Intracardiac myxomas comprise 50% of all benign cardiac tumours and have an estimated incidence of 0.5 per million population per year. Although the vast majority of myxomas (75%) occur in the left atrium, they are infrequently associated with mitral valve pathology. There have been very few case reports of left atrial myxoma associated with mitral valve pathology,1,2 and, to our knowledge, no such case has been reported in a pregnant patient.

Here we describe the case of a left atrial myxoma associated with mitral valve regurgitation in a pregnant patient, stressing the importance of transoesophageal or transthoracic echocardiography, preoperatively and during the operation, as a useful tool to rule out any associated valvular damage or mitral annular dilatation as an underlying cause of mitral regurgitation.

Case presentation

A 29-year-old Ghanaian lady presented with a 6-month history of palpitations, fatigue and increasing exertional dyspnoea. She was confirmed 6 weeks pregnant on ultrasound scan and routine blood investigations were all within normal limits. TTE showed a large pedunculated mass in the left atrium, attached to the interatrial septum, with an appearance suggestive of atrial myxoma. Mild mitral regurgitation (MR) was noted, which was thought to be due to the tumour prolapsing through the mitral valve. Left ventricular and left atrial dimensions were normal and left ventricular systolic function was good, with an ejection fraction of 60%. The patient was transferred to our cardiothoracic surgical centre for urgent surgery.

Intraoperative TOE confirmed the presence of a mass in the left atrium measuring 3.6 cm × 5.7 cm. The mass was attached to the interatrial septum, slightly inferior to the fossa ovalis and approximately 1.2 cm from the mitral annulus. The echocardiographic appearance of the large mass was typical of atrial myxoma. (Figure 1). There was mild MR associated with the prolapse of the tumour through the mitral valve causing a coaptation defect of the leaflets (Figure 2). There was thickening and possible calcification of the anterior (A2, A3) mitral leaflet. There was no prolapse of the mitral leaflets and the mitral annulus diameter was measured at 4.1 cm.
Through a median sternotomy, cardiopulmonary bypass was established using bicaval cannulation. The heart was arrested using antegrade cold-blood cardioplegia and the tumour was approached through a transeptal incision. A large smooth pedunculated mass with typical external appearances of a myxoma was encountered and was excised with its stalk from the interatrial septum (Figure 3). The interatrial septum and the roof of the left atrium were closed with 3-0 prolene and the right atrium was closed with 5-0 prolene. The patient was weaned from cardiopulmonary bypass without any difficulties. Intraoperative TOE demonstrated no residual left atrial mass and an intact interatrial septum with no shunting. However there was severe mitral regurgitation with a posteriorly directed jet (Figures 4 & 5) and a central jet. Thickening of the anterior leaflet and poor coaptation of the leaflets were noted. There was no mitral valve prolapse. Mitral annular dilatation was measured at 4.1 cm. The left ventricular function remained good and the patient maintained excellent haemodynamic stability.

Given the complex issue of valve choice (mechanical vs. bioprosthetic) in a pregnant woman should a valve replacement be necessary, and the patient’s excellent haemodynamic stability, it was decided that the best course of action was to transfer the patient to the intensive care unit. She was extubated the same evening and her condition and possible treatment options were discussed with her the next day, when she had regained complete consciousness. She agreed to undergo mitral valve repair and receive a bioprosthesis if required. Repeat surgery was performed the following day and the valve was approached using the previously performed transeptal incision. The anterior leaflet appeared thickened, with no palpable calcification, and the posterior annulus was dilated. The left atrium was not enlarged. A 32 mm Cosgrove annuloplasty band was implanted to reduce the size of the posterior mitral valve annulus. The patient was weaned from cardiopulmonary bypass easily and repeat TOE showed only mild residual MR (Figure 6).

The patient made an excellent recovery and there were no complications other than intermittent episodes of complete heart block, for which a permanent pacemaker was implanted three weeks postoperatively. Following that, she was discharged home. Due to her pregnancy she was not started on warfarin or aspirin and she remained free of thromboembolic incidents throughout her pregnancy. Histology
confirmed an atrial myxoma measuring $5.5 \times 2.5 \times 4.0$ cm. Her pregnancy remained viable and she delivered a healthy female infant in the 37th week via a caesarean incision. At one-year follow up she remains with excellent cardiac function and mild MR.

Discussion

The first case of successful surgical excision of left atrial myxoma was described by Crafoord in 1955. There are four case reports in the literature of left atrial myxoma removal during pregnancy. To our knowledge, there has been no previous case report of left atrial myxoma associated with mitral valve pathology in a pregnant patient requiring mitral valve repair.

