Primary Fibro Sarcoma of the Heart

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Case report

ABSTRACT

Primary malignant heart tumors represent rare entities where fibro sarcoma represents about 3% of all. Introducing the patient: A 15 years old patient with cardiac insufficiency (heart failure) symptoms, such as weakness, cyanosis, palpitations and breathing difficulties; enlargement of upper mediastinum and pleural effusion. Through echocardiography a

pericardial effusion and intracavitary thrombus in atrium was diagnosed. With computed tomography is diagnosed a tumoral mass in right atrium which is also spread in the right ventricle of the heart. Tumor is completely removed; pat histology result showed primary fibro sarcoma of the heart. At that time no metastasis was found. Conclusion. Primary malignant heart tumors may manifest like cardiac insufficiency or like systemic dis-

eases. Fibrosarcomas are rare and have bad prognosis. On average patients can live around six months after initial symptoms appeared and diagnosis of the tumor was done. In the case of cardiac insufficiency with differential diagnosis we should also think of heart tumors, which could certainly be proved for or eliminated by echocardiography.

Key words: Heart, tumors, fibro sarcoma, insufficiency, diagnosis, echocardiography.

1. INTRODUCTION

Findings about heart tumors were initially provided by Bonet in 18th century (1, 2). In 1936 first successful resection of cardiac tumor was performed (3). Depending on their origin heart tumors are classified in two main groups. First group consists of tumors that originate from heart tissues and thus are known as primary tumors whereas the second groups consist of secondary tumors which represent different malignant tumor metastasis that originate from others tissues and body organs. Primary heart tumors are very rare entity. According to autopsy results and surgical interventions their incidence is around 0.001 – 0.28 (3) while heart metastasis are 10-50 times more frequent (1, 3, 4, 6). About 75% of primary heart tumors are benign whereas others are malign. (2, 3). Sarcomas make about 75% of primary malign heart tumors (6) of which the most common one is angiosarcoma.

Fibrosarcomas are very rare tumors and make only about 3% of primary malignant heart tumors (4). In the following section we present a patient diagnosed with primary fibro sarcoma of the heart.

2. INTRODUCING THE CASE:

A 15 years old male patient, on the last week of September 2012, is hospitalized at the Pediatrics Clinic at the University Clinical Centre in Kosovo (UCCK) on the intensive care unit. He was showing some non-specific symptoms and signs such as weakness, fever; headache, nausea for about three weeks and gradually his situation worsen.

At the moment of check in the patient was cyanotic and had face edema particularly in the per orbital region. With heart and lung auscultation it was found that heart beat was weakened but rhythmical while breathing noise was weakened. He-

Figure 2. CT of thorax with i.v. contrast Axial plane

moglobin saturation was 67%, and arterial pressure was 80x60mmHg.

Lab data: WBC - 12.6, RBC - 5.08, Hgb - 14.3, Hct - 41.5, PLT - 188, MCV - 81.8, Gli - 4.8, ALT - 11, AST - 16, Chol - 2.5, CK - 40. Direct Bil. - 8.9, Total Bil. - 28.5, LDH - 507, Urea - 4.7, Urine: Prot. ++, Le. - 17. In the thorax radiography the PA view found pleural effusion of the left side and enlargement of upper mediastinum mainly on the right side of v. cava superior and v. azygos region. The ECG registered a sinus rhythm with a rate about 100/min, decreased voltage and AV block of grade I. With the transthoracic echocardiogram was

Figure 1. CT of thorax without i.v. contrast Axial plane

Figure 3. CT of thorax with i.v. contrast Coronal plane

Figure 4. CT of thorax with contrast, Coronal plane

Figure 5. CT of thorax with i.v. contrast. Coronal plane

found pericardial effusion, stasis on systemic veins and decrease of circulation on pulmonary artery. It is also found a hyperechogenic mass that nearly covers the right atrium and spreads towards the right ventricle, which was thought it represented a massive intracavitary thrombus. The patient was referred to the Radiology Clinic; a computed tomography of thorax with and without i.v. contrast was conducted. In this case a hyper-density mass was observed and the

majority of which after applying the contrast it showed a tumoral mass. This mass filled up almost all of the right atrium, which was widened and pressured the left atrium (Figure 1 and 2).

