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Case Report

A rare case of Ebstein's anomaly scheduled for elective caesarean section

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Abstract

Ebstein's anomaly is a rare congenital heart defect comprising less than 1% of patients with congenital heart disease. Among the congenital heart lesions, Ebstein's anomaly is one of the most diverse in presentation, severity and management. It involves the septal cusp of the tricuspid valve wherein, due to anomalous attachment of the leaflets of the valve the cusp is elongated causing downward displacement of the valve into right ventricle. This results in tricuspid regurgitation. The abnormally situated valve produces atrialization of the right ventricle. It may be associated with atrial septal defect and supraventricular arrhythmias. Here, we report a case of second gravida patient with Ebstein's anomaly. She was diagnosed with it a year back and was not on any treatment. Her echocardiogram showed grossly dilated right atrium grade III tricuspid regurgitation with mild pulmonary artery hypertension. Patient was taken up for elective caesarean section under spinal anaesthesia.

Key-words: caesarean section, spinal anaesthesia, bupivacaine.

Introduction

Ebstein's anomaly is a rare congenital heart defect involving the tricuspid valve leaflets was first described by Wilhelm Ebstein.[1] It is characterised by anomalous attachment of the leaflets of the valve causing cusp elongation which leads to downward displacement of the valve into right ventricle. This produces atrialization of the right ventricle. Among the congenital heart lesions, Ebstein's anomaly is one of the most diverse in presentation, severity and management. The incidence in general population is 1:110,000. This anomaly can pose risk in normal individuals and in pregnancy it carries a higher risk due to many physiological and hormonal changes. [2]

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Case History

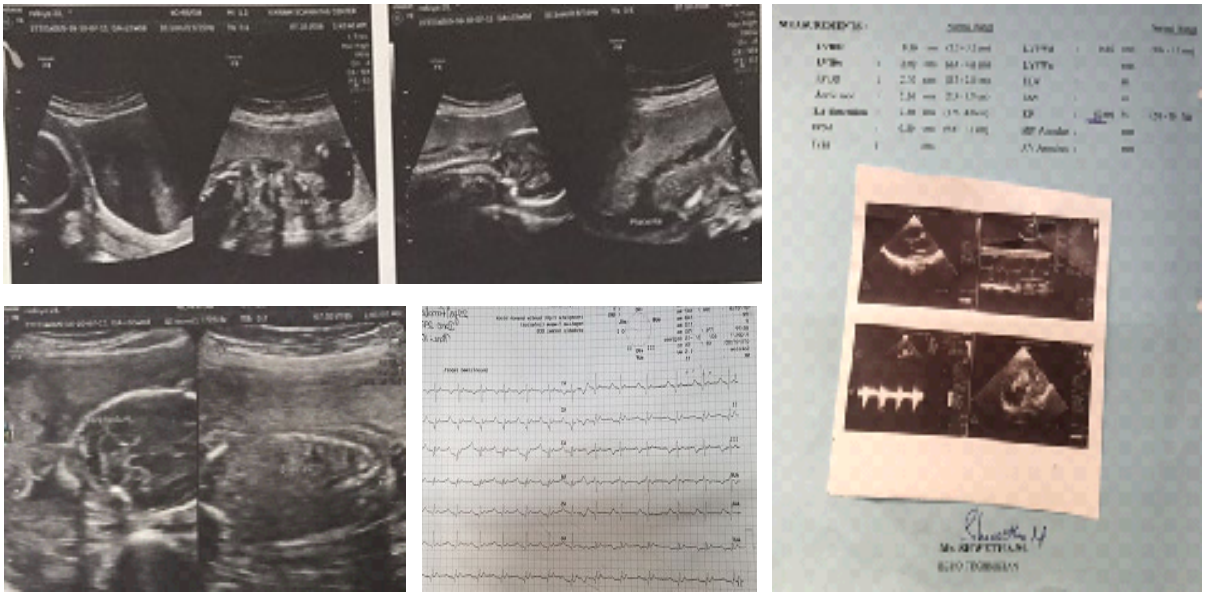
A 25-year-old second gravida diagnosed with Ebstein's anomaly was referred to our hospital for elective caesarean section. She had one previous spontaneous abortion one and a half years back and presented with 38 weeks of gestation She was not any medication for the same. She had exertional dyspnoea New York Heart Association Class 1. She had no history of recurrent chest infections, chest pain, cyanosis or palpitations. On examination, pulse rate was 80 beats per minute, regular in rhythm, normal in volume, and character with no radio-radio or radio-femoral delay. Blood pressure was 110/64 mm of Hg measured in right arm supine position. Cardiovascular examination revealed apex beat in sixth intercostal space in midclavicular line, Grade 2 parasternal heave on palpation. On auscultation pan systolic murmur, increasing on inspiration in tricuspid area with muffled heard sounds , pan systolic murmur in the mitral area and aortic area was heard .Electrocardiogram

showed sinus rhythm, large p waves indicating right atrial enlargement and incomplete right bundle branch block. Chest X-ray showed cardiomegaly with prominent right heart border. Recent echocardiogram showed atrialization of the right ventricle, dilated right atrium, Grade III tricuspid regurgitation, pulmonary artery systolic pressure of 35 mm of Hg, mild pulmonary artery hypertension with tricuspid septal leaflet apical displacement of 30mm. Routine investigations like total count, differential count, haemoglobin, bleeding time, clotting time and urine routine were within normal limits.

