CASE REPORT

Anaesthetic implications of Ebstein's anomaly

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ABSTRACT

Ebstein's anomaly is an uncommon congenital heart disease which involves the septal cusp of tricuspid valve. The cusp is elongated, so it embraces a part of right ventricle, thus atrializing a portion of the ventricle. It may be associated with atrial septal defect and supraventricular arrhythmias. Here we describe two cases of Ebstein's anomaly; one posted for elective Caesarean section and the other for total abdominal hysterectomy. The first patient was diagnosed to have this anomaly in her first pregnancy and was taking oral verapamil irregularly for supraventricular arrhythmias. Her 2D echo report showed an enlarged right atrium, severe tricuspid regurgitation and atrial septal defect. Post-operative leg stockings, low molecular weight heparin was given to prevent thromboembolic episodes. The second case was a middle aged woman in her late forties with fibroid uterus. She was not on any medication. General anesthesia was chosenfor both of the patients with preoperative aspiration prophylaxis and antibiotic prophylaxis for infective endocarditis. Both patients had an uneventful perioperative course.

Key words: Ebstein's anomaly; Caeserean section; congenital heart disease; general anaesthesia; pregnancy; tachyarrhythmias.

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INTRODUCTION

Ebstein's anomaly is a rare congenital heart disease; in which there is a downward displacement of the tricuspid valve into the right ventricle. It has a distal effective portion and an atrialized proximal portion[1]. It is frequently associated with intracardiac shunting, pulmonary hypertension, cardiac dysrhythmias and cyanosis[2]. Clinically, there is a wide spectrum of severity of disease with patients ranging from those who are asymptomatic to those who are debilitated[1]. Congestive heart failure and sudden collapse are the most common causes of death. The incidence in general population is 1:110,000 and there is no sex difference.[1,2]

CASE REPORT 1

A 25 years old female, who had undergone Caesarean section 3 years back, under general anaesthesia, presented with 9 months of amenorrhoea. She had been diagnosed to have Ebstein's anomaly in the previous pregnancy. After the first trimester, she had had dyspnoea, which gradually increased in intensity and at the time of presentation she was breathless even with minimal activity. However, she remained comfortable at rest and there was no history suggestive of paroxysmal nocturnal dyspnoea. She also had episodes of palpitations and occasional syncope, which were precipitated by exertion and stress. She was taking tab verapamil 40 mg BID since the previous pregnancy. There was no history of cyanotic spells. She had a recent history of mumps with fever, cough and running nose since 5-6 days. There was no significant family history and she had no addictions.

On examination, she was found to be an average-build lady with regular pulse, she was normotensive with bilateral pedal pitting edema and bilateral tender parotid swellings. Jugular venous pressure was not raised and

there was no pallor or cyanosis. Mouth opening was adequate with Mallampatti grade II. There was no evidence of inflammation at parotid duct opening in the mouth. Examination of cardiovascular system revealed the presence of a loud pansystolic murmur, best heard in the tricuspid area. Liver was not palpable and there were no other signs of heart failure. Investigations showed a haemoglobin of 11.2 gm%, normal blood counts, normal liver/ renal functions and blood sugar. Her ABG report showed pH=7.4, PCO_2=26.6, BE=-6.4, BE (ECF)=8.3, BB=41.5, HCO_3=16.3, PO_2=61.1 and O_2 sat=90.7, indicating a chronic metabolic acidosis. A 2D echo, done 6 months back, showed a hugely dilated right atrium and ventricle, an atrial septal defect of 13 mm size and grade-III tricuspid regurgitation. She had a depressed right ventricular systolic function. There was no evidence of pulmonary hypertension and the left ventricular function was normal. ECG showed right axis deviation but no arrhythmias.

The patient was referred to the physician for complete evaluation and management. She was instructed to continue taking verapamil on regular basis. In view of the recent history of mumps and upper respiratory tract infection, she was given antibiotics, steam inhalation and antiseptic mouth gurgles.

