CASE REPORT

Right atrial myxoma mimicking tricuspid stenosis

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Accepted 28 July 2015

SUMMARY

A 62-year-old man presented to the cardiology clinic with symptoms of fatigue, shortness of breath and swelling in the abdomen and legs. A pedunculated mobile mass with a short stalk in the right atrium was found to originate from the inferior vena caval opening and prolapse into the right ventricle through a tricuspid valve during diastole in echocardiography. The patient was referred to our department and surgery was planned for right atrial myxomas diagnosis. The mass with surrounding tissue was surgically removed using the cardiopulmonary bypass method. No postoperative complications were seen in the currently asymptomatic patient who is still under our follow-up schedule.

BACKGROUND

About more than half of the benign tumours diagnosed in the heart are myxomas. Development of cardiac myxomas are usually sporadic (90%) and seen in the left atrium in 60–80% of the patients. Cardiac myxomas are located in the right atrium in 15–28% of cases, in the right ventricle in 8% of cases, in the left ventricle in 3–4% of cases and rarely in both atriums. In addition, myxomas can originate from mitral leaflets, aortic valves and pulmonary vessels in rare cases.¹ Right-sided myxomas have a wider base on the atrium wall or septum compared with left-side myxomas. Although myxomas can occur in any age group, they are more frequently reported in females between 30 and 60 years of age. Myxomas are usually associated with cardiac symptoms associated with obstruction and embolic episodes. The most common symptom diagnosed in patients with atrial myxoma is shortness of breath caused by obstruction.²

Even though cardiac myxomas are mostly considered as benign tumours in histopathological classification, they require surgery as soon as they are diagnosed in patients due to their possible mortal complications. The standard treatment method is complete surgical excision of the tumour with surrounding healthy tissue.³

In this study, we present a relatively rare right atrial cardiac myxoma case with an unusual location (a pedunculated mobile mass with a short stalk in the right atrium, originating from the inferior vena cava and prolapsing into the right ventricle through a tricuspid valve during diastole) and mimicking tricuspid stenosis symptoms.

CASE PRESENTATION

A 62-year-old man presented to the cardiology department with symptoms of shortness of breath, fatigue, abdominal pain and swelling in the

abdomen and feet, which were ongoing for more than 1 month. Transthoracic echocardiography revealed ejection fraction as 70%, ventricle functions as normal, no detected pathologies in the aorta, mitral and tricuspid valves, an increased right atrium diameter (6.6 cm) and a mobile mass connected to the lower part of interatrial septum in the right atrium. Estimated pulmonary artery pressure was 70 mm Hg. The patient's transoesophageal echocardiography showed a 52×60 mm mobile mass which fills inside of the right atrium, pedunculated to the inferior vena cava (**figure 1**) with a short stalk that prolapses into the right ventricle during diastole through a tricuspid valve with bleeding areas (**figure 2**). The patient was diagnosed with an atypically placed myxoma and coronary angiography was indicated. During coronary angiography, the coronary arteries seemed normal with vascularisation of the tumour from the right coronary artery (RCA) (**figure 3**). Owing to severe tricuspid stenosis symptoms, the patient was referred to our department for immediate surgery. During physical examination of the patient, heart rate was 85 bpm and arterial blood pressure was 110/70 mm Hg. Pretibial oedema in the bilateral lower extremity, abdominal swelling and palpable hepatomegaly was also detected. During heart auscultation, tumour plop was heard during mesocardiac focus and widespread bilateral basal crackles were heard during lung auscultation. ECG showed normal sinus rhythm with no pathology. Bilateral pleural effusion was present with emphasis on the right side on posteroanterior chest X-rays. Abdominal ultrasonography showed medium-grade hepatomegaly. Intrahepatic veins were seen as larger than normal, possibly due to cardiac reasons.

Figure 1 Right atrial myxoma view of transoesophageal echocardiography (IVC, inferior vena cava; RA, right atrium; RV, right ventricle).

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To cite: Şaşkın H, Düzyol Ç, Özcan KS, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-210818

Figure 2 Entrance of right atrial myxoma to right ventricle through tricuspid valve view in transesophageal echocardiography (LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle).

Venous thrombosis was not detected in bilateral lower extremity venous Doppler ultrasonography. Laboratory results showed sedimentation rate as 112 mm/h, haematocrits as 30.4% (normal range 40–52%), haemoglobin as 10 g/dL (normal range 13.0–17.5), white cell count as 13 600 mm³, hypochromic microcytic erythrocyte morphology, and C reactive protein value as 22.8 mg/dL (normal range 0–0.5). Informed consent was taken from the patient. Under general anaesthesia, median sternotomy was performed. Following pericardiectomy, cardiopulmonary bypass was initiated with aortic and superior vena cava cannulation. Cardiac arrest was induced using medium-level hypothermia, hyperkalaemic antegrade blood cardioplegia and aortic cross clamps. The right atrium was opened after occlusion of superior and inferior vena cava with snare. The mass filling the right atrium was visualised and an inferior vena cava cannula was inserted. A red-coloured, gelatinous, short-stemmed mass with bleeding foci, pedunculated between the inferior vena caval opening, coronary sinus ostium and interatrial septum with a size of 6×5×4 cm, was removed in one piece with the surrounding and pedunculated tissue ([figure 4](#)).

Figure 3 Neovascularization view of right atrial myxoma from right coronary artery.

Figure 4 Myxoma removed from the right atrium.

