Right ventricle myxoma creating a partial obstruction of the pulmonary infundibulum: case report

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Citation

Abstract
A 13-year old female was referred to hospital for severe dyspnoea (stage III NYHA), with an oedema of the face and the lower limbs. She was then steadily putting on weight and gained up to 5 kilogram’s. The situation got worse and the patient referred to cardiovascular surgery. The interview indicated that there has never been a myxomatosis case in her family before. The physical examination revealed a worsening of the general health status and a clinical anaemia. However the pulse was regular, beating at 100 per minute; and the blood pressure was 100/60mmhg. There was no longer an oedema on the face, but the other bilateral pre-tibia oedema was still persistent and taking the gore. The examination also revealed a turgescence of the jugular veins and a hepatomegaly. The heart auscultation allowed a diagnosis of a pulmonary stenosis breath, which was not varying with the changes of postures. The patient did not present any neuro-endocrine clinical signs or any external skin problems. The chest cross section echocardiography revealed a cardiac tumour which implanted on the front lateral side of the right ventricle with an expansion into the pulmonary infundibulum. The patient was operated within 48 hours following her admission. The removal of the myxomatosis was achieved using technique of extra corporeal circulation (CEC). The heart was reached through a median sternotomy followed by a longitudinal right side atriotomy. The tricuspid valve was normal; the tumour was located across the tricuspid orifice. Time cross clamping was 15 minutes. The anatomy pathological examination showed a tumour myxomatosis-like tumour with two lobes measuring 4.2 x 2.3 centimeters. At its top there were zones and muscular tissue and hemorrhagic necrosis. The evolution was simple; the post operation scanning check ups were satisfactory. Right ventricle myxoma protruding into pulmonary artery is exceedingly rare. Obstruction of cardiac cavity is redoubtable complication of myxoma. We emphasize the importance of periodic follow up of young patient with echocardiography allowing early detection of recurrence.

INTRODUCTION
In cardiology tumours are very unusual and heart myxoma, which is generally a benign tumour, constitutes the most predominantly occurring cases. They actually represent 0.5 to 1 % of the tumours of soft tissues (1). The annual occurrence rate of this tumour is estimated at 0.5 / 1,000,000 / year (2) and it is usually located in the left auricle area (75%). It appears that its location in the right ventricle area is very rare (3).

Here is the case of a patient presenting a myxoma located in the right ventricle area creating subsequently a partial obstruction of the pulmonary infundibulum

PATIENT
The patient was a 13 years old girl who had been consulted at the NYHA (New York Heart Association) and diagnosed: stage III effort dyspnoea, with an oedema of the face and the lower limbs. Actually the early symptoms can be traced back to 6 months earlier and were discernable through a rather important effort dyspnoea. She was then steadily putting on weight and gained up to 5 kilogram’s. Two weeks ago, the situation got worse, requiring some cardiologic treatment. The patient referred to cardiovascular surgery. The interview indicated that there has never been a myxoma case in her family before. She was not feverish, but the physical examination revealed a worsening of the general health status and a clinical anaemia. However the pulse was regular, beating at 100 per minute; and the blood pressure was 100/60mmhg.

There was no longer an oedema on the face, but the other bilateral pre-tibia oedema was still persistent and taking the gore. The examination also revealed a turgescence of the
jugular veins and a hepatomegalia. The heart auscultation allowed a diagnosis of a pulmonary stenosis breath, which was not varying with the changes of postures. The patient did not present any neuro-endocrine clinical signs or any external skin problems. The chest x-rays showed a convexity of the right medium bow, a cardiothoracic index of 60 % and small-size left side pleurisy. The electrocardiogram indicated fast sinus rhythm.

The chest cross section echocardiography (figure 1) revealed an inner right ventricular bi-lobed tumour mass of 4.4 x 2 cm with a sessile basis.

**Figure 1**
Figure 1: the chest cross section echocardiography of the right ventricle myxoma.

That mass was implanted on the front lateral side of the right ventricle with an expansion into the pulmonary infundibulum, which it partially obstructs.

