

CALCIFIED ATRIAL MYXOMA AS A RARE CAUSE OF ATRIAL FIBRILLATION

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Primary cardiac neoplasms are very rare, and the most of those in adults are myxomas. We are reporting an interesting case of extensively calcified left atrial myxoma presenting by paroxysmal atrial fibrillation and angina pectoris like symptoms, mimicking coronary heart disease. The tumor was surgically removed and histological analysis confirmed pendicular myxoma with diffuse areas of calcifications, various in shape and size. Coronary heart disease was excluded by an invasive angiography. After the tumor removal atrial fibrillation disappeared, as well as other symptoms.

Key words: heart tumors, myxoma, calcifications, arrhythmia, atrial fibrillation

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INTRODUCTION

Primary cardiac neoplasms are very rare, three quarters of those are benign, and nearly half are myxoma (1,2). Cardiac myxoma usually occur sporadically, particularly frequent between the third and sixth decade of life, predominantly in females (1-4). Myxoma is usually pendicular and mobile (85%), arising from the interatrial septum typically on the left side (59-75%), but can also be located elsewhere in the heart: right atrium (15-28%), right ventricle (3-8%), left ventricle (0-4%), and can rarely be multifocal (5,6). Typical myxoma is 5-6 cm in size, oval with a lobular or smooth surface, in 16-34% of cases it can be irregular and villous with fragile gelatinous fronds that tend to fragment spontaneously, since they are usually composed of soft and friable tissue (5,6). Patchy calcifications are present in 10% to 20%, but massive calcification is uncommon (7,8). The clinical symptoms are often, over a longer period of time absent or non-specific, resulting in substantial growth prior manifesting symptoms (1,2,9). When myxoma become symptomatic, one or more typical manifestations are present: embolism (29%), intracardiac obstruction (67%), and/or constitutional symptoms (34%) which include: fatigue, fever, erythematous rash, arthralgia, myalgia,

and weight loss, causing significant morbidity and mortality, even sudden death (1,9). Other less frequent symptoms are: dyspnea, arrhythmia, syncope, chest pain, hemoptysis, ankle edema, and sudden death (9). We are reporting an interesting case of calcified left atrial myxoma presented by atrial fibrillation and single episode of angina pectoris like symptoms, mimicking coronary heart disease.

CASE REPORT

A 49-year-old female, Caucasian, presented with episodes of frequent chest discomfort and palpitations accompanied by chronic fatigue. Symptoms lasted for two months, and at one occasion she had intensive chest pain episode during moderate physical effort, lasting for 15-20 minutes, which has forced her to stop physical activity. Beside those symptoms, she was generally in good health, no history of chronic diseases or major surgeries, and family history was negative for heart or neoplastic diseases. Initial physical examination and blood tests did not reveal any serious disorders, electrocardiogram (ECG) was without abnormalities, while the chest X-ray showed only moderate

increase in cardio-thoracic diameter. Our working diagnosis was paroxysmal heart arrhythmia with possibility of coronary heart disease in the background. We ordered some additional tests: echocardiography, thyroid gland hormones, 24h heart ECG monitoring (Holter ECG) and exercise stress test. The patient was also instructed to measure blood pressure at home twice a day and to keep a record until next visit. The symptoms continued and the patient also added an observation of "feeling weight on the heart", especially during the night if she lied on her left side. Thyroid hormones and exercise stress test were normal, as well as blood pressure values measured at home. ECG monitoring showed normal sinus rhythm with single paroxysm of atrial fibrillation (longer than 30 sec.), 110 ventricular premature complex (PVC) per hour and occasional supraventricular extra systoles. Transthoracic echocardiography pointed to probable source of patient's problems; left atrium was dilated (47 mm) with oval tumor mass inside (33x45 mm). Tumor mass was pendicular, anchored at lower part of interatrial septum and imbued with small hyper echogenic areas, without mitral valve obstruction or possible clots. All other echocardiographic parameters were normal. With the working diagnosis of left atrial myxoma additional tests were done. Chest multi slicing computer tomography (MSCT) and heart magnetic resonance

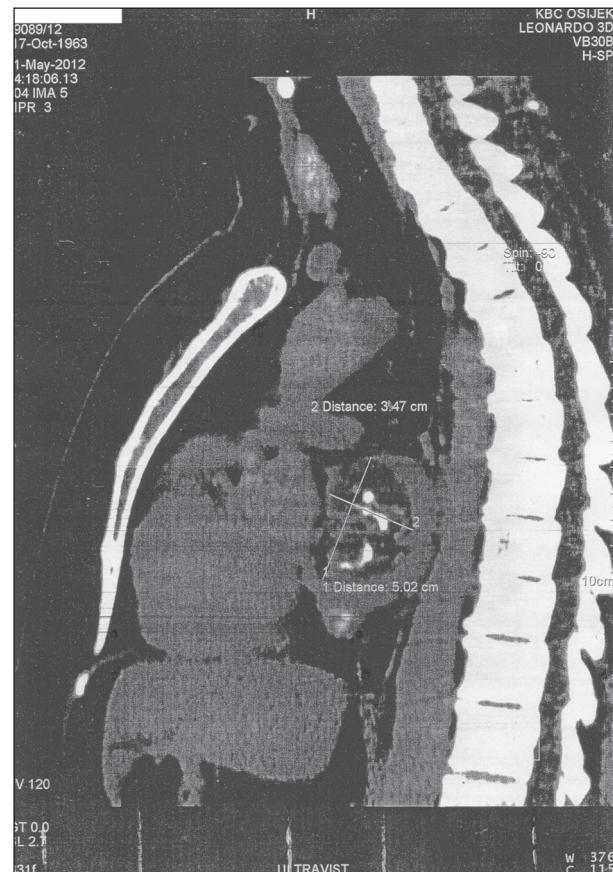


Fig. 1. Magnetic resonance image of atrial myxoma, bright areas represent calcifications.