Primary tumours of the heart are uncommon, with an estimated incidence of 0.17% to 0.19% in selected autopsy series. Of these tumours, 75% are benign and approximately 50% of these are myxomas. Myxoma is a neoplasm of unknown histogenesis, occurring most often in middle-aged, usually female patients, readily amenable to treatment, and with an excellent postoperative prognosis. Prompt diagnosis and intervention are important, as potentially fatal obstructive and embolic complications may occur.

Most myxomas (75%) arise inside the left atrium from the interatrial septum at the border of the fossa ovalis. Grossly, about two-thirds of myxomas are round or oval shaped with a smooth or slightly lobulated surface. Pinede et al, in their case series, reported that the average size of the myxomas varied from 1 to 15 cm. Most patients with myxoma present with symptoms of obstructive mitral valvular disease (mitral stenosis), congestive heart failure, signs of embolisation, systemic or constitutional symptoms of fever, weight loss or fatigue, and immunological manifestations of myalgia, weakness, and arthralgia. Central nervous system symptoms usually call for CT or MRI of the brain to exclude embolisation or aneurysm formation, which is sometimes associated with myxomas. It has been suggested that it is prudent to perform MRI of the brain in all cardiac myxoma cases to exclude central nervous system manifestations.

In our case, the patient was a 29-year-old pregnant female who presented with dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea, fatigue, and pain in her left arm. The actual dimensions of the tumour were $3.6 \text{ cm} \times 5.7 \text{ cm}$, in accordance with the usual dimensions described in large series (mean 5 cm). In these aspects, our patient fit within the classic picture of atrial myxoma.
There have been two case reports of left atrial myxoma with annular dilatation in the presence of left atrial and left ventricular enlargement. The rare feature in our patient was the presence of mitral annular dilatation without left atrial and left ventricular enlargement. This is an unusual finding and has rarely been reported previously in the literature. To our knowledge, this is the first of its kind to be reported in a pregnant patient.

Though mitral regurgitation is a frequent occurrence preoperatively, damage to the mitral valvular apparatus requiring mitral valve repair or replacement is rare. The striking feature in our patient was the acquired structural anomaly of the mitral valve, namely the posterior annular dilatation without chamber enlargement. From the TOE after tumour removal, the mitral regurgitation—which was initially thought to be due to tumour prolapse into the mitral valve—was actually mainly due to a coaptation defect secondary to posterior annular dilatation (Figure 4).

The possible causes of mitral regurgitation in a patient with left atrial myxoma are tumour prolapse causing volume overload, leading to left atrial and left ventricular dilatation and annular dilatation, and possible mechanical trauma to the mitral valve leaflets. In our patient the mitral valve looked absolutely normal, apart from mild thickening of the anterior leaflet, and there was no prolapse. The most likely explanation for this unusual finding is the repeated mechanical stretch of the mitral annulus by tumour prolapse into the mitral valve during systole and diastole (Figures 1 & 2). This patient had worsening dyspnoea with repeated episodes of paroxysmal nocturnal dyspnoea prior to admission. This could be due to an increase in cardiac output and blood volume expansion secondary to pregnancy in the presence of severe mitral regurgitation and the tumour obstruction.

In this patient, TOE played a significant role in diagnosing the annular dilatation as a cause of mitral regurgitation. Classical teaching suggests that simple excision of the left atrial myxoma resolves the symptoms and is usually the only treatment needed. In our experience this was not the case. It may be prudent to examine the mitral valve apparatus, irrespective of the chamber size, and discuss all the possible options for treatment in relation to the patient’s clinical condition before proceeding with surgical removal. This examination should be performed either with TTE, or if need be with TOE, in order to identify the occasional valvular pathology. Thus, the subsequent operation can be planned more accurately. In our case, despite the definite secondary mitral valve pathology, both chambers were of nearly normal size. To our knowledge, this is very uncommon.

The patient was not started on either warfarin or aspirin postoperatively. The decision was based on her pregnancy status. The current European Society of Cardiology guidelines support the use of warfarin for 3 months post-mitral repair, citing an absence of studies supporting the safety of omitting warfarin. Thus the decision was not fully justified, but the patient remained free of thromboembolic incidents and the infant was a healthy one.

The authors conclude that TOE is an invaluable tool in the diagnosis and understanding of the exact location of the neoplasm, associated mitral valve pathology, and the diagnosis and mechanism of mitral regurgitation after excision of a myxoma. Prompt understanding of the integrity of the mitral valve apparatus should be acquired before the operation, using either TOE or TTE, so that the surgical procedure may be planned accordingly. With hindsight, the other lesson to learn is that it may be prudent to examine the mitral valve apparatus as a routine procedure after tumour excision and discuss with the patient the possibility of mitral valve repair, with or without valve replacement, before proceeding with left atrial myxoma excision.

References

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