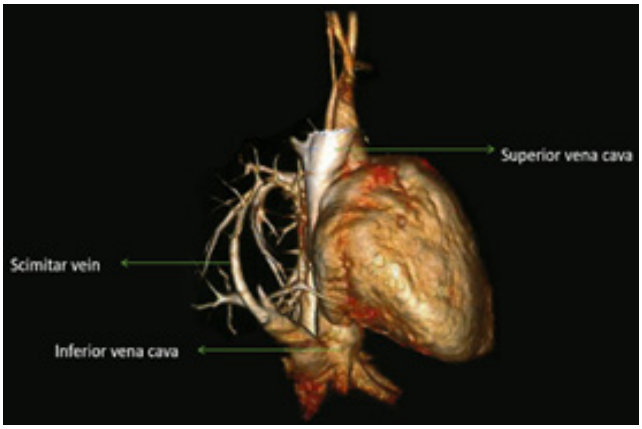

Figure 1 and 2. CT angiography shows the right upper pulmonary vein draining into the inferior vena cava.

respiratory infection and dyspnea on exertion. On examination, she was averagely built and nourished and had attained normal milestones for her age. Her vitals parameters showed a pulse of 110bpm regular, blood pressure of 100/60mm Hg and auscultation of lungs was unremarkable. The apex beat was at the 5th intercostal space and shifted 3cm medial to the left midclavicular line. All blood investigations were within normal limits. Airway examination revealed adequate mouth opening with Mallampati class II, bilateral tonsillar hypertrophy and neck movements were adequate. Due to exertional breathlessness, she was referred to a cardiologist for evaluation of her present cardiopulmonary status. Echocardiography revealed mesocardia, anomalous pulmonary venous return, ejection fraction of 66%, Pulmonary Artery Systolic Pressure (PASP) of 15mm Hg, normal biventricular function and no intracardiac shunt lesion were noted. Arterial blood gas was within normal limits. CT angiography showed right upper pulmonary vein draining into the inferior vena cava (Figures 1 and 2).

After a complete re-evaluation of the present cardiopulmonary status, the patient was taken up for adenotonsillectomy under general anaesthesia. Tablet Alprazolam 0.25mg was given the night before the surgery. After obtaining assent from the patient and consent from the parents, the patient was shifted to the operation theatre. Standard American Society of Anaesthesiologists (ASA) monitors like Electrocardiogram (ECG), noninvasive blood pressure, pulse oximeter and temperature probe were connected. Baseline vital parameters were noted. The intravenous line was secured with a 20G cannula and ringer lactate was started at 350 ml/hr for the first hour followed by 210 ml/hr for the second hour. Intravenous lines were meticulously de-aired to avoid entry of air bubbles into the systemic circulation and the same was taken care of during drug administration. The patient was preoxygenated with 100% oxygen for 3 minutes. After premedication with IV ondansetron 3mg and midazolam 1mg, opioid anaesthesia induction was done with IV fentanyl 120mcg and succinylcholine 60mg IV was used to facilitate orotracheal intubation with a size 6 cuffed endotracheal tube. Bilateral air entry was confirmed, the tube was fixed and throat packing was done. Anaesthesia was maintained with Intermittent Positive Pressure Ventilation (IPPV) with low tidal volume using oxygen in the air (FiO2-50%) sevoflurane at 1 MAC and intermittent injection of vecuronium was given for muscle relaxation and fentanyl 30mcg was repeated for intraoperative analgesia. Haemodynamic parameters were stable intraoperatively. Residual neuromuscular blockade was reversed with IV Neostigmine 1.5mg and glycopyrrolate 0.3mg. The trachea was extubated after adequate recovery was ensured and the post-operative period was uneventful.

3. Discussion

The major concerns with scimitar syndrome include the presence of pulmonary artery hypertension with or without atrial and ventricular septal defect, coarctation of the aorta, hypoplasia of the right bronchus, lung and right pulmonary artery and other congenital cardiopulmonary malformations[1]. Routine investigations

including Complete Blood Count (CBC), Arterial Blood Gas (ABG), chest radiograph and ECG among others help assess and optimise the clinical condition of these patients. Intraoperative monitoring should include ECG, oxygen saturation, ABG, capnography, temperature, urinary output and central venous pressure measurements[3]. Invasive blood pressure monitors are instituted depending on the clinical condition of the patient.

Intravenous induction with an opioid and a nondepolarising muscle relaxant can be used. Goal-directed fluid management is mandatory as hypotension can be detrimental in these cases. Wherever needed, analgesic management using regional anaesthesia techniques or intravenous opioids is preferred[4]. Perioperatively, all factors that precipitate pulmonary artery hypertension should be avoided which include hypovolemia, pain and any kind of stimulus to the sympathetic nervous system, hypoxia, hypothermia, hypercarbia and metabolic acidosis[4]. Atelectasis, high airway pressure and the use of nitrous oxide should be avoided as these factors influence the increase in pulmonary vascular resistance. Measures should be taken to prevent potential air embolism by strict removal of air bubbles from IV lines and stopcock if Atrial Septal Defect (ASD) is suspected[5]. In the presence of underlying lung hypoplasia, pressure-controlled mechanical ventilation with low tidal volume (4-8 ml/kg) and plateau pressure < 30 cmH_20 is recommended to avoid volutrauma[3].

Perioperative management aims at selecting an appropriate plan of anaesthesia, ensuring adequate depth and appropriate analgesia, avoiding an increase in pulmonary artery pressure, and optimising cardiac function with normal systemic blood pressure for age. In cases where there is right ventricular dysfunction, dobutamine and milrinone are considered inotropes of choice[4].

If pulmonary artery hypertension occurs, it should be treated with hyperventilation with 100% oxygen along with pulmonary vasodilators like nitric oxide and correcting metabolic and respiratory acidosis is beneficial[4]. For the management of hypotension, vasoconstrictors like noradrenaline can be initiated. Inotropic agents like dobutamine and milrinone help in increasing the inotropic function of the heart and simultaneously reducing pulmonary vascular resistance[3]. In some cases, pulmonary hypertension is developed postoperatively, and in such cases, ICU care and mechanical ventilation are required[5]. As this condition is similar to an 'adult' form of scimitar syndrome with no comorbidities, the case was successfully managed under general anaesthesia. No incidence of Pulmonary Hypertension (PAH) or systemic hypotension was noted and adequate post-operative analgesia was ensured.

4. Conclusion

Scimitar syndrome is a rare congenital disorder that is associated with a wide range of cardiac and pulmonary anomalies. In the absence of any comorbidities, anaesthetic management includes prevention and/or management of PAH and avoiding intraoperative and postoperative hypotension. Adequate analgesia in the postoperative period is essential to prevent any untoward cardiac events such as right ventricular dysfunction.

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