CASE REPORT

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Benign tumors of the heart: myxoma of the right atrium – A case report

Benigni tumori srca: miksom desne pretkomore

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Abstract

Introduction. Myxoma is the most common primary benign heart tumor. The most frequent location is the left atrium, the chamber of the heart that receives oxygen-rich blood from the lungs. Myxomas usually develop in women, typically between the ages of 40 and 60. Symptoms may occur at any time, but most often they are asymptomatic or oligosymptomatic for a long period of time. Symptoms usually go along with body position, and are related to compression of the heart cavities, embolization and the appearance of general symptoms. The diagnosis of benign tumors of the heart is based on anamnesis, clinical features and findings of the tumor masses by use of non-invasive and invasive imaging methods. Extensive surgical resection of the myxoma is curative with minimal mortality. Long term clinical and echocardiographic follow-up is mandatory. **Case report.** We reported a case of a 62-year-old male, presented with 15 days of intermittent shortness of breath, dizziness and feeling of heart palpitations and subsequently diagnosed with right atrial myxoma based on transthoracic echocardiography . The patient was emergently operated in our hospital. Long-term follow-up did not reveal recurrence. **Conclusion.** Our case was an atypical localisation of right atrial myxoma. Whether the intracardiac mass is benign or malignant, early surgery is obligatory in order to prevent complications.

Key words:
myxoma; heart neoplasms; diagnosis; echocardiography; echocardiography, transesophageal; histological techniques; cardiac surgical procedures; treatment outcome.

Apstrakt

Uvod. Miksomi su najčešći primarni benigni tumori srca. Najčešće su lokalizovani u levoj pretkomori koja prima krv bogatu kiseonikom iz pluća. U najvećem broju slučajeva sreću se kod osoba ženskog pola starosti od 40 do 60 godina. Simptomi se mogu pojaviti u svakom trenutku, ali su ovi tumori vrlo često asimptomatski ili oligosimptomatski tokom dužeg vremenskog perioda. Klinička slika zavisi od položaja tela i posledica je pritiska koji tumor vrši na srčane šupljine, embolizacije ili prisustva generalnih simptoma. Dijagnoza se postavlja na osnovu anamneze, kliničke slike i vizualizacijom tumora pomoću neinvazivnih i invazivnih dijagnostičkih procedura. Ekstenzivno hiruško uklanjanje je terapija izbora uz minimalnu stopu smrtnih ishoda. Dugotrajno kliničko praćenje je neophodno. **Prikaz bolesnika.** Prikazan je mušakrac star 62 godine, koji je 15 dana ranije imao povremeni osećaj kratkoće daha, vrtoglavice i lupanja srca. Dijagnoza miksoma desne pretkomore postavljena mu je na osnovu ehokardiografskog pregleda i tumor je hitno operativno uklonjen u našoj ustanovi. Tokom višegodišnjeg praćenja bolesnika nije uočen recidiv. **Zaključak.** Ovo je slučaj atipične lokalizacije miksoma u desnoj pretkomori. Bez obzira na to da li je intrakardijalna masa benignog ili malignog porekla, u cilju sprečavnja komplikacija neophodno je rano hiruško uklanjanje.

Ključne reči:
miksom; srce, naoplazme; dijagnoza; ehokardiografija; ehokardiografija, transezofagusna; histološke tehnike; hirurgija, kardijalna, procedure; lečenje, ishod.

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Introduction

In general, though cardiac tumours can be malignant or benign, later group is more common. Literature data report that although most cardiac tumours are benign, they could have malignant potential due to secondary impaired cardiac function (congestive heart failure, inflow/outflow tract obstruction, conduction system involvement) or peripheral embolization [1].

Myxomas are the most frequent type of benign cardiac tumors in all age groups. Appart from them, other less common neoplastic tissues like rhabdomyomas or fibromas, are mostly seen in children. In all cases, a definitive diagnosis is important because of its unpredictive nature. Namely, some cardiac tumors can be malignant, or they can present metastasis from a primary tumor [2].

As the most frequent, myxomas arise usually from the endocardium. Due to its slow growth, in about 20% cases, they are asymptomatic or oligosymptomatic for long periods of time. In 90% of cases, it appears in the period of 30 to 60 years of age, prevailingly in the sixth decade of life, and 75% of sporadic myxomas occur in females [3].

In everyday clinical practice, the early detection is of a great importance. This fact is important because clinical presentation is non-specific and depends on localization, growth rate and size of tumor. The most common clinical presentations are the symptoms and signs of heart obstruction and embolization. Also, literature data indicates that benign cardiac tumors increase the risk of an ischaemic strokes [4, 5].

Macroscopically, myxomas are clearly limited, gelatinous consistence, reddish-blue or yellowish coloration. They grow, most frequently, in the atria, especially in the left atrium. Although the most cases are sporadic, approximately 10% are familiar and are transmitted in an autosomal dominant mode [4].