(MR) detected heterodense calcified structure inside the left atrium, 5 cm in craniocaudal and 3.5 cm in anteroposterior diameter, without significant contrast imbibition (Fig. 1) - features pointing to atrial neoplasm, primarily myxoma. Coronary angiography was normal. The patient also did not report neurological or any other symptom which could be related to thromboembolism. Initially low weight molecular heparin (enoxaparine 2x1mg/kg) and beta blocker (bisoprolol 2.5mg/daily) were introduced. The patient was scheduled for surgery and the tumor was completely removed. Histological analysis confirmed the diagnosis - left atrial pendicular myxoma with diffuse areas of calcifications, various in shape and size, without osseous metaplasia. No additional treatment was recommended. Three months after surgery the patient had no symptoms or complications related to tumor or surgery. Control transthoracic echocardiography showed decrease of left atrial diameter (42 mm), and no atrial tumor or thrombotic masses were observed. 24h ECG monitoring was normal with rare PVC and occasional supraventricular extra systoles. Anticoagulant and beta blocker were discontinued. On a two-year follow up we found no myxoma, nor symptomatic or silent atrial fibrillation relapse.

DISCUSSION

Myxoma is the most common primary cardiac neoplasm found in adults (1,2). Depending on tumor location, size, and mobility symptoms are caused by tumor mass itself or complications (tumor fragmenting, thrombus formation, systemic response) (6,9). Typical symptoms are present in the most of patients, while others are less frequent. Systolic or diastolic murmur may be observed in about half of patients due to obstructed filling of ventricle ("tumor plop"), interference with closure of atrioventricular valves or narrowing the outflow tract (10). Our patient presented only with atypical set of symptoms - predominantly chest discomfort and palpitations, what makes this case a diagnostic challenge. According to series of Burke of 107 myxoma cases only 7 presented with atrial fibrillation and 4 with chest discomfort (9), asserting the low incidence of symptoms related to our case. The patient's assertion about feeling a „weight on the heart“ at a second visit may be biased, because she was informed about the tumor at the echocardiography examination, but we believe this observation was possibly caused by episodes of atrial fibrillation. Although symptoms initially directed our investigation to arrhythmias with suspected coronary heart disease as possible underlying cause, image tests unexpectedly pointed to left atrial myxoma, and the definitive confirmation of diagnosis was made at histological examination after surgical tumor removal.

Tumor morphology and location are all concordant with previous epidemiological and histopathological studies (1,2). Although patchy calcifications are present in 10% to 20% cases, massive calcification is uncommon (7,8). It remains unknown whether this case's somewhat atypical morphology - diffuse calcification, is related to the atypical presentation.

24h ECG monitoring revealed atrial fibrillation paroxysm and occasional PVCs, which is associated with atrial myxoma (9). After the tumor removal, atrial fibrillation symptomatic or silent disappeared, confirmed by multiple 24h ECG monitoring at follow up. This supports our conclusion about myxoma causing atrial fibrillation possibly by transitory left atrial dilatation. As there was no evidence of atrial fibrillation recurrence we decided to stop anticoagulant therapy.

Angiography and exercise stress test excluded coronary disease with high probability, stress echocardiography was unavailable. Cause of single angina like episode remains unclear, possible explanations may be: sudden atrial fibrillation with rapid ventricular response onset, transitory mitral valve obstruction or intercostal neuralgia. Nevertheless, the probability of sub massive pulmonary embolism in association with atrial fibrillation should also be considered.

In many myxoma cases different laboratory abnormalities were observed associated with myxoma itself, complications or other diseases (1,3). In this case all laboratory tests were continually normal.

CONCLUSION

In conclusion, only appearance of atrial fibrillation and its symptoms can be strongly associated to atrial myxoma in this case. Disappearance of atrial fibrillation after myxoma removal, strongly supports this

association. Due to reversible cause of atrial fibrillation we decided to stop anticoagulant therapy at follow up visit. Myxoma location and morphology are typical and only diffuse calcifications somewhat differs from its usual morphology. This might be associated with unusual symptom presentation. By reviewing literature and this case analysis we could not find a credible relation.

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S A Ž E T A K

KALCIFICIRANI MIKSOM ATRIJA KAO RIJEDAK UZROK FIBRILACIJE ATRIJA

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Klinički bolnički centar Osijek, Zavod za bolesti srca i krvnih žila, ¹Zavod za reumatologiju, kliničku imunologiju i alergologiju, Osijek i ²Opća županijska bolnica Našice, Odjel za patologiju, Našice, Hrvatska

Primarni srčani tumori su vrlo rijetki i većina su kod odraslih miksomi. Prikazujemo zanimljiv slučaj ekstenzivno kalcificiranog mksoma lijevog atrija koji se prikazao paroksizmalnom fibrilacijom atrija i simptomima angine pektoris oponašajući koronarnu bolest srca. Tumor je kirurški uklonjen, a histološka analiza potvrdila je pendikularni mksom s difuznim područjima kalcifikacija različitih oblika i veličina. Koronarna bolest srca isključena je invazivnom angiografijom. Nakon uklanjanja tumora atrijska fibrilacija kao i drugi simptomi nisu se više javljali.

Ključne riječi: tumori srca, mksom, kalcifikacije, aritmija, fibrilacija atrija