The right side cavities were dilated, and the right ventricle measured 44mm. The maximal gradient of right ventricle/pulmonary artery stood at 30mmhg whereas the average gradient stood at 15mmhg. The systolic function of the left ventricle was preserved and the ejection fraction was estimated at 70 %. The heart valves as well as the under valve located device were normal. Biology tests confirmed the clinical anaemia. The patient was operated within 48 hours following her admission. The exeresis of the myxoma was achieved using technique of extra corporeal circulation (CEC). That extra corporeal circulation system was installed in between two cava vena cannulas and an aorta cannula. The cold crystalloid cardioplegia was injected via the root of the aorta.

The heart was reached through a median sternotomy followed by a longitudinal right side atriotomy. The tricuspid valve was normal; the tumour was located across the tricuspid orifice. The operation confirmed the data collected from the scanning; i.e. that the mass was bi-lobed, and pedicle shaped. It was of pale pink colour, and dangling at the front right face of the right ventricle. As it can be seen (figure 2) the excision took away the pedicle.

**Figure 2**
Figure 2: surgical view of myxoma

The tricuspid valve was continent. After the operation, there was no difficulty stopping the external blood derivation system. The heart was beating at a sinus rhythm and the hemodynamic was stable. The CEC time span was of 238 minutes and the aorta time clamping lasted 15 minutes. The anatomy pathological examination showed a tumour myxomatosis-like tumour with two lobes measuring 4.2 x 2.3 centimeters. At its top there were zones and muscular tissue and hemorrhagic necrosis. The evolution was simple; the post operation scanning check ups were satisfactory. Check ups through scanners after 3 months, 6 months and one year showed no signs of recurrence.

**DISCUSSION**
Myxoma constitutes the most frequent case of heart tumours; it represents 50% of benign heart tumours (4, 5). Myxoma of the ventricle is rare. The occurrence of heart myxoma within patients of young age should make the doctor suspect the Carney syndrome. It is a syndrome that can be identified as follows: myxoma diversely located, together with external skin problems (skin and mucous membrane lentiginosis, schwannomis and skin myxomatosis) and endocrine tumours (6). Such symptoms have not been noticed in the case of our patient, and there has never been a myxoma case in her family either.
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In general it is located in the left auricle area (75%), statistics also indicate that 23% of myxoma is located in the right auricle area whereas 2% is located in the ventricles on an equal distribution basis between the right and the left ventricle (7). The gravity of the myxoma is a function of the risks of an arterial embolism, pulmonary embolism, but also of the level of risks of obstruction of heart cavities or pulmonary obstruction in the right side locations. These potential risks for embolism occurrence correlates with the type of myxoma. There are two types of polypoid myxoma with crunchy surface and the myxoma with smooth surface. Polyploid tumour presents a higher potential risk for embolism. The size of the tumour is one factor that increases the potential risk for embolism (8). Our patient presented a large tumour (4.4 x 2cm), but the surface of the tumour was not that crunchy. The surgical exeresis has to be made with precaution; and any inconvenient manipulation of the tumour should be banned. There is a potential risk of pulmonary obstruction during the anaesthesia induction phase and during the sternotomy. The risk of splitting up the mass is real during the fixing of cannulas into the right auricle, if the tumour tends to pass into the auricle through the tricuspid valves.

We chose to set cannulas directly in the upper vena cava and then the lower vena cava, leaving the right auricle free, so as the clampage of the pulmonary artery turned to be unnecessary to the process. The right side atriotomy allowed us to expose the tumour through the tricuspid orifice; the re-sectioning was effected without damaging the ventricle wall. Late relapses are rare regarding treatment of the simple forms if the re-sectioning is complete. Castello and col (9) noted 0.4 in 5 % of recurrences occurring 22 years after surgery. However if previous myxoma cases had occurred in the family, the rate of recurrence is 40 %. Turner recurrences increase with young age, multiplicity in the location of the tumour and the family medical background. That is the reason why a follow-up through regular scan is practised for this young patient.

CONCLUSION

Myxoma of the right ventricle is rare, and to avoid the risk for pulmonary obstruction due to the tumour we decided to achieve a semi emergency excision. The age of the patient as well as the location of the tumour prompted us to practise a regular surveillance to watch for recurrences.

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