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Case Report

# Right atrial tumor suspected myxoma in an 11-year-old girl

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Primary tumors of the heart are rare in children. The incidence is between 0.0017 and 0.19 percents in unselected patients at autopsy.[1,2] Three quarters of the tumors are benign. Benign cardiac neoplasms may lead to significant morbidity and mortality by affecting blood flow and causing arrhythmias and emboli.[3,4]

Previous autopsy study found that the most common primary cardiac tumors were rhabdomyosarcoma (45%), fibroma (25%), myxomas (10%), intrapericardial teratoma (10%), and hemangioma (5%).[5] Although myxomas are the most common tumor in adult, comprising from 60 % to 75% of primary cardiac tumors, they are rare in children. Myxomas may occur at all ages but are particularly frequent between the third and sixth decades of life. They usually occur sporadically.[6]

Myxomas are generally polypoid, often pedunculated, rarely sessile, and round or oval, with smooth or soft lobulated surface. The tumor size ranges from 1 to 15 cm in diameter; mostly are 5 to 6 cm.[7] Most myxomas originate from left atrium. Right-sided myxomas are extremely rare and can present with nonspecific signs and symptoms.[8] Despite the lack of symptoms and signs they should have a surgical resection. Surgical removal of the tumors should be performed as soon as possible because the long term prognosis is excellent and recurrences are rare.[7,8] This paper reports a case of right atrial myxoma in an 11-year-old girl.

## The case

An 11 year old girl was brought to Emergency Department, Sanglah Hospital, Denpasar on January 24th 2007 with complained of syncope which appeared suddenly while walking or standing. She woke up after about two minutes, spontaneously without any sequele. She felt much better when she was lying down. No complain of difficulty in breathing, or bluish color of lip, mouth, fingers or toes. There was no history of palpitation, cyanosis or dizziness. She did not complain of fever, cough or runny nose either. Her activity and appetite were normal, defecation and urination was also normal. This illness began when she was in the fourth grade of elementary school. She had never get any medication at all. A few hours before hospitalization, she had difficulty to breath insidiously. She was brought to a private hospital and after she got Oxygen, the symptom disappeared. She was brought to Sanglah hospital International Wing for further examination. Echocardiography was done by a cardiologist and she was suspected to have right atrium

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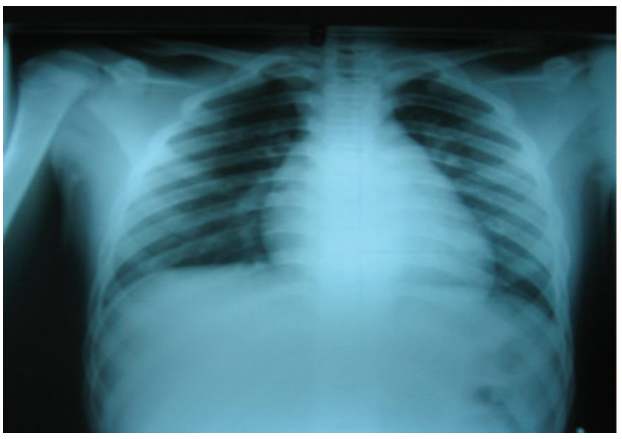

**Figure 1.** Enlargement of right atrium and right ventricle, also mild increased pulmonary vascularity was shown on chest X-ray.

myxomas. She suddenly had a syncope again and than brought to the emergency room on the same day.

On physical examination, the patient was alert, no cyanosis, regular pulse rate, 88 per minute, regular respiratory rate was 28 per minute. Temperature was 37 $^0$C and blood pressure was 110/70 mmHg. Her body weight and body height were 39 kg and 143 cm, respectively.

No abnormalities were found in her eyes ,pharynx and tonsils. There were no enlargements of lymph nodes.

Chest inspection showed no precordial bulging or ictus cordis. Ictus cordis was palpable and accentuated with impulse on the 3-4$^{th}$ intercostals space in left mid clavicular line. There was no thrill and RV heave. On auscultation, the first and second heart sounds were normal, regular with diastolic murmur in tricuspid valve area on the fourth intercostal space in left parasternal line. Lung examination showed symmetric chest movement. Vesicular respiratory sound was heard without wheezing or rales. The abdomen was normal. There was no sign of ascites.

The results of blood investigation (on January 24$^{th}$ 2007) showed; WBC count was 7.41 k/uL; haemoglobin was 12.3 g/dL; hematocrite was 36.3%; platelets count was 309 k/uL. Liver function test (on January 26$^{th}$ 2007) showed total bilirubin was 0.7 mg/dL, direct bilirubin was 0.00 mg/dL, AST was 22 u/L, ALT was 10 u/L and alkali phosphates was 184 u/L, albumin was 3.6 g/dL, globulin was 3.0 g/dL. BUN was 7 mg/dL, creatinine serum was 0.6 mg/dL. Bleeding time was 2'30", Clotthing time was 8'30". Protrombine time was 12.9", Plasma trombine time was 42.4".

