Anaesthetic management of a rare case of single ventricle heterotaxy syndrome for emergency caesarean section

INTRODUCTION

Heterotaxy syndromes refer to disorders of lateralisation whereby the arrangement of abdominal and thoracic viscera differ from normal and are mirror-images of normal.[1] They are classified into two main subgroups, right-sided isomerism with asplenia, and left-sided isomerism with polysplenia. The patients of right-sided isomerism with asplenia have more severe congenital heart diseases and lateralisation defects when compared with left-sided isomerism.[2] We present a primigravida with single ventricle heterotaxy syndrome for emergency caesarean section managed under graded lumbar epidural anaesthesia.

CASE REPORT

A 27-year-old primigravida at 36 weeks of gestation with pre-eclampsia in labour, presented to our hospital for safe confinement. Her past history revealed that she was a known case of complex cyanotic congenital heart disease. Though adviced, she had not undergone any corrective cardiac surgery. She had multiple hospital admissions due to chest infections for which she was treated with intravenous (IV) medications. She received infective endocarditis prophylaxis with benzathine penicillin 12 lakhs units deep IM from the age of 5 years till the present admission. She was also on tablet frusemide 10 mg twice a day for the past 1 week.

On examination, she was febrile with evidence of

marked central cyanosis, clubbing, and pedal oedema. Systemic examination revealed grade II parasternal heave and grade IV/VI ejection systolic murmur heard maximally at the upper left sternal border radiating over the entire precordium and neck with loud S_1 and single loud S_2. Auscultation of lung fields revealed crepitations in the basal region. Room air oxygen saturation (SpO_2) was 72%. Heart rate (HR) was 140/min and blood pressure of 140/98 mm of Hg in right upper limb in supine position.

Chest X-ray revealed moderate cardiomegaly of right ventricular predominance, dilated aorta with right-sided arch, and consolidation in the left upper and mid-zone. Echocardiography showed congenital heart disease of single ventricle physiology with mitral and pulmonary atresia, large non-restrictive ventricular septal defect with bidirectional flow, large non-restrictive atrial septal defect with bidirectional flow, severe tricuspid regurgitation with right ventricular systolic pressure of 115 mm of Hg, multiple aorto-pulmonary collaterals (MAPCAS) with normal ventricular systolic function [Figures 1 and 2]. Computed tomography (CT) scan of chest revealed enlarged liver in the midline with asplenia.

Patient was shifted to operating room after obtaining informed and written high risk consent. After securing IV access with 18 gauge IV cannula in left upper limb, she was administered cefotaxime 1 g IV for infective endocarditis prophylaxis. She was continuously monitored with five lead electrocardiogram, HR, non-invasive blood pressure, and SpO_2. Preloading was done with 300 ml of lactated Ringer's solution. Aspiration prophylaxis was given and oxygen was administered through facemask at 5 L/min. In left lateral position, at L1-L2 intervertebral space an 18 gauge epidural catheter was passed after locating the space with loss of resistance to normal saline. A test dose of 3 ml of 2% lignocaine was given, which confirmed the epidural placement. In supine position with left lateral tilt, 5 ml of 2% lignocaine with 50 μg of fentanyl was given epidurally, followed by 2 ml of 2% lignocaine after 10 min. A sensory level of T6 was achieved with stable haemodynamic parameters. Five minutes after skin incision, a live male baby weighing 2 kg was delivered. Baby cried immediately with APGAR score of 8 and 9 at 1 and 5 min, respectively. Midazolam 1 mg and fentanyl 50 μg were administered IV and 10 IUs of oxytocin was administered through infusion. Surgery was completed in 15 min. Post-operative analgesia was obtained with 0.1% bupivacaine and fentanyl

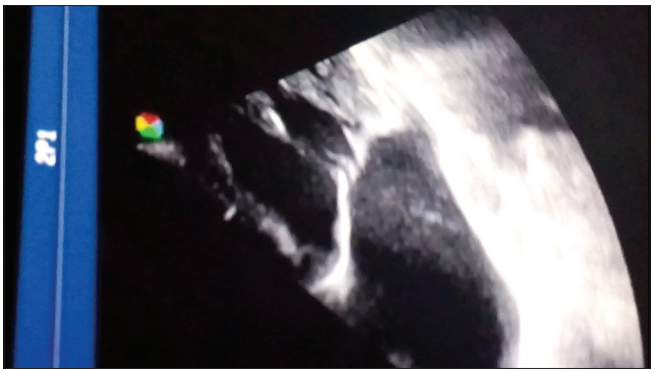

Figure 1: Echo showing rudimentary left ventricle with dilated and enlarged right ventricle

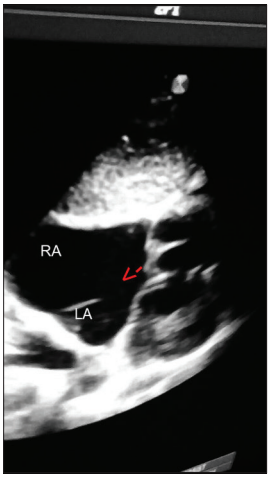

Figure 2: Two-dimensional echocardiography showing large non-restrictive atrial septal defect with bidirectional flow

1 μg/ml at 4 ml/h for 3 days. Patient was discharged on 20th post-operative day with the advice for follow-up with Cardiology and Obstetrics and Gynaecology consultants.

DISCUSSION

Heterotaxy results from failure of the developing embryo to establish normal left-right (L-R) asymmetry and is associated with a wide range of major cardiac and extracardiac congenital anomalies. The estimated incidence of heterotaxy syndrome is 1 in 6000 to 1 in 20,000 live births.[3] The cardiac lesion found in our patient was hypoplastic left heart syndrome (HLHS) with transposition of great vessels (TGA). HLHS was first described by Lev in 1952.[4] Without surgical intervention, this lesion is uniformly lethal.[5] The most common causes of mortality were arrhythmias, congestive heart failure, and sudden unexplained death.[1] The development of MAPCAS is probably one of the reasons, that our patient survived till adulthood without any palliative surgery.

Epidural anaesthesia was administered to our patient as it has a number of benefits in patients with single ventricle physiology. The sympatholysis associated with epidural block dilates the venous system and attenuates the large swings in blood volume seen at delivery. This sympatholysis was achieved gradually to avoid sudden changes in systemic vascular resistance and potential reversal of shunts leading to maternal hypoxaemia. In addition to excellent analgesia, regional anaesthesia avoids the negative inotropic effects of most anaesthetic agents.[6]

There is a paucity of literature regarding the anaesthetic care of adult patients with unrepaired cardiac anomalies. An adult female patient with congenital cyanotic heart defect had been palliated at 8 years of age. However, the major constituents of her anomalies remained unrepaired. General anaesthesia was administered to this patient for left-sided ureteroscopy with laser lithotripsy and stent placement.[7] Another case of a 29-year-old parturient with a single ventricle and TGA, who had lumbar epidural analgesia/anaesthesia for labour, emergency caesarean section and post-operative pain has been described.[8] A case of a pregnant woman with a functionally univentricular heart and double outlet right ventricle who underwent caesarean section was managed using an epidural technique.[9]

CONCLUSION

Successful pregnancy is difficult in women with complex congenital heart diseases. For optimum outcome, these patients should be managed by multidisciplinary team including obstetricians, cardiologists, and anaesthesiologists. We could successfully manage this case with graded epidural anaesthesia for emergency caesarean section and post-operative analgesia.

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