Successful Management of Maternal Left Atrial Myxoma in Pregnancy

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Received: 4 February 2021
Accepted: 12 September 2021

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DOI 10.5001/omj.2023.01

Abstract

A 29-year-old Gravida 3 Para 2 woman presented at 28 weeks + 2 days of gestation with 2 months' history of dyspnea associated with orthopnea and occasional palpitations. She was diagnosed with a 3.2x2.7cm left atrial myxoma on transthoracic echocardiography and underwent open surgical resection at 30 weeks of gestation after a multidisciplinary team input by obstetricians, cardiothoracic surgeons, anesthesiologists and neonatologists and detailed counselling and discussion with the couple. The woman did well and had an uneventful post-operative recovery. She was discharged on ninth day after surgery. She underwent cesarean section at 41 weeks of gestation for failed induction of labor and a healthy female baby was born with normal birth weight. Both mother and baby were discharged in a stable condition.

Keywords: atrial myxoma, pregnancy, cardiac tumors.

Background

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

The clinical manifestations are dependent on their 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 due to left ventricular inflow obstruction such as dyspnea, dizziness and syncope. [6-8] Other less common symptoms are neurological symptoms like stroke due cerebral emboli from the heart.[9]

Cardiac myxoma is rare in pregnant patients and have been reported in only 44 previous publications involving 51 patients. About 95% of them underwent transthoracic resection of the left atrial mass.[10] The indications for resection are the potential risk of embolic events and sudden death caused by myxoma obstructing the valve orifices.[11] Few previous publications suggested differing the surgery till after delivery or performing it in late pregnancy but outweighing the risks and benefits and with close monitoring and under medical management throughout pregnancy.[12,13]

Open heart surgery requires cardiopulmonary bypass which the standard therapy in such cases 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 flow.[14] Maternal mortality rate varies in the literature from 1% to 5%, with an average of 2.5% and does not differ from 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 and his group reported a case of myxoma resected two weeks postpartum using right anterior thoracotomy in order for the mother to have less pain and be able to hold and breastfeed her child.[12]

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

Case presentation

A 29-year-old woman Gravida 3 Para 2 woman presented to the cardiology clinic at 28 weeks +2 days of gestation, with a 2-month history of shortness of breath, increasing in severity in the preceding two weeks and associated with orthopnea and intermittent palpitations. She had two previous uneventful vaginal deliveries with gestational hypertension and gestational diabetes complicating both previous pregnancies. Her past surgical history was uneventful apart from laparoscopic appendectomy and cholecystectomy which were uncomplicated. Current pregnancy

was 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. She had a trans esophageal echo from another hospital at 26 weeks of gestation in view of her cardiac symptoms which showed a left atrial myxoma 3.2x2.7 cm attached to the interatrial septum with intermittent brief mitral in flow obstruction and Ejection fraction of 58%. She did not require any medications from cardiac point of view. She was given dexamethasone 6 mg 12^{th} hourly for 2 days for fetal lung maturity at 29 weeks of gestation. Further plan of management was discussed in a multidisciplinary meeting involving a cardiothoracic surgeon, a cardiologist, an obstetrician, a neonatologist and a cardiac anesthesiologist. Her echo was repeated in our hospital 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 28x27mm was noted in the left atrium with a thin stalk attaching to the inter atrial septum .The heterogeneous density and over all 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 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.

After discussing all possible options with the patient and her spouse including conservative management versus an urgent surgical procedure, it was decided to proceed with surgical excision of the myxoma. Fetal monitoring in the form of cardiotocogram (CTG) to be done prior to and after the surgical procedure to check for fetal wellbeing.

Outcome and follow up

The woman underwent left atrial mass excision at 30 weeks of gestation under general anesthesia. A median sternotomy incision was performed. Cross clamp time was 64 minutes. Surgery went uneventfully and she was shifted to the postoperative cardiac care unit (CCU) for monitoring. CTG post procedure showed fetal tachycardia and reduced beat to beat 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 8th day post excision, she had a bedside transthoracic echocardiogram, which revealed an intact interatrial septum. No myxoma remnant was seen. She was discharged on 9th postoperative day in a stable condition. She was advised to follow up the pregnancy at her regional hospital. She remained asymptomatic and at 41 weeks of gestation underwent a cesarean section for failed induction of labor. The outcome was a female baby with a birth weight of 3060 grams and an Apgar score of 9 and 10 at 1 and 5 minutes. Both mother and baby were discharged on the third day in a stable condition.