We also observe a spread on the right ventricle through ostium atrio-ventricular and an extension up on the superior caval vein, brachiocephalica dex. vein and jugularis dex. vein, and thus causing their dilatation (Figure 3 and 4).

On the axial plane the maximum diameter of the intra-arterial septum of this mass was 7x8.5 cm. In the pleural space there is a massive bilateral effusion. A greater enlargement of veins was also discovered which anastomose two kava veins: vena mammaria interna and vena azygos (Figure 5 and 6). No metastases were found. Free fluid is observed in the abdominal segment included in this

Figure 6. CT of thorax with i.v. contrast. Sagittal plane

Figure 7. CT of thorax with i.v. contrast (post surgery)

examination.

The patient was referred to a specialized Cardio-surgical centre in Tirana where he underwent a surgical intervention. The histopathological analysis proved that this was a case with fibrosarcoma tumor. A month

Figure 8. CT of thorax with i.v. contrast (post surgery)

Figure 9. CT of thorax, abdomen and pelvic with i.v. contrast – Coronal plane

and a half after the surgery another CT of thorax, abdomen and pelvic was conducted with an i.v. contrast (Figure 7, 8 and 9).

A pleural effusion was found on the left side. Pericardial effusion was also found. No signs of secondary mass were identified.

3. DISCUSSION

Heart tumors represent a big challenge when diagnosing them. Their clinical symptoms are nonspecific, and depend more on the size and location of tumor than on its histologic type. (6) They are caused by four main mechanisms: intracavitary enlargement of tumor can cause obstruction of blood circulation or malfunction of heart valves; local invasion of tumor can cause arrhythmia and pericardial effusion; a part of tumor can embolise and cause cerebrovascular and peripheral vascular accidents. Also, when tumor releases cytokines (IL-6) it can cause systemic or constitutional symptoms of weight loss, fever, arthralgia, myalgia, hemolytic anemia, thrombocytopenia, leukocytosis, elevation of CRP, faster SE (2, 6).

Primary heart sarcomas are found with people of all group ages, but more often in the 3-5 decade of their life with no observable gender differences (4). Our patient is 15 years old male. Primary heart sarcomas are usually found on the right atrium, and frequently are angiosarcomas (6).

Sarcomas found on the right side of heart usually infiltrate more, grow in the shape of cauliflower, and metastasize earlier compared to sarcomas located on left side of heart (6, 7). In our case we are dealing with a massive tumor located on the right atrium that spreads on the right ventricle towards vena cava superior, but no metastasis where found at the moment of diagnosis. Also, sarcoma located on right side of the heart rarely causes congestive heart insufficiency or disorderly hemodynamic status compared to left side sarcomas, which are very symptomatic at the moment of presentation (6, 7). In our case we have symptoms of heavy right side heart insufficiency NYHA III.

Through thorax radiography we can get nonspecific data. In our case we represented a widening of upper mediastinum and pleural effusion.

Echocardiography plays an important role on diagnosing heart tumors. It enables us to see the tumor, measure it, observe its location, movement and place where it is fixed. A computed tomography plays an important role in diagnosing heart tumors and allows us to see other thorax structures. In our case, through echography, the described mass is reported as thrombus. Also, it is also reported a pericardial effusion which was not observed in CT.

Magnetic resonance is very important in diagnosing heart tumors, too. We were not able to apply it because of patient's difficult situation.

Surgical intervention and complete tumor resection is main treatment. In case of malignant tumors chemotherapy and radiotherapy are applied too.

Primary malignant heart tumors which often attack young people have very bad prognosis: without a surgical intervention, survival between 9 to 12 months is 10% (6).

4. CONCLUSIONS

Heart tumors are rare; fibro sarcoma makes about 3% of all malignant heart tumors. Their diagnosis is very difficult not only because of their rareness but also because of their nonspecific clinical symptoms. In cases of acute heart insufficiency, in a differential diagnosis, we should suspect a heart tumor whereby every patient should be checked with echography. Heart sarcomas have bad prognosis; about a year after the diagnosis was made. Surgical excision, even if partial, is indicated for the purpose of decreasing symptoms of obstruction. In some cases of non-surgical sarcomas good results are achieved by heart transplanting.

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