On chest X-ray, there was no sign of cardiomegaly or double contour, but there was enlargement of right atrium and right ventricle with mild increased pulmonary vascularity (**Figure 1**). Electrocardiography (ECG) examination revealed normal sinus, P mitral (0.12 second) and P Pulmonal were negative, PR interval was normal limit (0.12 second), right ventricle hypertrophy (RVH) and left ventricle hypertrophy (LVH) was negative (**Figure 2**). Echocardiography examination revealed tumor in right atrium chamber, size 1 x 0.5 cm, mobile, pedunculated and the stalk incertion was on the atrium wall. According to the feature the echocardiography the patient was suspected to have of myxoma in right atrium (RA), which some time expulsion to right ventricle (RV), (**Figure 3**).

She was consulted to Thoracic Surgery Department. The surgeon advised to undergo excision

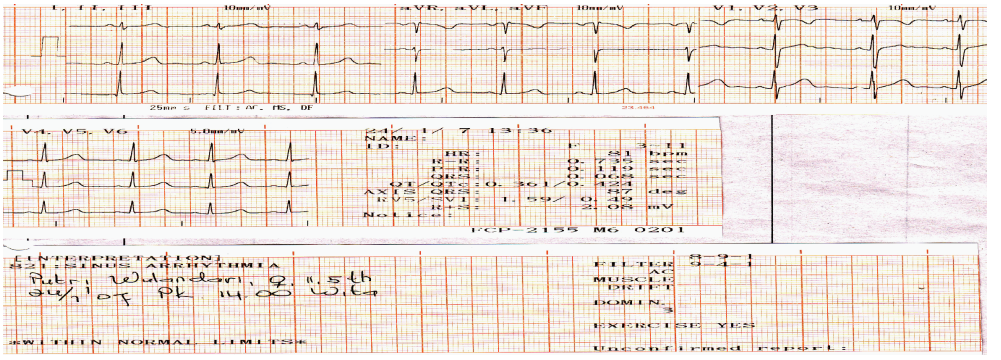

**Figure 2.** Electrocardiography revealed normal sinus, P mitral and P Pulmonal was negative , PR interval normal limit , and no shown RVH and LVH.

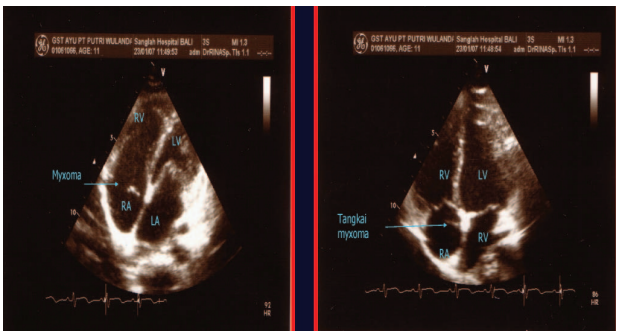


**Figure 3.** Echocardiography examination revealed Myxoma in right atrium (RA), which some time expulsion to Right ventricle (RV).

as soon as possible. Her father wanted the surgery to be done in Harapan Kita Hospital, Jakarta. the patient then referred to Harapan Kita Hospital ,Jakarta.

## Discussion

Cardiac tumors in pediatric age group are extremely rare. A primary cardiac tumor was diagnosed in 0.001% to 0.003 % of admissions at large children's referral centers. Autopsy studies found an incidence of 0.027% to 0.008%.[9] Myxomas are the most common type of cardiac tumors in adult, accounting for about 30% of all primary cardiac tumors, but they are very rare in infants and children.[5,9,10] There are no data about the incidence of cardiac myxoma in Indonesia. Our case was the first one in our hospital.

Myxomas are neoplasms of endocardial origin. The tumors usually projects from the endocardium into the cardiac chamber. The tumor cells are considered to be multipotential mesenchymal cells that persist as embryonal residues during septation of the heart and differentiate into endothelial cells, smooth muscle cells, angioblasts, fibroblasts, cartilage cells, and myoblasts. The rate of growth of myxomas is unknown, but they generally grow rather fast. The prevalence of myxomas in the atrial septum is therefore understandable. Cardiac myxomas usually develop in the atria. About 75 percents originate in the left atrium, and 15 to 20 percents in the right atrium.[5,7,9]

Most myxomas arise from interatrial septum at the border of the fossa ovalis, but they can also originate, in descending order of frequency, from the posterior atrial wall, the anterior atrial wall, and the atrial appendage. Myxomas on the heart valve are rare.[7-10] In our case the tumors were located in the right atrium, and echocardiography confirmed the diagnosed.