Discussion

Cardiac myxomas are commonly found on the left side and the patient usually presents with dyspnea such as in our case.[6-8] It is worth noting that dyspnea during pregnancy and 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. Therefore, they can be easily missed for the same reason. 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 reported three pregnant women diagnosed with cardiac myxomas, out of which two were complicated with cerebral infarctions requiring urgent surgical resection of the tumor.[11] Liu et al also reported a similar case that was complicated with cerebral infarction and retinal artery occlusion.[19] In pregnancy, and to balance the maternal and perinatal risks, the timing of surgical excision is important. Postponing surgical resection till after delivery will depend on the gestational age of pregnancy, worsening of the patient's symptoms and the findings of echocardiography. Traisrisilp et al reported a large cardiac myxoma of 9 cm in size diagnosed at 28 weeks that was removed 2 weeks postpartum.[20] The woman was diagnosed with a left atrial myxoma 2 years earlier but she lost to follow-up. She was evaluated during pregnancy and the transthoracic echocardiography showed a 9 cm mass in the left atrium obstructing mitral valve inflow, interfering with mitral valve closure, causing severe mitral regurgitation and severe pulmonary hypertension. Since the patient was asymptomatic and the couple wished to continue the pregnancy, the decision was made to deliver her at 32 weeks. The woman gave birth vaginally,

however she developed pulmonary edema during intrapartum period which was treated medically and 2 weeks postpartum she underwent the resection without complications.

In our patient, the initial plan was to delay the surgery to ensure that the fetus will be more mature. But a repeat Echo done after one week of admission were worrisome as the myxoma was resulting in a moderate degree of restriction to diastolic LV filling at 29 weeks of gestation which is likely to get worse with the expected plasma volume expansion 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 catastrophic potential complications to the fetus and the mother.

Thus, the decision was made by the multidisciplinary team to proceed with the surgery as soon as possible. During surgery, the patient is particularly at risk of circulatory obstruction, hypotension and probably vulnerable to prolonged operative time and compromise of the placental blood flow, leading to premature labor, non-reassuring fetal status, long term disabilities and fetal loss.[10] Open heart surgery as well as the use of cardiopulmonary bypass may cause premature labor and endanger the baby. The fetal mortality rate during maternal cardiac surgery with cardiopulmonary bypass is approximately 18.6%[15,16.]The obstetrician decided to forego fetal monitoring intra operatively as non-reassuring fetal status are common during cross clamp time and intervention can cause more harm than good to both mother and the fetus. Fetal heart was heard before and after the procedure. CTG was done when the patient was shifted to CCU which also showed reduced beat to beat variability which settled by itself. We did not act on that also as other features in CTG were reassuring and patient was on continuous intravenous infusion of morphine and the reduced beat to beat variability could be explained by that. As expected, the CTG returned to normal once morphine infusion was stopped. The patient had an uneventful post-operative recovery followed by a normal antenatal period.

Yuan and his group reported the largest review on cardiac myxoma in pregnancy including 44 articles and 51 patients.[10] In their review, the most common symptoms were dyspnea and palpitations and the cardiac myxoma was diagnosed more in the second trimester, similar to our patient. Transthoracic echocardiography was the most common technique used for diagnosis as in our patient. Most women underwent the myxoma resection during pregnancy (95.9%) with 47.2% of them resected in the third trimester and resulted in good maternal and fetal -neonatal outcomes.

They concluded that proper timing of cardiac surgery, being non-emergency in nature, shorter operation duration and improved cardiopulmonary bypass conditions may result in even better maternal and feto-neonatal outcome.[10]

Conclusion

Cardiothoracic surgery in pregnancy especially in the third trimester is associated with termination of pregnancy along with it. However, in this patient we have continued the pregnancy till term from her cardiac surgery at 30 weeks with efficient input from the multidisciplinary team. The authors have no conflict of interest.

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