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Anaesthesia and orphan disease:Anaesthetic management of a child with Scimitar syndrome scheduled for release of temporomandibular joint ankylosis

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ABSTRACT

Scimitar syndrome is a relatively rarecongenital heart defect with a complex combination of cardio pulmonary abnormalities characterized mainly by right lung hypoplasia and partial anomalous pulmonary venous return. We report a ten years old female child diagnosed with Scimitar syndrome at the age of four years with recurrent chestinfections, bilateral temporomandibular joint ankylosis,mandibular hypoplasia and a grossly restricted mouthopening for release of ankylosis. Anaesthetic management of thispatient was done successfully with use of fibreoptic bronchoscopyfor securing the airway.

KEYWORDS Scimitar syndrome, Partial anomalous pulmonary venous return, Right lung hypoplasia, temporomandibular joint ankylosis, Mandibular hypoplasia

INTRODUCTION

Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies consisting of a partial anomalous pulmonary venous connection of the right lung to the inferior vena cava, right lung hypoplasia, dextroposition of the heart, and anomalous systemic arterial supply to the right lung.[1, 2, 6]The first anatomical observation of this anomalous pulmonary venous return was made by George Cooper an English pathologist andRaoul Chassinat from France separatelyin 1836.[3] Scimitar syndrome accounts for 0.5 to 1 % of congenital heart diseases. This rare anomaly has an incidence of approximately 1 to 3 per 100,000 live births.14However, the actual prevalence rate is likely to be higher because in many cases the patients remain asymptomatic for most of their lives.The term was first given by Dr Catherine Neill et al in 1960.[8] Females are more likely to develop the syndrome compared to males.[10]The hallmark of Scimitar syndrome is an anomalous right pulmonary vein that drains part or all of the right lung into the inferior vena cava .[8, 16, 27] 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 described by Halasz and colleagues in 1956.[11]This is the so-called Scimitar sign. In adults and older children it is usually detected during an investigation for dyspnoea, fatigue, recurrent respiratory infections, or as an incidental finding on a routine chest radiograph. This adult form of Scimitar syndrome usually is not associated with pulmonary hypertension and typically has mild symptoms and a benign prognosis.[11, 12] The infantile group of patients are symptomatic soon after birth,develop severe pulmonary hypertension and cardiac failure, making management difficult and the mortality is high.[11, 12] Haemodynamically, there is an acyanotic left to right shunt. The anomalous vein usually drains into the inferior vena cava most commonly or into theright atrium, superior vena cava, the azygos system, the hepatic vein or portal vein.[8,16,,18,19,20,27]

The scimitar syndromeforms about 3–5% of all PAPVC (partial anomalous pulmonary venous connection).[21, 22] About 70% of these patients with scimitar syndrome have an associated atrial septal defect.[23]The other associations with this syndrome areventricular septal defect (VSD)29,patent ductus arteriosus (PDA),[8,25,26] Tetralogy of Fallot,[1, 25] coarctation of aorta,[30,31,32,] hypoplastic left heart syndrome,[8] total anomalous pulmonary venous connection,[30]cor triatriatum,[12] bicuspid aortic valve, [26, 31] and subaortic stenosis.[30] Overall, 19% to 31% of patients with scimitar syndrome have associated cardiac anomalies.[1,26,28,34,35,] Other associations include psilateraldiaphragmatic anomalies, accessory diaphragm, diaphragmatic hernia,localised bronchiectasis, bronchogenic cysts, horseshoe lung,vertebral anomalies like hemivertebrae and genitourinary tract abnormalities.[28,37,38] Surgical correction should be considered in the presence of significant left to right shunting and pulmonary hypertension.[17,39]This involves creation of an inter-atrial baffle to redirect the pulmonary venous return into the left atrium. Alternatively, the anomalous vein can be re-implanted directly into the left atrium.[12, 34,40]

Ankylosis of the temporomandibular joint (TMJ) most often results from trauma or infection, idiopathic or iatrogenic, but it may be congenital or a result of Rheumatoid Arthritis .[9] Chronic painless limitation of motion occurs. This interferes in speech, mastication, maintenance of orodental hygiene, aesthetic and psychosocial problems.[9, 41] It may be a life threatening problem to establish airway in an emergency. Children with bilateral temporomandibular joint ankylosis tend to develop secondary mandibular hypoplasia.[9,41] Whenankylosis occurs in children while the jaws are still in phase of growth it will cause restriction of growth of mandible. The bird face deformity is responsible for obstructive sleep apnoea in these patients.[9,41]