The defect caused by the removal was closed using a pericardial patch. Ligation of the greater branches coming from the RCA which feed the tumour was performed. The vessels from the RCA were also ligated. The operation was successfully completed without any complications. The patient was discharged after 5 days without any postoperative complications. The specimen was sent to the pathology department for analysis. Pathology results reported the mass as a benign myxoma with stellate cells and isolated immature spindle-shaped cells seen within loose stromal embryonic connective tissue stromal cells which are covered in part with a single layer flat epithelium and in part with bleeding capsules ([figure 5](#)).

We followed up the patient with clinical and echocardiographic examination at 1, 3, 6 and 12 months.

The patient had no symptoms and recurrences on echocardiography.

DISCUSSION

Primary cardiac tumours are quite rare. Seventy-five per cent of heart tumours are benign while 25% of them are malignant. The most common benign tumours of the heart are myxomas. Myxomas are divided into two forms as sporadic and familial form. Sporadic form is the most frequently diagnosed form, whereas the familial form is reported to be around 7% of the cases.⁴ Myxomas are usually placed in the left atrium. More rarely, they are seen in the right atrium, ventricles and heart valves. In autopsy series, right atrial myxomas were reported to have a low incidence rate. One of such reports is the Mayo Clinic series which consists of 23 673 patients between 1954

Figure 5 Histopathological view of the removed myxoma (H&E stain, ×200 magnification).

and 1979. In all those patients, 41 (0.17%) were diagnosed with primary heart tumours and 28 were diagnosed with myxomas. Seventeen of those myxomas were placed within the left atrium and four patients had (14%) right atrium myxoma.⁵

Myxomas found in the right atrium are usually pedunculated to the right atrium wall, atrial septum or fossa ovalis with a wide base and include calcification and bleeding foci.² Cardiac myxomas can present a wide range of different clinical symptoms depending on their place, size and mobility. Patients might show different symptoms according to tumour-caused obstruction and cerebral or peripheral embolism caused by myxoma or increased inflammatory response. Even though right atrial myxomas can be asymptomatic, right heart failure (dyspnoea, haemoptysis), venous congestion symptoms (hepatomegaly, acid and peripheral oedema), pulmonary hypertension and pulmonary embolism findings can also be seen in such cases.⁶ In our case, a red-coloured, gelatinous, short-stemmed mass with bleeding foci, pedunculated between the inferior vena caval opening, coronary sinus ostium and interatrial septum, with a size of 6×5×4 cm which caused right heart failure and venous congestion symptoms (shortness of breath, swelling in the abdomen and legs), right atrial myxoma was diagnosed.

Two-dimensional echocardiography is used to confirm all myxoma diagnoses in patients. Advanced imaging techniques such as CT, MRI and coronary angiography can also be employed for additional diagnosis tools. During daily clinical practice, atrial myxomas are usually confused with valve diseases, infective endocarditis, coronary artery disease, pulmonary embolism, pulmonary hypertension, malignancy, intracardiac thrombus and rhabdomyolysis.³ We also diagnosed our patient using echocardiography.

It is not unusual to see vascularised structures of myxomas by vascularisation from coronary arteries. Vascularisation of myxomas detected during angiography is seen in 40% of the patients and this is usually found by chance during coronary angiography for coronary artery disease suspicion. Myxomas usually get arterial supply from right coronary artery, followed by circumflex artery. Moreover, myxomas that receive blood from both arteries were reported in the literature. Even though clinical significance of vascularised myxomas are not completely understood, they are thought to be sources of thrombus formation with acute bleeding within the tumour and embolism.⁷ Our patient's coronary angiography results showed vascularisation of the tumour during right coronary artery screening.

Myxomas should be surgically removed as soon as they are diagnosed due to risks of valve obstruction and systemic embolisation. Surgical treatment method is total excision of the mass with surrounding healthy tissue. The main point that should be considered in right atrial myxoma surgery is placement of the caval cannula near the tumour after pump insertion in order not to cause intraoperative tumour embolisation during cannulation. Myxomas usually end with full recovery following careful surgical excision and the non-presence of traumatised mass or tumour inoculation.¹ In our case, in order to minimise tumour embolisation risk, the inferior vena caval cannula was inserted after opening the right atrium following cardiac arrest considering the size and nearby placement to the inferior vena caval

opening of the tumour. Moreover, if the tumour's blood supply comes from the coronary artery, this can change the surgical strategy drastically. Like in our patient, surgical ligation of arterial branches is crucial, since incomplete identification and ligation of those branches may cause the free branches to empty into left atrium, causing myocardial ischaemia and cardiac steal syndrome development. Recurrence after surgical resection of primary lesions has been observed in 1–4% of sporadic and 12–22% of familial cases.⁸ The perioperative mortality ranges from 0% to 3% in multiple series.⁹

In our centre, we operated on nine patients with myxoma (7 originating from the left atrium, and 2 originating from the right atrium). None of these patients had recurrences, mortality and complications at follow-up.

In conclusion, right atrial myxomas are rare tumours which can cause tricuspid valve stenosis and right heart failure symptoms due to their location. In these cases shortness of breath, embolisation and symptoms of venous congestion are commonly observed and right atrial myxomas should be definitely considered during differential diagnosis.

Learning points

- ▶ Right atrial myxomas are rare tumours which can cause tricuspid valve stenosis and right heart failure symptoms due to their location.
- ▶ In these cases shortness of breath, embolisation and symptoms of venous congestion are commonly observed and right atrial myxomas should be definitely considered during differential diagnosis.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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