

CASE REPORT



The riddle of the right ventricle

ANA-MARIA BALAHURA^{1,2}, ANDRADA CAMELIA GUȚĂ^{1,3}, VALENTIN ENACHE⁴, CRISTIAN BALAHURA^{2,5},
ALEXANDRA EMMA WEISS^{1,2}, CRISTINA JAPIE², ELISABETA BĂDILĂ^{1,2}, DANIELA BARTOȘ^{1,2}

¹Clinical Department No. 5, Carol Davila University of Medicine and Pharmacy, Bucharest, Romania

²Department of Internal Medicine, Emergency Clinical Hospital Bucharest, Romania

³Department of Cardiology, Prof. Dr. C.C. Iliescu Emergency Institute for Cardiovascular Diseases, Bucharest, Romania

⁴Department of Pathology, Emergency Clinical Hospital Bucharest, Romania

⁵Department of Gastroenterology, Emergency Clinical Hospital Bucharest, Romania

Abstract

Right ventricular (RV) myxoma is a very rare finding. Its differential diagnosis includes cardiac thrombus, and its risk of life-threatening complications mandates early diagnosis followed by surgical resection. We report the case of a patient with an incidental RV mass and a difficult differential diagnosis. A 66-year-old woman, first assessed in neurosurgery due to a lumbar herniated disc, was referred to cardiology for examination before proceeding to surgery. She complained of dyspnea on exertion present for the last few months and reported no fainting or syncope. Clinical examination showed intermittent pulmonary systolic murmur. Transthoracic echocardiography revealed an oval-shaped sessile mobile mass (42/18 mm) attached to the anterior RV wall. Computed tomography confirmed the presence of a RV mass with lower attenuation than the myocardium and extension towards the pulmonary trunk, without other abdominal or pulmonary masses that would suggest a thrombus. Cardiac magnetic resonance imaging described an ovoid mass (47/16 mm) in the right ventricle, "clinging" to the apical trabeculae, swinging during the cardiac cycle, causing partial obstruction of the pulmonary valve during systole. The patient underwent surgical resection of the tumor. Macroscopic specimen showed a translucent polypoid mass with hemorrhagic areas. Microscopy confirmed the diagnosis of RV myxoma. The case illustrates the difficulty of establishing the correct etiological diagnosis of a cardiac mass, especially when located in the right ventricle. Multimodality imaging remains the cornerstone of noninvasive tissue characterization of cardiac masses, still requiring histopathological confirmation, particularly in the setting of conflicting imaging results.

Keywords: cardiac mass, myxoma, echocardiography, cardiac magnetic resonance imaging.

Introduction

Cardiac tumors represent 0.2% of all tumors found in humans [1]. Of all primary heart tumors, approximately 75% are benign, with myxoma accounting for at least half of these. Approximately 75% of myxomas arise from the left atrium [2], and only a very few cases of right ventricular (RV) myxoma have been described in the literature thus far [1, 3]. Depending on the size and location of the tumor, presenting symptoms may vary. The classic triad found in patients with cardiac myxoma is characterized by obstruction of blood flow (dyspnea, syncope), constitutional symptoms (fever, weight loss), and thromboembolic events (pulmonary or systemic) [4]. Right sided myxomas embolize in approximately 10% of cases and have the potential to cause fatal pulmonary artery obstruction. It has also been theorized that multiple emboli from a right-sided myxoma may lead to the development of pulmonary hypertension [5]. The risk of life-threatening complications indicates the importance of early diagnosis and prompt surgical resection.

Aim

We report the case of a patient with an incidental RV

mass, in whom conflicting results on imaging examinations were further clarified by histopathological (HP) analysis.

Case presentation

We present the case of a 66-year-old woman assessed in neurosurgery for lumbalgia due to a L4–L5 lumbar herniated disc. Having a history of systemic hypertension and a grade 2/6 systolic ejection murmur at the left upper sternal border, she was referred to cardiology for further investigations before proceeding to surgical treatment of the neurological condition.

Detailed anamnesis revealed progressive dyspnea upon moderate exertion in the last few months, but no episodes of fainting or syncope. Patient had no history of major cardiac or thromboembolic events. At the time of the cardiovascular examination, no murmurs were detected, heart sounds were normal at 75 beats/min, and there was no jugular venous distension, peripheral edema, and no pulmonary rales. Oxygen saturation in ambient air was 96%, blood pressure was 135/80 mmHg and Lasègue's sign was positive, with no other pathological findings upon clinical examination. The electrocardiogram (ECG) performed upon admission showed sinus rhythm,

intermediate QRS axis and no ST-T changes. Chest X-ray findings were unremarkable. Routine laboratory tests revealed thrombocytopenia, hypercholesterolemia, and hypertriglyceridemia. Inflammation markers were negative.

