

Case Report

Right Ventricular Myxoma

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Abstract

We present a case of a right myxoma in a 13-year-old boy. Echocardiography revealed a huge mobile heterogeneous mass, suggestive of myxoma, in the right ventricle (RV) protruding across the tricuspid valve into the right atrium (RA), causing partial RV inflow obstruction with a sessile mass around the tumor mass, suggestive of thrombus. The masses were completely removed under cardiopulmonary bypass. The RV masses were histo-pathologically confirmed to be a myxoma and a thrombus, respectively.

Introduction

A myxoma is a myxoid tumor of primitive connective tissue [1] It is the most common primary tumor of the heart in adults, but can also occur in other locations. 75% of all myxomas are found in the left atrium, and almost all of the myxomas are found in the right atrium. It is very rare for a myxoma to be found in either of the ventricles. The tumor takes one of two general shapes: a round, firm mass, or an irregular shaped, soft, gelatinous mass. They are attached to the endocardium, the inside lining of the heart.

The cells that make up the tumor are spindle-shaped cells and are embedded in a matrix rich in mucopolysaccharides (a group of carbohydrates). Myxomas may contain calcium [2], which shows up on x rays. The tumor gets its blood supply from capillaries that bring blood from the heart to the tumor. Thrombus may be attached to the outside of the myxoma. There are three major syndromes linked to myxomas: embolic events, obstruction of blood flow, and constitutional syndromes. Embolic events happen when fragments of the tumor, or the thrombi attached to the outside of the tumor, are released and enter the blood stream. Gelatinous myxomas are more likely to embolize than the more firm form of this tumor [3].

Myxomas may also obstruct blood flow in the heart, usually at a heart valve. The mitral valve is the heart valve most commonly affected. Blood flow restrictions can lead to pulmonary congestion and heart valve disease. Embolization can lead to severe consequences. In cases of left atrial myxoma [4], 40-50% of patients experience embolization. Emboli usually end up in the brain, kidneys, and extremities. Stroke is more common in young patients. Aseptic symptoms are often non-specific, minor embolic phenomena, such as transient visual loss and transient loss of consciousness are often overlooked. The third syndrome linked to myxomas are called constitutional syndromes, nonspecific symptoms caused by the myxoma (Figure 1).

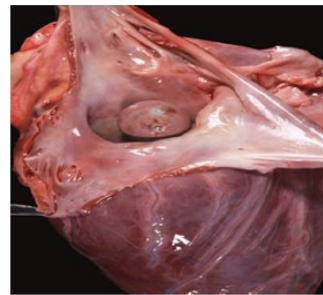


Figure 1: Intraoperative view of the myxoma which rise from RVOT and goes through the tricuspid valve into the right atrium

There is no known causative agent for myxoma. The main symptoms, if any, produced by myxoma are generic and not specific. These include fever, weight loss, anemia, elevated white blood cell (WBC) count, decreased platelet count and Raynaud's phenomenon. Myxomas are usually seen in adults. They are rarely seen in children, accounting for only 9-15% of all cardiac tumors from birth to adolescence. They are often found attached to the atrial septum and mitral valve apparatus in the left atrium (>85%). Three times less is incidence of the right ventricular myxoma. Myxomas can appear sporadically or as part of the syndrome myxoma or Carney syndrome, which includes endocrine neoplasms, tumors in other organs, and skin with spotty hyperpigmentation. This type of familial cardiac myxoma [5] accounts for less than 10% of the myxomas appearing in the heart. They have an autosomal dominant transmission and most commonly appear in females. Patients with syndrome myxoma tend to be younger than those with sporadic myxomas. Myxomas may embolize; this may be their first clinical presentation. Peripheral embolization is reported to occur in as many as 70% of patients with myxomas and may even occur in utero. Diagnosis is made following a suspicion that a myxoma might be present, and can usually be confirmed by echocardiogram. Surgery is used to remove the tumor. Myxomas can re-grow if they are not completely removed. The survival rate for this operation is excellent. Successful removal of the tumor rids the patient of this disease. Emboli from a myxoma may survive in other areas of the body. However, there is no evidence that myxoma is truly metastatic (able to transfer disease from one area to another), causing tumors in other areas of the body.

Case Report

A 13-year-old boy was presented in our hospital with a 40-days history of fever, sweating, dyspnea during physical activity and weight loss. A physical examination revealed a left side basal pneumonia, diastolic murmur on the pulmonary valve, joint pain and giant A-wave on jugular venous pulse. Echocardiography (transthoracic and 3D transesophageal-TOE) (Figure.2)

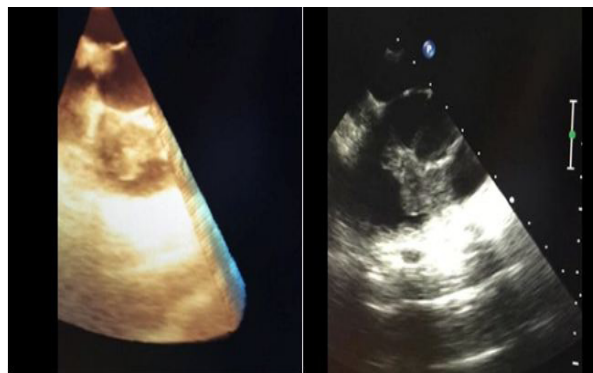


Figure 2: Transesophageal image of the Right outflow tract fulfilled with myxoma.