CASE REPORT

A ten years old girl presented to the paediatric out patient department at our hospital with history of recurrent chest infections and inability to open her mouth.She had restricted movements of jaws,inadequate chewing,restricted mouth opening,and problems in speech,poor maintenance of oro dental hygiene,malaligned teeth,and emotional,social and psychological disturbances. Her main complaints were related to her jaw movements and then on elicitation her parent revealed that she suffered from recurrent respiratory infections since early childhood. Presently child had no history of cough,fever, chest pain, palpitations or haemoptysis. The child had a weight of 20 kgs and height of 130 cm. Her heart rate was 80 per minute and regular, blood pressure was 90/60 mmhg and respiratory rate was 18 per minute. Her exercise tolerance was good and she had a MET value of 8. Chest radiography showed a hypoplastic right lung and an emphysematous left lung, rightward shift of trachea, mediastinum and heart.ECG revealed right axis deviation. Echocardiogram revealed moderately elevated pulmonary artery systolic pressure (44 mmHg),a mildly dilated right atrium and a hypoplasticright pulmonary artery.Computed tomographyscan of chest showed the right pulmonary vein(a large scimitar vein)draining into the inferior vena cava below the diaphragm and dextroposition of the heart. There was no other congenital anomaly.Pulmonary function tests showed moderate obstruction.She was referred to the oromaxillo facial surgery

department for further management.

After obtaining an informed consent from her parents she was posted in the elective surgical listfor release of ankylosis and interpositionalarthoplasty with temporalis muscle. As for any elective surgery the child was fasted for six hours. 1 gm of Cephalexin was given intravenously an hour prior to the surgery .The child was very apprehensive and uncooperative even though she was explained why she would be operated and what would be done was the best for her. In the operating room0.5 mg of midazolam, 0.1 mgglycopyrrolate, and 2 mg of ondansetron were given intravenously.Asleep fibreopticnasotracheal intubation was planned as direct laryngoscopy was impossible with the 3 mm inter incisor distance.As a backup plan we had the ENT team for tracheostomy in case it was needed.Xylometazoline drops and 2% xylocaine viscous jelly was instilled into her nostrils. The child was nebulised with 4% xylocaine for fifteen minutes. She was oxygenated with 100% oxygen for five minutes and then inhalational induction was done with sevoflurane in 100% oxygen. Then a size 5 nasopharyngeal airway lubricated with 2% xylocaine viscous jelly was passed into her right nostril gently and a breathing circuit was connected with an endotracheal tube connector and spontaneous respiration was ensured with 100% oxygen and 5% sevoflurane. The fibreoptic bronchoscope was introduced through the left nostril and the vocal cords were visualised and it was advanced gently into the tracheal lumen to the carina. The airway was adequately anaesthetised so the child did not have any reflex response. A 5 size cuffed endotracheal tube was already passed and fixed at the proximal end of the scope. The tube was passed over the bronchoscope into the trachea just above the carina. The endotracheal tube was connected to the breathing circuit and its position was confirmed by capnography trace and bilateral chest auscultation. Intravenous 2 mg of vecuronium was given for muscle relaxation and intravenous 40 mcg fentanyl was given for analgesia. Anaesthesia was maintained with 50:50 air oxygen mixture with 1 MAC isoflurane due to its lesser effect on hypoxic pulmonary vasocontriction compared with other inhalational agents. Intermittent positive pressure ventilation was given on a closed circuit and a PEEP of 5 cm of water. Her right radial artery was cannulated and connected to a pressure transducer for blood gas sampling and continuous blood pressure monitoring. Considering her fairly reasonable MET value and being a smaller child an invasive central venous pressure line was not considered. The patient was connected to a multipara monitor displaying ECG, NIBP, IBP,$ETCO_2$, SPO_2 and core temperature. Throat pack was inserted by surgeon after surgical access to the oral cavity was established. The procedure lasted for 80 minutes andintraoperative fluid therapy was given restrictively and in a targeted manner, with adequate hemodynamic monitoring to optimize right-ventricular preload. Her vital parameters were stable throughout surgery.Her arterial blood gas values during peri operative period showed a pH of 7.4, PaO_2 of 138 mmHg and $PaCO_2$ of 30 mmHg.Low doses of a vasoconstrictor infusion; noradrenaline 2–5 µg and phenylephrine were prepared and kept available as an emergency measure but the systemic pressures were stable peri operatively.

