CASE REPORT

Huge cardiac myxoma in pregnancy

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SUMMARY

A 28-year-old pregnant woman presented at 28 weeks of gestation. She was diagnosed to have a left atrial myxoma 2 years earlier, but was lost to follow-up. During this pregnancy, 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. However, there were no clinical signs of pulmonary and systemic congestion or obstruction. Based on the clinical symptoms of the patient, the echocardiographic findings and the term of her pregnancy, the patient decided to schedule for a vaginal delivery with surgical correction after delivery. She gave birth at 32 weeks of gestation. During labour, pulmonary oedema developed but was detected early and it responded to therapy. Two weeks after delivery, a right anterior thoracotomy was performed to facilitate the removal of the left atrial myxoma and repair of the mitral valve.

woman. The treatment involved a multidisciplinary approach and the outcome was successful.

CASE PRESENTATION

A 28-year-old hill tribe woman from Northern Thailand, gravida 4 para 3 in the last 6 years, first presented at 28 weeks of gestation for cardiac disease evaluation. All of the three previous pregnancies were unremarkable with uneventful deliveries. Two years prior to this current pregnancy, she had symptoms of progressive dyspnoea without orthopnoea and was diagnosed with left atrial myxoma and scheduled for surgical correction. However, she was lost to follow-up because of associated economic problems. The current pregnancy was unplanned and there had been no antenatal care.

INVESTIGATION

At the first visit, transthoracic echocardiography revealed a moderately dilated left ventricle with borderline systolic function, a dilated left atrium with a large atrial mass 9×4 cm attached to the interatrial septum by a stalk and obstruction of the mitral valve inflow with a mean diastolic mitral valve gradient of 5 mm Hg (figure 1). The mass also interfered with proper mitral valve closure, causing severe mitral regurgitation. Severe pulmonary hypertension (estimated systolic pulmonary pressure of 65 mm Hg) with borderline right ventricular systolic function was also noted. However there were no clinical signs of pulmonary or systemic congestion. Her cardiac status was considered as high risk for pregnancy.

Obstetric ultrasound revealed a normal fetus consistent with 28 weeks of gestation and growth was within normal limits.

TREATMENT

After counselling, the couple opted to continue the pregnancy and the women was closely monitored for cardiac decompensation and fetal well-being by the multidisciplinary team (cardiac surgeons, cardiologists, anaesthesiologists, obstetricians and neonatologists). She was scheduled for vaginal delivery at 32 weeks of gestation or caesarean section based on obstetric indications, and surgical correction after delivery. In case of maternal heart failure or haemodynamic deterioration, emergency caesarean section with surgical correction was planned to be performed in the same setting. Fetal surveillance with non-stress testing and umbilical artery Doppler was carried weekly.

BACKGROUND

Cardiac tumours are very uncommon. Myxomas are the predominant cardiac tumours in adults, occurring in adults and occur more frequently in women between the third and sixth decade of life; therefore, they are occasionally encountered during pregnancy.^{1,2} The clinical manifestations are usually associated with their size, mobility and location.² The most common symptoms are constitutional symptoms such as myalgia, fever, fatigue and weight loss. About half of the cases present with symptoms due to intracardiac obstruction symptoms such as dyspnoea, dizziness and syncope.²⁻⁴ Other less common symptoms are cerebral symptoms due to the ischaemia secondary to emboli.⁵ Cardiac myxoma can be life threatening and usually requires urgent surgical removal to prevent potentially serious emboli and haemodynamic deterioration or even sudden death.⁶ Cardiac myxomas during pregnancy are even rarer and have been reported in only 44 previous publications involving 51 patients.⁷ Several physiological changes in the cardiovascular system during pregnancy can theoretically worsen the disease and threaten maternal and fetal life. The overall perinatal mortality of pregnant women undergoing cardiac surgery is, to date, nearly 20% with the risk increasing in cases of cardiopulmonary bypass adding to the concern surrounding the case of this patient.⁴ The aim of this report is to describe this case, which to our knowledge of the largest cardiac myxoma, ever reported in a pregnant

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Figure 1 Preoperative echocardiography showed a 9×4 cm hyperechoic lesion occupying the left atrium which interfered with the closure of the mitral valve.

OUTCOME AND FOLLOW-UP

At 32 weeks of gestation, after corticosteroid administration to encourage fetal lung maturity, induction of labour with membrane stripping and oxytocin was performed. The fetus tolerated the procedures well. The patient had a normal delivery, giving birth to a healthy preterm male baby weighing 1900 g, Apgar score 7 and 9 at 1 and 5 min. During labour and postpartum, haemodynamics were monitored by the cardiologists and anaesthesiologists. Pulmonary oedema developed during labour but was detected early and responded to fluid restriction and diuretic drug. The patient was stable at functional class I–II in the postpartum period.

Two weeks after birth, she underwent a minimally invasive right anterior thoracotomy to enable removal of the left atrial myxoma and repair the mitral valve with an annuloplasty ring. Images of the gross and microscopic examination are shown in figures 2 and 3, respectively. The right anterior thoracotomy approach was chosen instead of a sternotomy so the patient would not be subjected to the postoperative limitations of a sternotomy in particular facilitating her ability to hold her child in the first 3 to 6 months after surgery. Immediate postoperative echocardiography showed left ventricular dysfunction with an ejection fraction of 20% but this gradually improved, a left ventricular ejection fraction of 42% being registered 2 months

Figure 2 Gross examination revealed a pedunculated soft gelatinous grey-tan mass (9 cm at greatest dimension) with a short stalk. Cut sections showed a variegated appearance with a gelatinous yellow-white cut surface and areas of haemorrhage and degeneration.

