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Case report

Right ventricular myxoma

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Abstract Right ventricular myxoma are primary rare benign cardiac tumors with each new case contributing to the new evidence in a management and treatment of the disease. We present an 80 year-old male patient with symptoms of fatigue on exertion and previous history of essential hypertension. Right ventricular myxoma was diagnosed on echocardiography and it was confirmed by futher diagnostic procedures. The most serious complication is arrhythmia and sudden cardiac death. A patient was referred for the urgent cardiosurgical intervention and successful extirpation of tumor was performed.

Kew words primary cardiac tumor, right ventricular myxoma, echocardiography

Background

Myxoma are round or oval-shaped, pedunculated, mobile, intra-cavity tumors[1]. They are rarely found in the ventricle, especially the right ventricle. The frequency of cardiac myxoma is 7 cases per 10,000 within the population and they account for 25-40% of all cardiac tumors in adults[1,3]. Primary cardiac neoplasms are relatively uncommon, with an overall incidence of 0.0017 to 0.19%[2]. Myxoma, benign primary tumor, account for 25-40% of all cardiac tumors in adults[2]. In regard to localization, 75-85% of myxomas originate within the left atrium, 15-20% in the right atrium[2,4], and only 2.5-4% within the ventricular chambers[2,4,5]. Right ventricular myxomas are particularly uncommon, with only 30 cases reported since 2010[6]. From a pathohistology perspective, myxoma consist stellate cells on a background of myxoid stroma and may be homogenous or present with central areas of hemorrhage, necrosis, calcification, and thrombosis[7].

Symptoms of right ventricular myxoma vary based on size and location within the cavity, but are often uncharacteristic which may lead to delayed diagnosis and unfavorable outcomes[7,8]. Clinical manifestations typically present with a triad of obstructive, embolic, and constitutional symptoms[4]. Patients with right ventricular outflow tract obstruction may present with symptoms of obstruction, or those similar to that in right sided congestive heart failure such as ascites, lower leg edema, and Superior Vena Cava Syndrome[4,7,8]. Patients may also exhibit dyspnea secondary to pulmonary embolization or constitutional symptoms such as fever, weight loss, and infection[4].

Echocardiography, computed tomography, and cardiac magnetic resonance imaging are the diagnostic modalities of choice, as well as preoperative imaging modalities crucial in surgical resection which is currently the treatment of choice[9]. Gold-standard diagnosis is mostly done post-resection with pathohistology analysis.

Case presentation

An 80 year old man with a history of essential arterial hypertension presented to the emergency department with complaints of fatigue upon exertion that became more pronounced in the previous 6 months. On examination, the patient was alert, well-oriented in all directions, afebrile, and eupneic. Significant physical examination findings included slight pretibial edema without varicosities and a discrete systolic murmur heard over the heart apex. All other parameters were normal. The patient underwent electrocardiography which showed sinus rhythm with a frequency of 70/min and in V1 and V2 slight negative T waves. Blood pressure was 140/80 mmHg. Laboratory results revealed sideropenic anemia and hypercholesterolemia. The transthoracic echocardiography showed an ovoid hypermobile solid tissue mass located at the right ventricular base, with a bipolar diameter of approximately 40mm, obstructing the right ventricular outflow tract (Figure 1, 2) It was determined that the mass was a right ventricular myxoma. During echocardiography, the severe tricuspid regurgitation and pulmonary hypertension was diagnosed with peak systolic pressure in the right ventricle of 88mmHg. The ejection fraction of the left ventricle was 60%. Cardiac surgery procedure and extirpation of myxoma was performed. Post-operative procedure and further follow-ups revealed no complications and patient stability. The right ventricle systolic pressure decreased to 45mmHg with now moderate tricuspid regurgitation.

Discussion

Cardiac tumors are rare and are mostly secondary while myxoma is the commonest primary cardiac tumor[10]. It is

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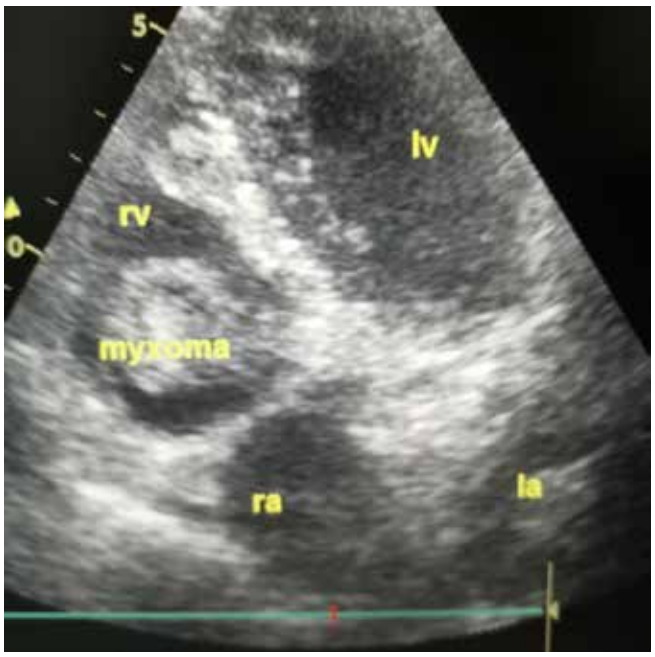

