Pediatric Anesthesia and Critical Care Journal 2016;4(2):89-90
doi:10.14587/paccj.2016.18

PACCJ

A case of scimitar syndrome: anesthetic considerations regarding non cardiac surgery

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Keypoints

- Possiblity of pulmonary hypertension must be kept in mind.
- Meticulous attention should be given to monitoring, pain and fluid management
- Patient often require postop ICU care.

Abstract

Scimitar syndrome is a rare congenital heart disorder characterized by curvilinear pattern visible on the chest radiograph, resembling a sword (scimitar) created due to abnormal drainage of pulmonary veins into the inferior vena cava instead of draining into heart. We intend to report overall management of a child suffering from scimitar syndrome planned for surgical correction of rectovaginal fistula.

Keywords: congenital, anesthesia, scimitar, infant, pediatric, general anesthesia

Introduction

Scimitar syndrome is a rare congenital heart disorder.[1] Incidence is around 1 per 100000 births. This syndrome is referred to as scimitar syndrome because of a curvilinear pattern visible on the chest radiograph, resembling a sword (scimitar) created due to abnormal drainage of pulmonary veins into the inferior vena cava instead of draining into heart.[3] Other cardio-pulmonary anomalies may also be present in such patients. There are very few case reports of Scimitar syndrome. Thus we intend to report overall management of a child planned for surgical correction of rectovaginal fistula, incidentally diagnosed as scimitar syndrome during preanaesthetic checkup.

Case report

A 13 months old female child, having recto-vaginal fistula was posted for surgical correction (posterior-sagittal ano-rectoplasty). The child was of average built with normal milestones for the age. However she was tachypnoeic with decreased air entry over right side of chest. Apex beat was also shifted towards right side. Pediatric cardiologist was consulted and on further evaluation and investigating diagnosis of scimitar syndrome was made. Chest radiograph showed typical scimitar sign, echocardiography revealed cardiac dextroposition, anomalous pulmonary venous return of the right upper and lower pulmonary veins to the inferior vena cava and mild pulmonary hypertension. Patient was crying and uncooperative hence 50 mg ketamine with 0.2 mg glycopyrrolate intramuscularly was given in the preoperati-

ve area in mother's lap. Once sedated the child was carried into operation theater, and standard monitors including pulse oximetry, non-invasive blood pressure, electrocardiogram and precordial stethoscope were attached. Intravenous line was secured with 22 G intracath inside OT and baby was kept on mask. Patient was medicated with Midazolam 0.03 mg/kg and Fentanyl 2 mcg/kg. After preoxygenation with 100 % oxygen for 3 minutes; the patient was induced with 50 mcg of fentanyl and 1 mg of midazolam and relaxed with intravenous succinylcholine 20 mg.

Tracheal intubation was done with 5 uncuffed Endotracheal tube using Airtaq. Air entry on both sides confirmed, then tube was fixed with proper sticking and throat packing was done. Maintained with 60% N2O, 40% O2 and 2 % sevoflurane and intermittent injection atracurium. Temperature monitoring was done throughout the procedure via axillary temperature probe. Patient was then placed in lateral position and caudal block was given using 7ml of 0.25% bupivacaine. Patient was then placed in prone position.

Hemodynamic parameters were stable intraoperatively. Reversal of the neuromuscular blockade was done with Neostigmine 0.4 mg and Glycopyrolate 0.1 mg and adequate recovery was ensured after which trachea was extubated which was followed by an uneventful postoperative period. All due anesthetic and surgical considerations were taken into account and PSARP was done successfully.

Discussion and conclusion

Scimitar syndrome has varied presentations.[2] From asymptomatic to severe symptoms due to right lung hypoplasia, pulmonary hypertention, left to right shunt, atrial or ventricular septal defects and coarctation of the aorta etc. In adults, the syndrome is of mild variety, generally without pulmonary hypertension, treatment is not required. However, when the syndrome appears in the neonatal period, it presents as heart or respiratory failure secondary to pulmonary hypertension and generally requires surgery but may be asymptomatic[4] as in our case.

Diagnosis can be made with an echocardiogram showing the anomalous connection in 70% of cases. Three-dimensional tomography or cardiac magnetic resonance imaging are fundamental for visualizing the anomalous pulmonary vein and clarifying the patient's anatomy.

Treatment depends on age and symptomatology.

In newborns, it starts with management of the pulmonary hypertension. Surgical correction in scimitar is recommended if (a) patient having symptoms of heart failure, recurrent infectious diseases, or refractory pulmonary artery hypertension (PAH) and in (b) asymptomatic patient with significant Left-Right shunting >1.5:1. Non cardiac surgery in a case of scimitar syndrome can be performed safely however during perioperative period we have to avoid any air in IV lines, tubings, stopcocks especially in presence of ASD.

Meticulous attention should be given to monitoring, pain and fluid management. Nitrous oxide should be avoided. For pain relief, one may use caudal epidural. Intraoperative and postoperative factors that precipitate PAH such as hypoxia, hypercarbia, hypothermia, acidosis, atelectasis, high airway pressure should be avoided. If child develops PAH postoperatively, patient may need postoperative ICU care and mechanical ventilation. Weaning from ventilator must be gradual in such cases.

References

1. Elsallabi O et al. Scimitar Syndrome. J Gen Intern Med 2015 May 5.
2. Bo I et al. Variants of the scimitar syndrome. Cardiol Young 2015 Sep 16:1-7.
3. Çiçek S et al. Scimitar syndrome: the curved Turkish sabre. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2014;17:56-61.

4. Atik E. Case 6 / 2014 - Five-year old child with Scimitar syndrome and pulmonary sequestration of right lower lobe. Arq Bras Cardiol 2014;103:e45-7.