Successful Management of Maternal Left Atrial Myxoma in Pregnancy

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ABSTRACT

A 29-year-old woman (gravida 3, para 2) presented at 28 weeks+2 days of gestation with a two-months history of dyspnea associated with orthopnea and occasional palpitations. On transthoracic echocardiography, she was diagnosed with a 3.2 × 2.7 cm left atrial myxoma. The patient underwent open surgical resection at 30 weeks of gestation. She had an uneventful postoperative recovery and was discharged on the ninth day. At 41 weeks of gestation, she gave birth by cesarean to a healthy baby of normal weight. Both the mother and the baby were discharged in stable condition.

Cardiac tumors are very uncommon. Myxomas are the predominant cardiac tumors in adults, occurring more frequently in women between the third and sixth decades of life,[1,2] and are occasionally encountered during pregnancy.[3] Cardiac myxoma is extremely rare, with a reported incidence of around 0.5 per one million people.[4] However, they are being more frequently diagnosed and managed surgically due to the widespread use of echocardiography.[5]

The clinical manifestations of myxoma are dependent on its size, mobility, and location.[6] They are commonly found on the left side.[6] The most common symptoms are constitutional in nature such as myalgia, fever, fatigue, and weight loss. About half of the cases present with symptoms suggestive of left ventricular inflow obstruction, such as dyspnea, dizziness, and syncope.[6–8] Other less common symptoms are neurological, such as stroke due to cerebral emboli of cardiac origin.[9]

Cardiac myxomas are rare in pregnant women and have been reported in only 44 publications involving 51 individuals. About 95% of them underwent transthoracic resection of the left atrial mass.[10] The indications for resection include the risk of embolic events and sudden death caused by myxoma obstructing the valve orifices.[11] A few authors have suggested deferring the surgery till after delivery or performing it in late pregnancy.[12,13]

Open heart surgery (the standard therapy) requires cardiopulmonary bypass which can cause alterations in coagulation, the release of vasoactive substances, activation of the complement system, emboli, non-pulsatile flow, hypotension, and hypothermia.[5,14] Hypothermia can lead to uterine contractions and reduction of placental blood flow.[14] Maternal mortality rates vary in the literature from 1% to 5%, with an average of 2.5%, and does not differ from those for non-pregnant women with similar diseases.[14] Fetal mortality rate due to surgery is 18.6%.[15,16] Median sternotomy is the standard surgical approach for cardiac myxoma removal. However, Taksaudom et al,[12] reported a case of myxoma resected two weeks postpartum using right anterior thoracotomy for the mother to have less pain and be able to hold and breastfeed her child.

We are presenting the first case of left atrial myxoma reported in Oman which was managed surgically in pregnancy without interfering with its progression and resulted in an overall good outcome.

CASE REPORT

A 29-year-old gravida 3, para 2 woman presented to the cardiology clinic at 28 weeks+2 days of gestation, with a two-month history of shortness of breath, increasing in severity in the preceding two weeks, and with associated orthopnea and intermittent palpitations. She had two previous

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uneventful vaginal deliveries with gestational hypertension and gestational diabetes complicating both those pregnancies. Her past surgical history was uneventful apart from a laparoscopic appendectomy and cholecystectomy which were uncomplicated. The current pregnancy was also uneventful with a well-grown fetus. She was on low-dose aspirin since the 12th week of gestation, in view of previous gestational hypertension. All her investigations were normal in this pregnancy including full blood count, screening for gestational diabetes, Hepatitis B, and HIV serology, and anatomy scan done at 22 weeks of gestation. A transesophageal echocardiogram done at another hospital at 26 weeks of gestation showed a left atrial myxoma 3.2 × 2.7 cm attached to the interatrial septum with intermittent brief mitral inflow obstruction and ejection fraction of 58%.

When she presented at our hospital, she did not require medications from a cardiac point of view. At 29 weeks of gestation, she was given dexamethasone 6 mg/12-hourly for two days to enhance fetal lung maturity. We repeated the echocardiogram after admission which showed normal biventricular dimensions, cavity size, resting wall motion, and systolic function. The left ventricular ejection fraction was 67%. A large highly mobile mass measuring 28 × 27 mm was noted in the left atrium with a thin stalk attaching to the interatrial septum. The heterogeneous density and overall appearance were consistent with a left atrial myxoma. This mass was noted to intermittently obstruct the left ventricular inflow as it prolapsed across the mitral valve annulus and into the left ventricle during the diastolic phase of the cardiac cycle. The effective mitral valve orifice in diastole was reduced to 1.9 cm^2 and generated a mean diastolic gradient of 4 mmHg at a heart rate of 75/min. This was consistent with moderate left ventricular inflow obstruction. There was no evidence of pulmonary hypertension.

Further plan of management was discussed in a multidisciplinary meeting involving a cardiothoracic surgeon, a cardiologist, an obstetrician, a neonatologist, and a cardiac anesthesiologist. After discussing all possible options with the patient and her spouse, it was decided to proceed with surgical excision of the myxoma.