Figure 1. TTE 4-chamber view

observed more often in females than in males, with the mean age of presentation being 53 years[11]. Right ventricular myxoma is of unknown etiology, but genetics may play a role in disease development. Symptoms are varied and depend on multiple factors including location, size, mobility, and fragility[12]. Symptoms present with right ventricular outflow tachycardia obstruction such as shortness of breath and fatigue upon minimal exertion[13] as was observed in our case. These symptoms were secondary to pulmonary hypertension, a significant consequence of RV myxoma in our patient.. Although the tumor itself is benign, complications such as embolisation[15], pulmonary hypertension or sudden death may occur most significantly due to mechanical obstruction caused by the myxoma[15]. In 30-40% of all cases, embolism occurs[16] due to constant agitation of the tumor and may result in detaching of parts of the tumor or the whole tumor[17]. Specific characteristic of myxoma is its ability to mimic systemic autoimmune diseases due to secretion of interleukin 6 and 8 [18]. IL-6 is a pleiotropic cytokine that increases B cell differentiation and leads to increased synthesis of polyclonal immunoglobulins[18]. One of the rarest forms of cardiac malignancy, right ventricular myxoma, manifests with RVOT obstruction leading to syncope and it is essential that the diagnosis is made early, patient is sent to surgery and even though remission is rare, continuous follow up is needed[19]. In our case this 80 year old patient was not sent to echocardiography testing earlier in his lifetime and focus was on treatment of pulmonary hypertension. It is essential that clinicians recognize and identify right ventricular myxoma, define its cause and treatment through echocardiography as soon as possible. It also needs to be taken into consideration for differential diagnosis of B-cell lymphoma. B-cell lymphoma and right ventricular myxoma may give the same symptoms, such as right ventricular inflow obstruction and low cardiac output. Misdiagnosis can be made on echocardiography as well since both appear as a ventricular mass on the image. Treatment of the right ventricular myxoma needs

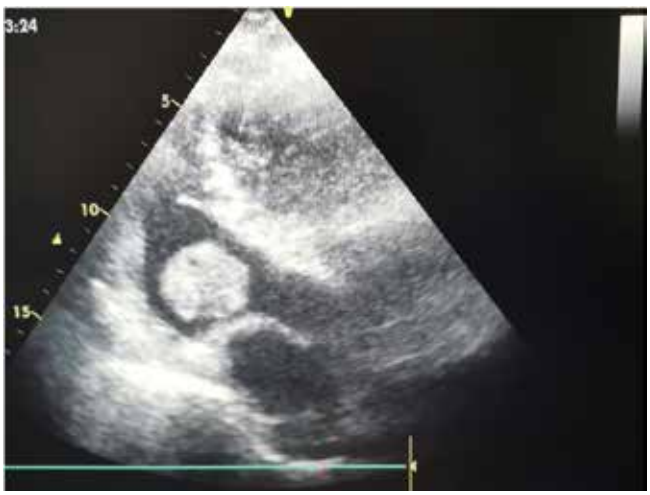

Figure 2. TTE

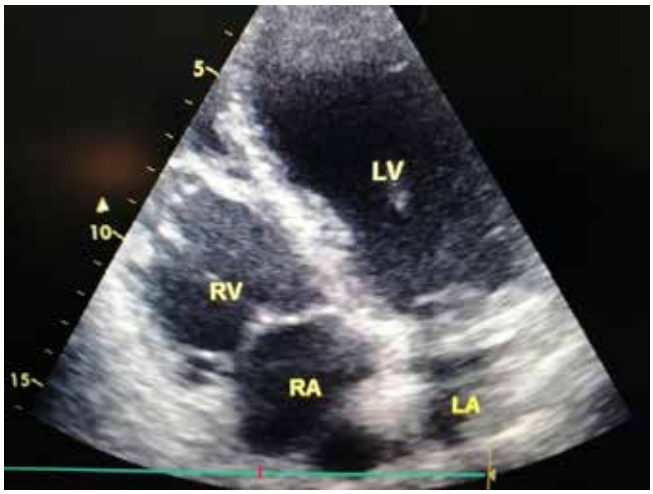

Figure 3. Post-operative TTE

to be urgent and tumor mass should be surgically removed. However, radiofrequency ablation may also be used for patients in poor clinical conditions where cardiac operation can be risky or it can be used as a supplementary technique for the treatment of obstructive cardiac tumors, when only partial resection is possible[20].

Conclusion

Cardiac myxomas filling the entire RV cavity, resulting in right ventricle inflow and outflow tract obstruction is very uncommon. Diagnosis can be achieved with echocardiography, and the patient should be referred for surgical intervention. These tumors should be excised on an urgent basis to avoid risk of embolization or even sudden death.

Patient Consent Form: Patient was informed about publication
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Sažetak

Miksom desnog ventrikula

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Miksomi desnog ventrikula su veoma rijetki primarni tumori srca i time svaki novi slučaj doprinosi novom dokazu u dijagnozi i liječenju ovog oboljenja.. U ovom radu prikazali smo 80 godina starog muškarca sa simptomima zamora u naporu te od ranije anamnestički prisutnoj esencijalnoj hipertenziji. Miksom desnog ventrikula dijagnosticiran je ehokardiografijom i potvrđen odgovarajućim dijagnostičkim procedurama. Najčesća ozbiljna komplikacija je aritmija srca i iznenadna srćana smrt. Pacijent je upućen na urgentnu kardiohiruršku operaciju i urađena je uspješna ekstirpacija tumora.

Ključne riječi: *primarni kardialni tumor, miksom desnog ventrikula, ehokardiografija*