

Surgical Excision of Left Atrial Chondrosarcoma and Mitral Valve Repair in a 30 Weeks Pregnant Female

Mohsin Nazir Butt^{1*}, Aurangzeb Durrani² and Jahanzeb Khan²

¹Department of Anaesthesia and ICU, Aga Khan University Hospital Karachi, Pakistan

²North West General Hospital and Research Center, Peshawar Pakistan

Abstract

This case report is regarding a 32 years old female with 30 weeks gestation, presented to the obstetric department first time with gestational amenorrhoea. She has history of exertional dyspnoea, orthopnoea and attacks of syncope on and off from last three years. On further work up, she has got left sided intra atrial mass of about 41×36 mm in size attached to the lateral wall of left atrium, protruding into left ventricle through mitral valve, during diastole causing severe obstruction. Left ventricular systolic function was normal, estimated ejection fraction of 58% and no regional wall motion abnormality.

Fetal heart rate and other parameters of fetal wellbeing was assessed, they were within normal limits. Patient and her family were counseled about her critical condition.

In the presence of obstetrician within the operating room, cardiac surgery was started with all essential monitoring. A successful excision of atrial mass and mitral valve repair was done with a cross clamp time of 45 minutes and total cardiopulmonary bypass time of 105 minutes. Postoperatively, patient was shifted to cardiac ICU and extubated when met the criteria of extubation and she was kept in cardiac ICU for 48 hours. In CICU, fetal cardiac activity was assessed continuously. All parameters regarding fetus were remained within normal limits and Obstetrician decided to plan for Caesarian section vs. spontaneous vaginal Delivery after four to six weeks, with regular weekly visits to labor suit. At 36th week, patient was admitted for induction of labor or possible caesarean section. The whole of the labor went uneventful and she delivered a male baby with good APGAR score. There were no postpartum complications. Histopathology of the excised mass showed a very rare intra cardiac tumor i.e. Chondrosarcoma.

Keywords: Atrial Chondrosarcoma; Cardiac mass excision in pregnancy; Mitral valve replacement

Introduction

Primary cardiac tumors are rare, with an autopsy incidence ranging from 0.001% to 0.030% [1]. Chondrosarcoma is a malignant tumor of cartilaginous tissues that has been exceptionally described in the heart. Primary malignant tumors of the heart are uncommon comprising mainly of primary sarcomas, predominantly angiosarcoma, fibrosarcoma, and rhabdomyosarcoma [2].

Chondrosarcomas of the heart are very rare and the majority of them are secondary, where the primary site can be identified. Metastatic cases are usually located in right atrium [3]. Primary chondrosarcoma of the heart has not been described in WHO classification of cardiac tumors [4]. The prognosis of patients with cardiac chondrosarcoma is poor; survival is measured in weeks or months [5]. In this case report we are going to discuss the surgical excision of atrial Chondrosarcoma in a 30 weeks pregnant female. The important aspect of this case report is a malignant cardiac tumor which was operated during third trimester of pregnancy. It was a challenge to a multidisciplinary team of Anaesthesiologist, cardiologist, cardiac surgeon, perfusionist and obstetrician. Along with the specific anaesthesia concerns of pregnancy, like aorto-caval compression, maternal or fetal hypoxia, risk of aspiration, probability of difficulty in maintaining airway, uterine contraction, maintenance of utero-placental blood flow, fetal viability and outcomes, there were additional concerns regarding cardiopulmonary bypass like alterations in coagulation, the release of vasoactive substances, and activation of the complement system, air and particular emboli, nonpulsatile flow, hypotension, and hypothermia. The maternal mortality ranges from 1% to 5%, while fetal mortality ranges from 16 % to 33% [6]. According to some studies the mortality rates depends upon the preoperative functional class, i.e. increasing functional class proportionally increase mortality rates [6].

Case History

Patient was seen by a team of obstetrician, Cardiac surgeon and Anesthesiologist preoperatively. Antepartum fetal assessment was within normal limits. After a combined discussion of medical team and her family, the patient was planned for surgical excision of left atrial mass, mitral valve repair followed by caesarean section.

Patient and her family were counseled about the critical condition of patient and possible adverse outcomes of this surgery. High risk informed consent was taken from the patient and her husband. Possibility of fetal demise was also discussed. The patient and her family members gave the consent.

Preoperative preparations include Nothing per oral for six hours preoperatively, blood and blood products, postoperative Cardiac ICU booking, (anxiolytic) tablet midazolam 7.5 mg one hour before surgery and aspiration prophylaxis (i.e. Inj. Zantac 50 mg one hour before surgery, Inj. Metoclopramide 10 mg half hour before surgery and sodium citrate 30 ml preoperative) in order to prevent aspiration of gastric contents specifically in pregnant patient. Before the induction

***Corresponding author:** Mohsin Nazir Butt, Department of Anaesthesia and ICU, Aga Khan University Hospital Karachi, Pakistan, Tel: 0092-321-2227083; E-mail: mohsin.nazir@aku.edu

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of anaesthesia, fetal cardiac monitoring with cardiocography (CTG) was done (Figures 1-4).