The woman underwent left atrial mass excision under general anesthesia at 30 weeks of gestation. A median sternotomy incision was performed. Cross clamp time was 64 minutes. Cardiotocogram (CTG) was taken before and after the surgical procedure to monitor fetal well-being. Surgery was uneventful and she was shifted to the postoperative cardiac care unit (CCU) for monitoring. CTG post-procedure showed fetal tachycardia and reduced beat-to-beat (BTB) variability which settled by 12 hours postoperatively. This was attributed to the prematurity of the baby and continuous intravenous infusion of morphine given to the mother for pain relief. On the eighth-day post excision, a bedside transthoracic echocardiogram revealed an intact interatrial septum with no myxoma remnant. She was discharged on the ninth day in stable condition with advice to keep the pregnancy monitored at her regional hospital. She remained asymptomatic and underwent a cesarean section at 41 weeks of gestation for failed induction of labor. The outcome was a female baby with a birth weight of 3060 g and Apgar scores of 9 and 10 at 1 and 5 minutes. Both mother and baby were discharged on the third day in stable condition.

DISCUSSION

Cardiac myxomas are commonly found on the left side of the heart and the patient usually presents with dyspnea as in our case.[6–8] It is worth noting that dyspnea during pregnancy and the postpartum period is a relatively common symptom. The differential diagnosis is broad and includes etiologies such as pulmonary embolism, pneumonia, severe preeclampsia, and physiological changes in pregnancy.[17] Primary cardiac tumors such as atrial myxomas, given their rarity, are not usually considered as an initial diagnosis, and can be easily missed. Cardiac myxoma can be life-threatening and usually requires urgent surgical removal to prevent potentially serious embolic events, hemodynamic deterioration, or even sudden death.[18] Wang et al,[11] reported three pregnant women diagnosed with cardiac myxomas, out of which two were complicated by cerebral infarctions requiring urgent surgical resection. Liu et al,[19] also reported a similar case that was complicated with cerebral infarction and retinal artery occlusion. In pregnancy, and to balance the maternal and perinatal risks, the timing of surgical excision is important. The decision to postpone it to after delivery depends on the gestational age of pregnancy, worsening symptoms, and the echocardiography findings. Traisrisilp

et al,[20] reported a large (9 cm) cardiac myxoma in the left atrium diagnosed at 28 weeks gestation. Being asymptomatic, the patient was allowed to continue her pregnancy to full term. Two weeks postpartum, she underwent a successful resection of the myxoma.

The largest ever review of cardiac myxoma in pregnancy (based on 44 articles and 51 patients) was published by Yuan et al.[10] They found the most common symptoms to be dyspnea and palpitations and that cardiac myxoma was more likely to be diagnosed in the second trimester, as in our patient. Transthoracic echocardiography (which we adopted) was the most common diagnostic technique. The vast majority (95.9%) underwent myxoma resection during pregnancy with 47.2% of cases resected in the third trimester, with good maternal and fetal-neonatal outcomes. The authors concluded that proper timing of cardiac surgery, being non-emergency in nature, shorter operation duration, and improved cardiopulmonary bypass conditions would result in even better maternal and fetal-neonatal outcomes.[10]

Our initial plan was to delay the surgery till the fetus became more mature. But a repeat echocardiogram at the 29th week of gestation showed that the myxoma was causing moderate restriction to diastolic left ventricular filling. This was likely to get worse as plasma volume expanded towards the latter part of pregnancy. Moreover, the highly mobile nature of the mass and its rather unstable attachment to the interatrial septum portended a high risk for systemic embolization with potential catastrophic complications to the fetus and the mother. Based on the above, our multidisciplinary team decided to conduct the surgery early.

During open heart surgery with cardiopulmonary bypass, the patient is at risk of circulatory obstruction and hypotension during the prolonged operative time. This in turn may compromise the placental blood flow, leading to premature labor, non-reassuring fetal status, long-term disabilities, and fetal loss.[10] The fetal mortality rate during maternal cardiac surgery with cardiopulmonary bypass is approximately 18.6%.[15,16]

The obstetrician decided to forego intraoperative fetal monitoring, as episodes of non-reassuring fetal status are common during cross-clamp time and intervention can cause more harm than good to the mother and the fetus. Fetal heart was heard before and after the procedure. CTG done after the patient was shifted to CCU showed reduced BTB variability. We did not act on that also as other features in CTG were reassuring and the patient was on continuous intravenous infusion of morphine which accounted for the reduced BTB variability. As expected, the CTG returned to normal once the morphine infusion was stopped. The patient had an uneventful postoperative recovery followed by a normal antenatal period.

CONCLUSION

This is the first case of excision of left atrial myxoma in a pregnant woman in Oman. With input from a multidisciplinary team, the myxoma was managed by cardiothoracic surgery in the 30th week of pregnancy without interfering with the progression of pregnancy and resulted in overall good outcome.

Disclosure

The authors declared no conflicts of interest. Oral consent was obtained from the patient to publish this case report.

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