Patient had hemodynamic stability with good effort tolerance. Contemplated technique of anaesthesia was subarachnoid blockade. All routine and emergency drugs and equipments were kept ready in operation theatre. Patient was kept nil by oral for eight hours for solid foods and four hours for clear fluids. Tab. ranitidine 75mg was given the previous night. On the day of surgery in the preoperative area,informed written consent was taken. 18G IV cannula was secured. Infective endocarditis prophylaxis was administered with Inj. ampicillin 2g and Inj. gentamicin 80 mg intravenously. Aspiration risk prophylaxis was given with Inj. ondansetron 4 mg, Inj. ranitidine 50 mg, Inj. metachlorpromide 10 mg IV was given. Precautions were taken to avoid air bubbles in the peripheral venous lines.

In the operation theatre, electrocardiogram, pulse oximeter and non-invasive blood pressure monitors were attached. Apart from these routine monitors we used invasive blood pressure monitor canulating the radial artery. Under aseptic precautions, lumbar puncture was done in left lateral position with 26 G needle. Inj. Bupivacaine 0.5% hyperbaric (5mg/ml) 1.8ml with Inj. Buprenorphine 30 micrograms (0.1ml) was administered intrathecally after confirming CSF backflow. Sensory blockade was achieved till T6 level checked by loss of sensation to 23 G hypodermic needle. Motor blockade was Bromage scale 3. Oxygen was supplemented with facemask at 5 litres per minute rate. Incision to delivery time was 6 minutes. A live female baby weighing 2.39kgs was extracted with an Apgar score of 8 at first minute and 10 at 5 minutes. Soon after extraction of baby, 15 units of Inj. oxytocin were given by slow IV infusion. Inj. midazolam 1mg and Inj. fentanyl 30 micrograms IV were given. Patient was hemodynamically stable throughout the procedure with mean arterial pressure between 70 to 100 mm of Hg and heart rate between 70 to 90 beats/min and oxygen saturation 98-99%.

Surgery lasted for 30 minutes. Post operatively patient was shifted to ICU for observation. Patient was monitored for her vitals until two segment regression till T10. Patient was administered with post operative analge-



sia with diclofenac 75mg infusion when VAS score was equal to or more than four. Early ambulation was advised.. Post-operative period was uneventful. The patient and baby were discharged home on 7th post-operative day with a referral for management by cardiology.

Discussion

Ebstein's anomaly is a rare congenital cardiac condition with an incidence of 1 in 100,000 in general population. It was described by Wilhelm Ebstein in 1866. [3] Ebstein's anomaly has its pathology in the tricuspid valve leaflets. There dysplastic abnormalities of the basal and free attachments of the leaflets. It results in tricuspid regurgitation and sometimes stenosis. Also there is downward displacement and elongation of the septal and anterior cusp leading to atrialization of the ventricle wherein part of the right ventricle effectively forms part of atrium. This compromises the right ventricle contractility. [4]

Also there can be right to left shunting due to some interatrial communication. Progressive cyanosis can be seen in case of shunting. Symptoms of right ventricular dysfunction like pulmonary hypertension may be seen. Paroxysmal atrial tachyarrhythmia with or without atrioventricular bypass tracts that is Wolff Parkinson White syndrome is the commonest dysarrythmia. Other complications include pulmonary emboli, systemic emboli, congestive cardiac failure and sudden cardiac collapse. [5]
Mostly Ebstein's anomaly present in neonates and infants with cyanosis and congestive cardiac failure. If the patient survives till adulthood then it can get precipitated due to any stress event most commonly pregnancy. Though it is found to be well tolerated in pregnancy, complications like prematurity, foetal loss and congenital heart disease in the off spring are seen. Patients showing symptoms like arrhythmias, cyanosis and preeclampsia have increased foetal and maternal risk. Our patient was acyanotic and not in failure. She had cardiomegaly with mild pulmonary hypertension. Our anaesthetic aim in these patients must be to maintain a sinus rhythm, heart rate, preload, contractility, prevent rise in pulmonary vascular resistance, and maintain systemic vascular resistance. [6]
Our contemplated technique of anaesthesia was subarachnoid blockade.

Complications with general anaesthesia include supraventricular arrhythmias during induction whose incidence is 20% which may not respond to drugs. It also has increased risk of shunting, polypharmacy and aspiration.
Hence neuraxial blockade is a better technique of anaesthesia with advantages like controlled hemodynamics, post-operative analgesia, and early recovery. Epidural has its disadvantages in pregnancy like difficulty in space identification, difficulty in space identification and increased chance of bloody tap and dural puncture. Also the time of blockade onset is delayed.
With proper preloading, fluid management and usage of low dose local anaesthetics subarachnoid blockade can be considered a better method of anaesthesia. The above precautions can prevent the hypotension as well. If in case of hypotension graded doses of phenylephrine is the ideal treatment. Ephedrine, commonly used vasopressor during caesarean section may precipitate supraventricular tachycardia in these patients. Paradoxical emboli which can be life threatening must be prevented by avoiding air bubbles in the peripheral venous lines. Since our patient was hemodynamically stable we avoided central venous catheterization as it can cause arrhythmias and infective endocarditis, in addition to being technically difficult.[7,8] Certain precise cardiac monitoring like bedside echocardiography and transesophageal echocardiography are useful during surgery in such cases,[9] but were unavailable in our institute.

Oxytocin was used judiciously as rapid infusions or boluses can cause severe vasodilation and hypotension since they increase pulmonary vascular resistance methylergometrine and prostaglandins are better to be avoided. Even though regional anaesthesia is a safe alternative, there have been reports of patients being operated safely under general anaesthesia for both obstetric and other incidental surgeries. General anaesthesia with intubation enables control of oxygen deliver. [10]
In our patient since she had good effort toler-

ance and hemodynamic stability, we used subarachnoid blockade.

Conclusion:

Subaracnoid blockade can be a better technique of anaesthesia in hemodynamically stable ebsteins anomaly cases as it provides post operative analgesia, avoids polypharmacy, early rooming in and better and faster recovery.

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