General anesthesia was planned for Caesarean section. Tab ranitidine 150 mg was given on the night prior to surgery and in the morning with sips of water. Verapamil was also continued till the day of surgery. Intravenous line was secured with an 18G cannula and urinary catheterization done under strict asepsis. Inj. Ondansteron 4 mg IV was given. Antibiotic prophylaxis for infective endocarditis was given (inj ampicillin 2 gm and inj. gentamicin 80 mg IV). Routine monitoring was done. Rapid sequence induction was done after preoxygenation for 3 minutes with inj. propofol 80 mg and inj. ketamine 30 mg given in slow incremental doses. Inj. rocuronium was used for intubation and as a relaxant throughout the surgery. Cricoid pressure was continued till the inflation of the tube cuff. Anesthesia was maintained on sevoflurane 1.5-2.5% in oxygen and nitrous oxide through a Bain's circuit. After delivery of the baby, inj. midazolam 1 mg and inj. fentanyl 50 µg were given. The baby was a 2.6 kg male child, who cried well at birth with APGAR scores of 8, 9 and 9 at 1, 5 and 10 min respectively. Patient's vital signs remained stable throughout the procedure with pulse in the range of 70-90 beats/ min, blood pressure in the range of 100/70 to 140/80 mmHg. Intraoperative blood loss was estimated to be about 600ml. Patient received 1 litre of Ringer lactate intraoperatively. She was extubated uneventfully at the end of the procedure. Inj. tramadol and paracetamol rectal suppositories were given for post-operative analgesia, and low molecular weight heparin and leg stockings to prevent thromboembolic episodes. Early ambulation was advised. Her postoperative stay in the hospital was uneventful.

CASE REPORT 2

A 47 year old lady presented with history of pain in abdomen and mennorhagia since 5-6 months. She had breathlessness (New York Heart Association-grade II-III) and giddiness on and off for 2-3 years. On examination, she was pale with a regular pulse 90/min, blood pressure 120/60 mmHg. There were no signs of cardiac failure; a pansystolic murmur was audible in mitral and tricuspid areas. 2D echocardiogram revealed the presence of grade-I Ebstein's anomaly and mild mitral regurgation. ECG showed a sinus rhythm and an evidence of right ventricular hypertrophy. Routine blood investigations were within normal limits. Blood gas analysis was also normal. She was transfused three pints of blood to correct her severe anaemia (Hb-6.5). General anesthesia was planned for the scheduled abdominal hysterectomy. The course of anaesthesia, monitoring done and postoperative management was carried out on the lines of case-1 described above.

DISCUSSION

Ebstein's anomaly is a very rare congenital heart disease. There are few cells in Ebstein's right ventricle than are normally found, which contribute to ventricular dilatation in addition to tricuspid regurgitation.[3] Left ventricle may also be poorly contractile with mitral valve prolapse. In severe form of disease, because of atrialized right ventricle and poor ventricular function, tricuspid regurgitation is severe, raising right atrial pressure above left atrial pressure resulting in right to left shunt. Also because of a large atrium, there is potential for varying degrees of blood pooling in this hypocontractile portion of the heart[4]. The abnormal development of tricuspid valve results in several activation abnormalities including delayed intra atrial conduction, right bundle branch block, ventricular pre-excitation etc. Because of this the patients are subject to paroxysmal arrhythmias which may be supraventricular or ventricular. The Wolff-Parkinson-White syndrome (WPW) occurs in up to 20% of cases[1], because the right atrium gets incorporated into the ventricle, bypassing

the annulus fibrosus.[3] WPW in Ebstein's anomaly is less responsive to routine medications. Atrial fibrillation associated with WPW may degenerate into ventricular arrhythmia which may be malignant and fatal.[2] DC cardioversion may be required and it can be done safely in pregnancy[2]. Patients with intra atrial communication are at risk of paradoxical embolism, brain abscess, CCF and sudden death.[4]

In pregnancy several physiological changes occur within the cardiovascular system. Plasma volume increases with a smaller rise in red cell mass. Cardiac output rises by 40% with an increase in both stroke volume and heart rate and with on accompanying rise in circulating catecholamines. Systemic blood pressure is lower than in non pregnant state indicating a considerable reduction in peripheral vascular resistance and placental shunting.[5]