Figure 3 Microscopic examination showed polygonal, bipolar or stellate cells embedded in an abundance of myxoid substance. These cells have bland cytology and are usually present as single cells, or anastomosing cords or nests (H&E, ×200). 1 February 2017.

after surgery. Neurohormonal blockade medication for heart failure was prescribed. She chose progestin-only pills for contraception.

DISCUSSION

Cardiac myxomas are usually sporadic and occasionally recur. Ninety per cent occur in the atria and 75%–80% in the left side,² as found in this case. Myxomas usually originate from the interatrial septum. Based on a comprehensive review including all 44 relevant publications,⁷ surgical resection of a tumour during pregnancy is a relatively safe procedure, possibly due to the short operative time. Fetal outcomes were no different between the treatment options, timing of cardiac surgery or delivery modes, though a caesarean section was carried out in most cases. Timely surgical removal and improved cardiopulmonary bypass may be associated with better obstetric outcomes. The tumour size was also directly associated with the severity of obstruction. Note that the mean size of tumours in previous reports was 55.1 ± 24.5 mm (median 55 mm), while the size of the tumour in our case was 90×40 mm.

There is a close association between high perinatal mortality rate and preterm delivery,^{7,8} so if at all possible, delivery should be postponed to as late into the pregnancy as possible leading to maximum fetal maturity. However, 30%–40% of cases have been found to have an embolic episode in the systemic arterial circulation producing ischaemic or haemorrhagic cerebrovascular events,² and causing haemodynamic deterioration from an obstructive-like lesion. In such situations, an urgent surgical removal of the tumour is warranted. As there are a limited number of cases in the literature, it is unclear whether surgical removal should be performed during pregnancy or after delivery when cardiovascular changes secondary to pregnancy become normal. However, in many cases, the choice to interrupt the pregnancy and follow-up with cardiac surgery was made.^{9,10}

To manage the balance between the maternal and perinatal risk, the timing of surgical treatment is important. If diagnosis is made in the first trimester, our recommendation is that the surgery should be carried out early in the second trimester.^{11,12} If the diagnosis is made after the first trimester but before the late third trimester, surgery should be carried out without delay. If the

diagnosis occurs late in the third trimester, delivery by caesarean section should be carried out once fetal maturity has been established and the mass should be excised in the postpartum period.^{3 11} Compared with other cases in previous reports, our case was likely to have a higher risk for both mother and fetus because of the very large nature of the tumour with pulmonary hypertension at the first visit. This made the patient particularly at risk of circulatory obstruction and probably vulnerable to prolonged operation time and compromise of the placental blood flow, leading to a non-reassuring fetal status. However, our patient had no clinical symptoms of embolic events for 2 years after the diagnosis suggesting that the tumour was less friable and the lack of haemodynamic deterioration reduced the need for emergency surgery. In this case, the huge myxoma in the left atrium with evidence of pulmonary hypertension mean that a caesarean section would be potentially catastrophic if an intra-partum or postpartum haemorrhage occurred, possibly leading to low cardiac output. On the contrary, increased venous return immediately postpartum could aggravate pulmonary oedema which had already developed prenatally.

The processes of managing our patient are as follows: (1) prolonging of the pregnancy time for as short a period as possible to avoid potential embolism or further deterioration of pulmonary hypertension; (2) waiting for fetal lung maturity, that is, 32 weeks of gestation when our neonatal intensive care unit would give care with a high possibility of success; (3) close monitoring of the mother and fetus with the team always available for early detection and intervention for cardiac decompensation or sudden haemodynamic changes; (4) labour induction

with intensive monitoring rather than caesarean section to avoid more haemodynamic changes during the operation and to reduce the chance of unexpected excessive intraoperative bleeding and (5) surgical removal of the tumour after delivery when maternal haemodynamics returned to non-pregnant state.

The management of this case was very challenging as concern regarding cardiac failure, pulmonary oedema or severe left heart obstruction could develop at any time as an increase in the cardiac output associated with normal pregnancy may possibly not be achieved, especially in late pregnancy, during labour or immediately postpartum. The case presented here illustrates the successful outcome of a multidisciplinary approach in treating a life-threatening cardiac disorder during pregnancy.

Contributors KT: drafting and revising the manuscript. RK: writing manuscript in the part of echocardiography. NT: writing manuscript in the part of surgery. SL: writing manuscript in the part patient care during labour.

Competing interests None declared.

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Learning points

- Cardiac myxoma during pregnancy is a rare condition in which the physiological haemodynamic changes associated with pregnancy can increase the threat of the disease and also increase the risk to maternal and fetal life.
- There is no consensus about the timing of surgical treatment. A balance between maternal and perinatal risk is a critical and needs to be discussed with the patient.
- A multidisciplinary approach is essential in the formulation of both the treatment plan and patient care to maximise the chances of a good outcome.

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