The clinical features of myxomas rarely depend on their location, size and mobility. Most patients present with one or more the triad of embolism, intracardiac obstruction, and constitutional symptoms.[5,7,11,12] Occasionally they are symptomless, particularly these with small tumors. Embolism occurs in 30% to 40 % of patients with myxomas. Since most myxomas are located in the left atrium, systemic embolism is particularly frequent. In cases of right atrial myxomas, clinical evidence of embolic events is uncommon. Peripheral emboli occur in more than 70% pediatrics patients with myxomas. Emboli are related to fragmentation of tumor substance or embolization of thrombi adherent to the tumor external surface. Left sided tumors are associated with systemic and right sided ones with pulmonary arterial emboli.[7,8,10] About 80% of pediatric patients present with symptoms of valvular obstruction. Symptoms and physical finding are often positional. When atrial myxomas obstruct the atrioventriculer valves, the patient may experience dyspneu, dizziness, or syncope when sitting or standing, with reducing of symptoms on lying down. Constitutional disturbances, such as fatigue, fever, erythematous rash, athralgia, myalgia and weight loss can be found. Recent finding suggest that the production and release of the cytokine interleukin 6 by the tumor itself may be responsible for the inflammatory and autoimmune manifestations.[3,7,10,12] In this case, the patient had syncope suddenly while walking or standing for 2 minutes and then woke up spontaneously. The symptoms reduced if she was lying down. This mechanism could be caused by the tumor obstructing the atrioventricular valves.

On physical examination, systolic or diastolic murmur may be heard in more than half the patients with myxomas, depends on the location, size, and mobility of the tumor and on the body position. Diastolic murmurs are due to obstructed filling of the left or right ventricle; systolic murmurs occur if the myxoma interferes with the closure of the atrioventricular valves or narrows the outflow tract.[7,9,10] The

auscultatory findings are characteristically variable and depend on body position in cases of mobile tumors. In about one third of the patients, protodiastolic murmurs can be heard 80 to 150 msec after the second heart sound ("tumor plop"). Moreover, pericardial friction rubs may sometimes be present in patients with right atrial tumors.[7,12] In our case auscultation showed first and second sound normal, regular with diastolic murmur in tricuspid valve area on the fourth intercostal space in left parasternal line.

Laboratory findings showed anemia, thrombocytopenia, elevated sedimentation rate and gamma globulin. Anemia generally normochromic or hypochromic, but the hemolytic anemia may also be found because of the mechanical destruction of erythrocytes by the tumor. Less common findings are leukocytosis, thrombositopenia.[7,10,12] Our case, showed normal levels of WBC, hemoglobin and thrombosit. Globulin was also normal.

The chest radiography may be normal or may demonstrate cardiomegaly with pulmonary edema. Right sided myxomas show right atrial and right ventricle enlargement. Classification that makes the tumor visible on routine examination is unusual and is considered to be more frequent in cases with right atrial tumors.[9,10,12]

In this case, chest radiography showed enlargement of right atrium and right ventricle with mild increased pulmonary vascularity.

Electrocardiography (ECG) findings are nonspecific. They may reflect the hemodynamic alterations caused by the tumor, which result in atrial overload or ventricular hypertrophy. The cardiac rhythm usually shows normal sinus; in contrast to the findings in mitral valve disease, atrial fibrillation is uncommon. Bundle branch block, repolarization abnormalities, commonly seen in cases with intramural rhadomyomas and fibromas, are rarely seen in those with myxomas.[5,7,12]

In our case, ECG showed normal sinus, no P mitral (0.12 second) and P Pulmonal ; PR interval was within normal limits (0.12 second), RVH and LVH was negative.

The mainstay in the diagnosis of cardiac myxomas is two dimensional Doppler echocardiography. This method can generally be used to determine the location, size, shape, attachment, and mobility of a myxoma and is superior to one-dimensional echocardiography.[5,7] On echocardiography, the characteristic narrow stalk is the most important distinguished feature of a myxoma, followed by tumor mobility and distensibility. When these features are seen, myxoma can be diagnosed with high degree of confidence, especially if the tumor is also attached to the interatrial septum. Myxomas demonstrate variable internal echocardiography features.[5,8,13] In our case, echocardiography revealed tumor in right atrium chamber, with size of 1 x 0.5 cm, mobil, pedunculated and the insertion of the stalk is on the atrium wall but sometimes expulsed to the tricuspid valve. According to that feature the confirmed diagnosis was myxoma in right atrium, which sometimes expulsed to right ventricle.

The treatment of choice of atrial myxoma is early surgical resection with cardiopulmonary bypass. After diagnosis has been established, the surgery should be performed promptly, because of possibility of embolic complications or sudden death. This treatment is curative; the earlier the diagnosis is confirmed and the earlier surgery is performed and the better the prognosis of the patient.[7,8,14] Most myxomas are pedunculated and must be removed as one mass, as intraoperative fragmentation may lead to embolization or recurrence. Operative mortality is 0% to 3% in multiple series. Recurrence rate are 1-3% for sporadic myxomas and are thought to be a result of incomplete resection.[14-16]

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