At the end of the procedure neuromuscular blockade was reversed with neostigmine 1.0mg and glycopyrrolate 0.2 mg. The pharyngeal pack was removed and the throat was thoroughly suctioned. She was extubated awake with all protective airway reflexes and shifted to the post operative recovery area.

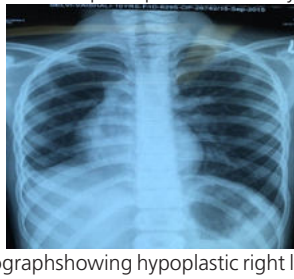

Fig1-chest radiographshowing hypoplastic right lung,dextrocardia and scimitar vein

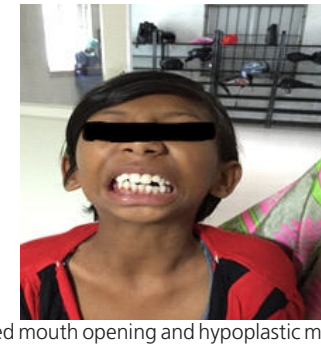

Fig 2. Restricted mouth opening and hypoplastic mandible

DISCUSSION

Anaesthetic considerations were focused on difficult airway, pulmonary hypertension and right lung hypoplasia. With a near total trismus, an uncooperative child who would not allow awake fibre optic intubation and associated Scimitar syndrome with moderate pulmonary hypertension it was more difficult to establish the airway although a standby tracheostomy set was kept in the operating room in case fibre optic guided intubation failed. Awake fibreoptic bronchoscopy is a safe method of securing airway in these patients.Tracheostomy is obviously associated with severemorbidity and mortality and longterm side effects and hence was reserved as the last option in case ofemergency.

Scimitar syndrome is a very rare congenital heart disease with a partial anomalous pulmonary venous return, pulmonary hypertension andright lung hypoplasia. At the time of surgery, the patient should be optimised in a stable condition for an extended period of time.Nitrous oxide and ketamine should be avoided as they aggravate pulmonary hypertension.Hypoxia, acidosis, hypercapnia, hypothermia should be avoided as it may also aggravate existing pulmonary hypertension. A target PaCO2 of 30–35 mmHg was achieved with moderate hypoventilation without allowing the pH value to fall below 7.4.Intraoperative fluid therapy should also be carried out restrictively in a targeted manner to optimize right-ventricular preload. Intraoperative management should ensure that depth of anesthesia and analgesia is always sufficient, as stress and pain during awareness may contribute to pulmonary vasoconstriction. An essential goal is to maintain systemic blood pressure above pulmonary arterial pressures, thereby preserving coronary blood flow.Due to hyploplastic right lung we considered use of isoflurane which has minimum effects on hypoxic pulmonary vasoconstriction. Histamine releasing muscle relaxants like atracurium and mivacuriumshould be avoided as they may further increase pulmonary resistance. Our patient recovered well and was shifted to high dependency unit after surgery for a day and was given adequate analgesics. On follow up after discharge she had a good weight gain and is also doing exercises to prevent recurrence.

CONCLUSION

Scimitar syndrome is a rare congenital heart defect.Pulmonary hypertension is a major reason for elevated perioperative morbidity and mortality, even in noncardiac surgical procedures. The associated right hypoplastic lung should be kept in mind and the patient should be evaluated for the presence of other associated anomalies. Our patient had a near total trismus and mandibular hypoplasia, was an uncooperative child and did not understand the situation and risks involved. Thoughfibreopticn-asotracheal intubation is the standard method of airway establishment an alternative option of tracheostomy should be always be kept if the first plan fails. Hypoxia, hypercapnia, hypothermia, systemic hypotension should be avoided in the peri operative period.

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