Continuous five leads ECG (to detect heart rate, rhythm and ischemic changes), non-invasive blood pressure, oxygen saturation, nasopharyngeal temperature monitoring, urine out- put, end tidal carbon dioxide, gasses and inhalational anaesthetic measurement were done. Before the induction of anaesthesia, invasive intra-arterial

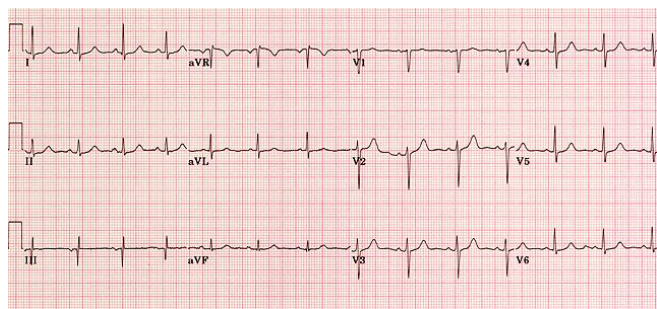


Figure 1: Preoperative ECG and Chest X-ray.



Figure 2: Preoperative Chest X-Ray.

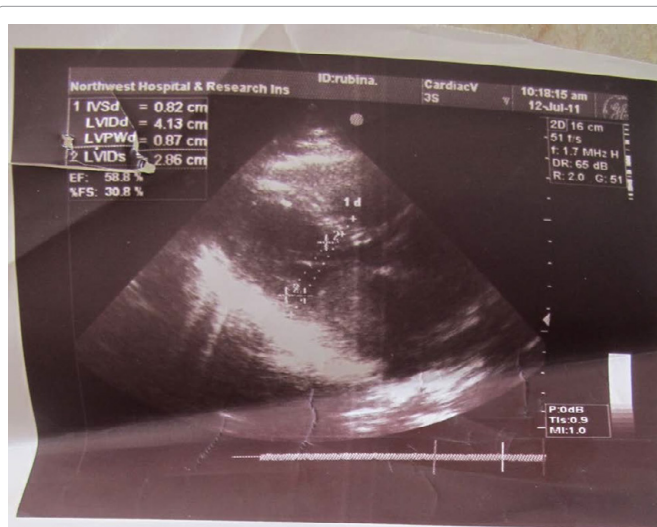


Figure 3: Preoperative Echocardiography.



Figure 4: Left Atrial Tumor (Chondrosarcoma).

line was inserted in left radial artery for more precise and beat to beat arterial blood pressure monitoring, which is one of the crucial monitoring during cardiac surgery. Preoxygenation with 100% oxygen was done for three minutes. Anaesthesia induction was done with injection midazolam 3 mg and Morphine 10 mg intravenously, once the loss of verbal response was achieved cricoid pressure was applied in order to prevent gastric aspiration. Injection rocuronium 60 mg was given intravenously and trachea was intubated after one minute with endotracheal tube of 7.5 mm internal diameter. Once we confirmed the proper placement of endotracheal tube, cricoid pressure was released and tube was fixed at 20 cm. Central venous line and pulmonary artery catheter were inserted from right internal jugular vein. Central venous pressure and pulmonary artery pressure was also monitored, in order to manage the patient according to the parameters of pulmonary artery catheter and central venous pressure throughout the surgery and post operatively. Additionally, fetal monitoring was done with preoperative ultrasonography to check fetal viability and continuous fetal heart rate monitoring specially during cardiopulmonary bypass phase, was also done with cardiocography (CTG).

Preoperative central venous pressure was 20 mmHg and pulmonary artery (PA) pressure of 60/28 mmHg, pulmonary capillary wedge pressure (PCWP) was 30 mmHg, cardiac output 5.6 L/min and cardiac index of 2.8 l/min/m². Patient was maintained on Isoflurane 1.2% with oxygen 50% and Air 50%. A minimum alveolar concentration (MAC) of 1 was obtained. The Patient remains stable after induction of anaesthesia. Base line activated clotting time (ACT) was 155 seconds. After all aseptic measures and draping, median sternotomy was done and pericardium was opened. Heparin 20000 units intravenous was given. After three minutes ACT was repeated. The repeated (ACT) came out to 550 seconds. Patient's circulation was put on cardiopulmonary bypass machine with average flow rate of 3.85 l/m. Cold Cardioplegia was given through coronary sinuses. Patient was cooled down to 30 degree centigrade. Total cross clamp time was 45 minutes and bypass (CPB) time was 105 minutes. Right atrium was opened and through trans-septal approach, left atrial tumor was excised and mitral valve was repaired. Throughout the CPB duration, fetal cardiac activity was