Case report

A 62-year-old man, admitted to the Coronary Care Intensive Unit, was presented with intermittent shortness of breath, dizziness, heart palpitations and progressive weight loss. The above symptoms were present for 15 days before admission. Personal history was positive for hypertension and smoking. Family history was positive for cardiovascular diseases. Upon clinical examination, the patient's heart rate was regular at 100 bpm, blood pressure was 150/100 mmHg, and body temperature was normal. The lips and finger nails were neither cyanotic nor clubbed. No thrills were detected in the precordial region. Lung sounds were normal, with soft systolic heart murmur (2/6) at Erb's point. Routine laboratory data showed high erythrocyte sedimentation rate [100 mm/h (reference range, < 20 mm/h)] and high D-dimer level (1,616 ng/mL). Electrocardiography (ECG) and chest X-ray were within normal limits (Figure 1). Transthoracic (Figure 2) and transesophageal echocardiography (Figure 3) revealed an occupying mass in his right atrium of approximately 5.3 x 4.6 cm and comfirmed moderate tricuspid obstruction.

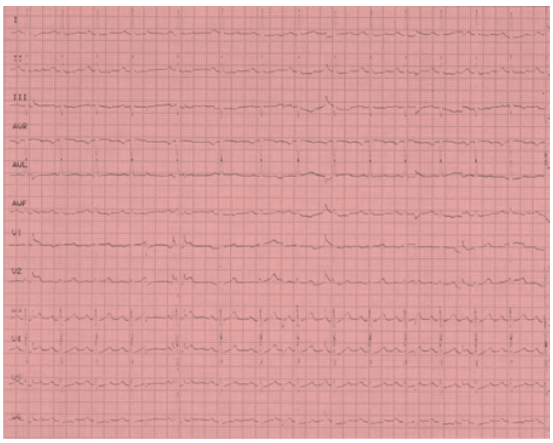

Fig. 1 – The patient's electrocardiogram (ECG) findings on admission.

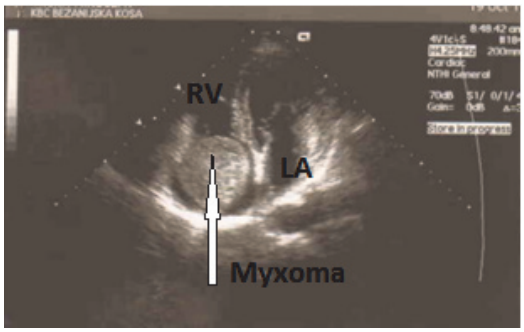

Fig. 2 – Echocardiogram shows the right atrial myxoma (highlighted). (LA – left atrium, RV – right ventricle).

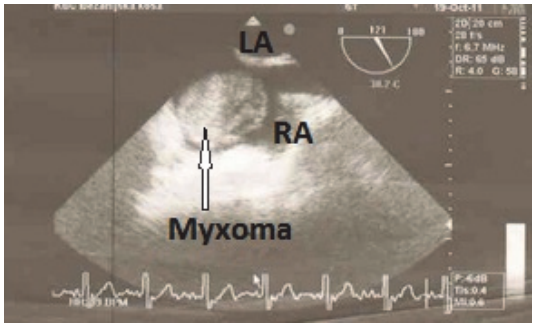

Fig. 3 – Transesophageal echocardiography revealed a large echo dense mass in the right atrium (LA = left atrium, RA = right atrium).

During his stay in the Coronary Intensive Care Unit he was hemodynamically stable with occasional episodes of paroxysmal supraventricular tachycardia.

He was scheduled for emergent surgical removal. Following preoperative preparation, the patient was taken for cardiac surgery. The heart was approached through median sternotomy. During surgery, the right atrium was opened and occupying mass was found in the posterior wall of right atrium and completely removed. Macroscopical examination of the mass revealed a smooth mass of 50 x 45 x 30 mm with a stalk arising from the posterior wall of the right atrium. The diagnosis was confirmed histologically.

Seven days after the operation, the patient was discharged from the hospital. Months and years later, further reexa-

minations, revealed no residual tumour. Three years after the operation, due to the permanently present atrial flutter type I, radiofrequency catheter ablation was done. The patient was returned to a sinus rhythm. On the last check-up, 5 years after the cardiac surgery, interatrial septum aneurysm was maintained (Figure 4).

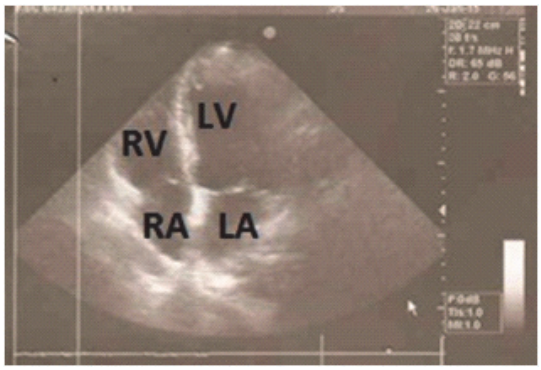

Fig. 4 – Four-chamber view of patient's transthoracic-echocardiographic study revealed interatrial septum aneurysm in a follow-up after 5 years. (LV – left ventricle; RV – right ventricle; RA – right atrium; LA – left atrium).