Revealed a huge mobile heterogeneous mass (2,5x3,5cm), suggestive of myxoma, in the right ventricle (RV) with origin from a fibrous part of intraventricular septum, protruding across the tricuspid valve into the right atrium (RA) (1x1,5cm), causing partial RV inflow obstruction with a sessile mass around the tumor mass, suggestive of thrombus. Cardiovascular CT scan confirmed the diagnosis of myxoma. No familiar anamnesis and no other signs for Carney complex were notified in our patient (Figure.3).

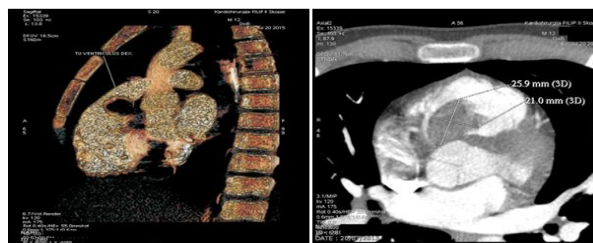


Figure 3: CT image of the myxoma into the RVOT.

Following sternotomy, routine cardiopulmonary bypass (CPB) was established through aortic and bicaval cannulation. The heart was arrested using cold blood cardioplegia. The right atrium was opened and a 2,5x3,5 cm mass was identified under the septal leaflet of the tricuspid valve, protruding into the PA. The base of the tumor mass was attached to the pars membrane of the interventricular septum (IVS). Septal leaflet was sized and tumor mass was resected with a small part of

membranous IVS. The hole on the IVS was closed with a direct suture. Right ventricle was visually inspected through tricuspid valve and no other masses were found. Postoperative TOE did not show the presence of any residual mass. After cardiopulmonary bypass weaning patient was in a AV block third degree, so epicardial pace maker was active DVI/90/min. After 10 days sinus rhythm was recovered completely. 21st day after surgery patient developed an AV block III rd degree, there why permanent pace maker Medtronic adapter was implanted. Histopathology confirmed the diagnosis of myxoma (Figure 4).

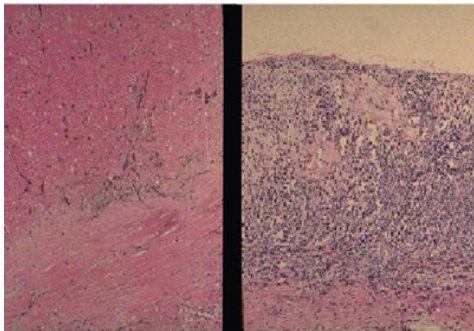


Figure 4: Pathohistology results, abundant mucopolysaccharide matrix and vascular channels with varying numbers of distinctive stellate or plump “myxoma” cells (left side). Tumor-associated thrombus, degeneration, hemorrhage, calcification, inflammatory infiltrate (basophils) or superinfection are showed on the right side

Patient is with one year survival period, with normal quality of life. Control echocardiography was in normal ranges, right ventricle with normal morphology.

Discussion

The diagnosis of myxomas is challenging and before the echocardiography diagnostic they were discovered only at autopsy. However, the development of TTE and TOE has facilitated the diagnosis of these tumors. Especially 3D TOE permits the location, origin, size, shape, attachment, mobility, valve compromise and hemodynamic consequences to be determined the presence of this tumor immediately under septal tricuspid leaflet with an origin from pars membrane of the IVS has been infrequently reported in the literature. In the case of our patient tumor was clearly demonstrated by means of the 3D TOE and therefore it was successfully excised during surgery. There have been reports of patients with myxomas in the LA and RV but diagnosed with a time interval. False-negative echocardiograms have been previously reported and it is more frequent

with right sided myxomas.

However, surgeon removed small membranous part of the IVS which was the base of the tumor attachment performing surgical reconstruction of that part. Transient AV block third degree and temporary pace-maker stimulation had been override after ten days in our case. Nevertheless, in spite of these wide resections myxomas can recur in different areas in the a trial cavity w10x. Some other groups support resection of the tumor alone with low incidence of recurrence after this approach w9x. Reported recurrence rates vary ranging from 1% to 14% for sporadic cases and up to 40% for familial cases. It is more common in young males, patients with multifocal masses and familial variant.

In our case the position of myxoma was so specific that it's extirpation was connected with high degree possibility for damage of tricuspid valve or appearance of AV block. Immediately after surgery using a TOE we realize that tricuspid valve stayed completely competitive, but AV block was transformed from temporary in permanent. The pathohistology analysis showed that the mixomatous tissue was incorporated in the normal septal tissue, where is the normal space of the Hiss bundle and its branches. Follow-up with TTE varies depending whether it is a familial or sporadic case. Some authors recommend a biannual follow-up, others annual with some groups suggesting up to 10 years or a lifetime 3D TOE continues to be an invaluable diagnostic modality for cardiac masses. It offers accessibility and crucial information on mass morphology, position and mobility.

Cardiac CT scan is another important non-invasive diagnostic tool that provides detailed information on tissue characterization therefore helping in the differential diagnosis of these masses.

Given our patient had all the characteristics of a familial myxoma, such as young age, right chamber involvement and multiple sites, we have recommended follow-up with echocardiogram for the patient and all first degree relatives.

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