closely monitored. Initially, the fetal heart rate was slightly dropped down for a while but it recovered spontaneously within a few minutes. It was decided earlier that if the fetal heart rate dropped persistently then emergency caesarean section would be done and obstetrician was also with in the operating room and she herself was monitoring fetal activity. After completion of mitral valve repair, the patient was off from cardiopulmonary bypass and heparin was reversed with protamine sulphate. Cardiac output, cardiac index, systemic vascular resistance, pulmonary vascular resistance and central venous pressure was monitored during surgery and infusions of vasoactive, vasodilator and inotropic drugs were titrated according to the above mentioned monitoring. One of the targets was to maintain cardiovascular parameters in order to prevent utero-placental insufficiency. She remains stable post bypass and fetal heart rate was also within normal limits. She was on inotropic support with epinephrine at the rate of 0.05 mcg/kg/min and glyceryl trinitrate (GTN) at the rate of 0.5 mcg/kg/min. Hemostasis was secured and chest was closed.

Postoperative pulmonary artery pressure was 35/19 mmHg with PCWP of 20 mmHg. Cardiac output was 5.1 L/min. and Cardiac Index of 2.2 l/min/m². Postoperatively, patient was shifted to cardiac ICU. She was kept on ventilator support on assist control mode, and sedated with intravenous infusion of propofol 25 mcg/kg/min. After achieving post cardiopulmonary bypass parameters of extubation i.e. chest drains less than 200 ml/min, normothermia, serum potassium more than 4 meq/l, no acidosis and minimal inotropic support. Propofol infusion was stopped and when patient gradually woke up and spontaneously breathing, she was kept on trial of spontaneous breathing on pressure support mode at 15 cm/H₂O initially and gradually dropped down to 8 cm/H₂O. On successful trial of spontaneous breathing with minimal changes in hemodynamics, patient was extubated and kept on oxygen supplementation with face mask at 5-10 l/m.

Postoperative complete obstetric and fetal examination and postoperative ultrasonography were done which were normal. She remained in ICU for one week and then shifted to ward for further management.

At 36th week, patient was admitted for induction of labor or possible caesarean section. On examination of patient fundal height corresponded to 36 weeks gestation, longitudinal position and cephalic presentation. Eight hours after induction of labor, Patient delivered spontaneously. She spontaneously delivered a male baby of good APGAR score and birth weight of 2.8 kg. The vital signs of the patient remained stable throughout the labor.

Histopathology of intracardiac tumor showed malignant neoplastic lesions with dimorphic growth pattern comprising of island of well differentiated cartilage alternate with undifferentiated stroma. Features are those of Mesenchymal chondrosarcoma.

Later on patient was referred to oncologist for chemotherapy. Chemotherapeutic regimen was suggested but the patient and her family refused for chemotherapy.

Discussion

The most common sarcoma is vascular in origin, particularly angiosarcomas; however a variety of bony, neurogenic, cardiogenic and soft tissue variants have been reported [7]. Primary heart sarcoma is rare, constituting less than 25% of all cardiac neoplasia. Among the malignant tumours, sarcomas are the more frequent histological type, and they present dissemination potential and local invasion [8]. Cardiac sarcomas are poorly differentiated at presentation; the

histopathologic classification is extremely difficult [9]. The prognosis is generally poor and usually measured in weeks or months [9]. A study reported a mean survival rate of 11 months and a median survival of 6 months in patients presenting with primary cardiac tumor [10]. Simpson et al. found a 17-month median survival for patients with complete surgical excision and 6-month median survival for patients in whom complete surgical excision could not be achieved [11]. The incidence of heart diseases in pregnant women ranges from 1% to 4% [6]. The most frequent pathology is valve disease (93.2%). Mitral valve disease is the most prevalent (72.9%), and mitral commissurotomy or replacement is required in 78% of the cases while the ideal gestational age for cardiac surgery is 13th to 28th week [6]. Heart surgery during pregnancy is associated with significant maternal and fetal mortality rates. There are several identified risk factors which increase maternal and fetal mortalities, such as maternal age, gestational age, functional class, emergency surgery, myocardial protection and anoxic time. Few measures to reduce maternal and fetal mortality include avoiding functional deterioration, earlier surgery to prevent these patients from requiring an emergency procedure; performing surgery with minimal extracorporeal circulation time; adequate fetal monitoring and perform surgery in the second trimester of pregnancy.

In this case patient presented late to the antenatal clinic (i.e. in her third trimester of pregnancy) with significant signs and symptoms of cardiac disease and had a large intracardiac tumor. During surgery we tried to control all possible risk factors to reduce the maternal and fetal mortality. Post operatively patient was seen by oncologist and chemotherapeutic regimen was suggested but the patient and her family refused to take chemotherapy. Three months Postoperative Echo showed normally working mitral valve with no remaining intra cardiac growth. But bone scan that was done six months postoperatively showed different metastatic deposits in different areas.

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