The study was approved by an Institutional Review Board and the patient gave his informed consent.

Discussion

Although most commonly cardiac myxomas arise from the interatrial septum, in our case the localization was rare – from the posterior wall of the right atrium [6].

Generally, myxoma is insidious primary cardiac tumor, commonly presented with nonspecific symptoms, such as fatigue, fever, myalgia, erythematous rash, arthralgia, weight loss, or laboratory abnormalities. These facts makes it difficult to diagnose and makes it potentially fatal if untreated [7].

Additionally, although they are regarded as mild proliferative lesions, with low metastatic potential and without modulation of tumoral suppressing genes or oncogenes, its malignancy may be determined by its specific behavior [8].

The literature data point on the fact that the presence of clinical symptoms depends on the tumor size. Namely, left atrial myxomas become symptomatic when they reach about 70 years, while right atrial myxomas may grow up twice this size before becoming symptomatic [9, 10].

Clinical presentation depends mostly on size, location and mobility of the tumor and thus may mimic arterial embolism or symptoms of cardiac, infectious, malignant and immune diseases, making its diagnosis more challenging [11].

Namely, the most common cause of complication is systemic embolization. It appears in 25%–50% of cases and can occur to any end organ leading to ischaemia and possible infarction [11]. The embolism end point depends on the location of myxoma and the presence of intracardial shunt.

Also, study conducted by Ha et al. [12] pointed on the association between polypoid cardiac myxomas and the occurrence of systemic embolism. Namely, the incidence of systemic embolization was significantly higher in patients with polypoid compared to round shaped myxomas. Because of previously mentioned fact, cardiac tumors should be in the differential diagnosis in every case of embolization.

Additionally, in some cases, benign tumors can manifest with arrhythmia, inflow/outflow obstructon, or even as a heart failure. Also, polypoid tumors more frequently prolapse through the mitral or tricuspid valve into the ventricle, causing the destruction of the annulus of valve leaflets [12].

Clinical significance also lies in the fact that they could be neovascularized by a branch of coronary artery, which can lead to bleeding in the myxoma with the progressive mass size increasement and consecutive blood flow obstruction [13].

Myxomas can produce numerous growth factors and cytokines, such as vascular endothelial growth factor that accelerates angiogenesis and results in tumor growth and increase the expression of proinflammatory cytokines [9, 14]. Thus, the period from the onset of the symptoms to the accurate diagnosis could be long and ranges from 1 to 126 months [15].

In some cases, cardiac myxomas can be associated with Carney complex, rare autosomal dominant syndrome, characterized with spooty skin pigmentation, endocrinopathy and endocrine or neuroendocrine tumors [16]. Although family members often have the same gene mutation, it is usually presented with distinct phenotypes as a result of various genetic and environmental factors. Recent studies identified mutations in the PRKAR1A gene that encodes the protein kinase A regulatory subunit 1-alpha (R1α) on chromosome 17q [16].

In terms of adequate diagnosis, two-dimensional echocardiography is in most cases sufficient. It provides an assessment of tumor location, size and mobility. On the other hand, transesophageal echocardiography is more sensitive, with better specificity and 100% sensitivity compared to transthoracic echocardiography. It also provides better resolution of heart chambers which makes it the imaging modality of choice [17]. Also, in patients older than 40 years of age it is very important to perform cardiac catheterization and angiography to exclude coexistent coronary artery disease or in order to evaluate neovascularization [9].

Immediately after the diagnosis is made, operative resection of the myxoma is the treatment of choice. After the surgical resection, diagnosis is comfirmed by histologic examination. Namely, myxomas are characterized by the presence of myxoid stroma fulfilled with the lipid cells [18]. Surgery is usually safe and curative, with an early postoperative mortality below 2% [19]. Complications are very rare and mostly presented as a postoperative atrial fibrillation, postsurgical neurologic complications or as it can be associated with unprofessional interatrial septum revision and consecutive bleeding which require exploration [19, 20].

Generally, the possibility of myxoma reccurence is rare, except in younger patients or in cases of Carney complex, where the possibility of the tumor recurrence is high. In these cases it is reasonable to do a biannual transthoracic echocardiography [12]. According to recent literature data, in all other cases, because of rare myxoma recurrence rate, continuous annual echocardiography is called into question [21].

Conclusion

We presented a case of a patient with an atypically located myxoma arising from the posterior wall of the right atrium.

Diagnosed by echocardiogram and after emergently surgical removal comfirmed by histological analysis.

Although rare, cardiac myxomas are the most common benign cardiac tumors. Its appearance is very important since it has nonspecific symptoms. Given the high embolic potential of myxomas, surgical removal is always the best treatment option. Prior the surgery, transthoracic echocardiography represents the single and the most important diagnostic method which gives the accurate anatomic information and identifies a precise surgical strategy.

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