In patients with Ebstein's anomaly these physiological changes may have appreciable adverse haemodynamic consequences. In the presence of impaired right ventricular size and function, increased blood volume may be poorly tolerated and result in worsening of tricuspid incompetence, raised right atrial pressure and increased right to left shunting. Complications of pregnancy and delivery such as haemorrhage may not be well tolerated, where right ventricular dysfunction may impair patient's capacity to cope with the large volume shifts associated with fluid resuscitation. The patients may have a variety of arrhythmias. The raised concentration of circulating catecholamines in pregnancy will exacerbate any predisposition to arrhythmias; major arrhythmias may occur specifically during maternal hypoxia or stress.[1,5] CCF may occur, particularly during 2nd trimester because of increased circulating blood volume and again shortly after delivery following uterine retraction and placental separation.[6] Pulmonary and systemic embolism is a constant threat during later stages of pregnancy and in the perioperative period.[1,6]

Basic principles of management of Ebstein's anomaly cases are[1]; to maintain prelaod and after load and maintain sinus rhythm, to prevent increased right to shunting which may occur if there is decrease in systemic vascular resistance or increase in pulmonary vascular resistance or with increased intrathroacic pressure and avoidance of tachycardia as this leads to impaired right ventricle filling.[7] Antibiotic prophylaxis is advisable to prevent infective endocarditis and invasive monitoring is to be avoided as far as possible because of high risk of infective endocarditis, paradoxical embolism and arrhythmias. Also the procedure is likely to be technically difficult with high failure rate in these patients.[1,2,7,8] IV fluids are to be restricted and given with meticulous titration because of risk of right ventricular failure.[2] Air in IV lines should be meticulously removed.

These patients pose challenges for both neuraxial and general anesthesia. General anesthesia is preferred in patients with severe forms of disease. The advantage of general anesthesia is that hypotension is usually avoided and fluid balance is easier to control.[1] Although epidural has reportedly been given successfully to patients with Ebstein's anomaly, in our case we chose general anesthesia over epidural, because she had chronic metabolic acidosis and associated atrial septal defect suggesting that she had an element of right to left shunt. So there was the risk of exacerbation of this shunt in the event of decreased systemic vascular resistance and hypotension due to epidural.[1] Further, the patient had Grade III tricuspid regurgitation and a depressed right ventricular systolic function. So there was a risk of right ventricular failure in case of fluid resuscitation required in the event of epidural induced hypotension or obstetric haemorrhage.[1,2,9]

In the second case the reason of choosing general anesthesia was the risk of right ventricular failure with volume loading had the patient had right ventricular hypertrophy and tricuspid regurgitation. A possibility of a poorly contractile left ventricle could not be ruled out as the patient had mitral regurgitation. Reduced peripheral resistance associated with epidural or spinal anesthesia could have compromised blood supply to the peripheral areas.

Preoperative preparation of the pregnant patient was done by starting her on antiarrhythmics, antibiotics and steam inhalation. A rapid sequence induction with reduced doses of propofol and ketamine can prevent sudden hypotension caused by propofol alone, which may exacerbate the shunt fraction.[3,7] We used thiopentone in slow incremental doses for induction in the second patient. Thiopentone was relatively contraindicated in the first case due to a history of recent upper respiratory tract infection.

Synthetic oxytocin must be given by slow infusion to avoid vasodilation.[1] Ergometrine should be avoided because of its adverse effect on pulmonary vasculature.[1,6] Prostaglandin F2 a is also a useful alternative.[6]

These patients are prone to cardiac arrhythmias. Hence E.C.G was continuously monitored and a defibrillator was kept ready. Factors which are known to precipitate

arrhythmias, e.g. light plane of anesthesia or a fluid or acid base disturbance need to be avoided.[9]

We conclude that the successful management of a patient with Ebstein's anomaly requires a deep understanding of pathophysiology of this cardiovascular disease and its effects on pregnancy. A multidisciplinary approach is the key to successful outcome in these patients.

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