Transthoracic echocardiography (TTE) (Figure 1, A–C) was performed showing mild left ventricle (LV) hypertrophy, with preserved LV ejection fraction (LVEF 55%) and normal LV filling pressures. There was no hemodynamic

significant valvular disease to explain the intermittent ejection murmur. The right heart chambers had normal dimensions and function. Notably, an oval-shaped mobile sessile mass, measuring 42/18 mm, was attached to the anterior wall of the right ventricle, intermittently protruding into the RV outflow tract, and causing mild stenosis, with no hemodynamic impact on the tricuspid valve. However, TTE did not allow definite discrimination between a cardiac tumor and thrombus.

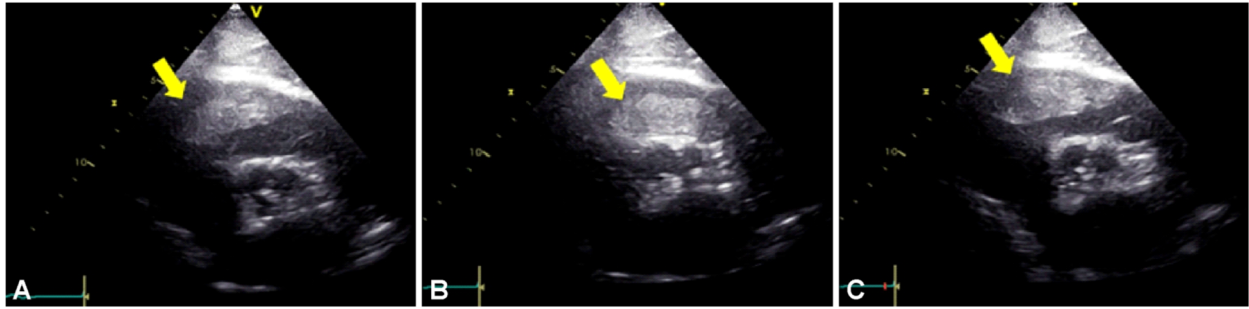


Figure 1 – (A–C) Transthoracic echocardiography, parasternal short axis at the great vessels: systolic and diastolic movement of an oval-shaped sessile, mobile mass (arrow), 42/18 mm diameter, located in the right ventricle. No hemodynamic impact on the tricuspid or pulmonary valves was noted.

A contrast computed tomography (CT) scan of both thoracic and abdominal cavity (Figure 2, A–C) confirmed the presence of a RV mass with lower attenuation than the rest of the myocardium, extending towards the pulmonary trunk, with no other pulmonary or hepatic masses, a description pleading for a ventricular thrombus.

There were no signs of thrombus in any of the venae cavae, right atrium, or pulmonary arteries ramifications, nor was there on venous Doppler ultrasound of the lower limbs. A magnetic resonance imaging (MRI) was considered mandatory before any decision regarding treatment.

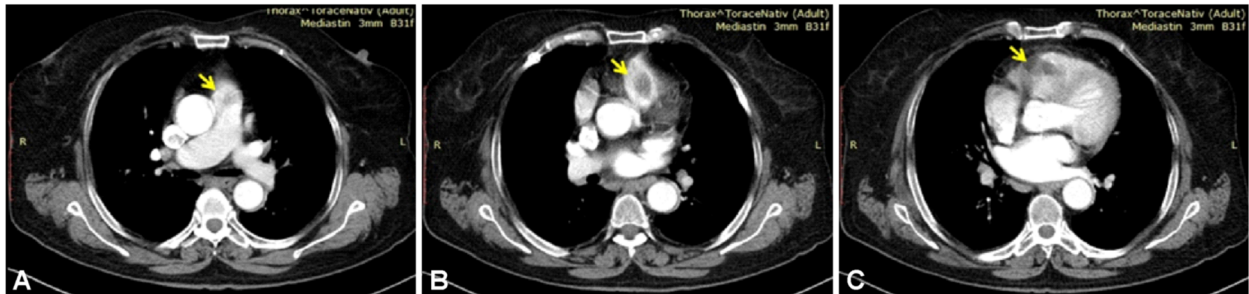


Figure 2 – (A–C) Thoracic computed tomography (CT) with intravenous iodinated contrast showing a filling defect corresponding to an oval-shaped mass in the right ventricular (RV) outflow tract. The mass has no contrast uptake. The right ventricle inflow and the pulmonary arteries are completely opaque upon contrast enhancement.

Further, cardiac MRI (Figure 3, A and B) described the ovoid mass of 47/16 mm in the RV outflow tract, “clinging” onto the apical trabeculae and swinging during cardiac cycle, causing partial obstruction of the pulmonary valve during systole. This mass was isointense with the myocardium in the unenhanced T1-weighted image and remained hypointense in the contrast-enhanced images. The absence of contrast enhancement was consistent with an avascular tumor. Estimated LVEF was 59% and RVEF 64%, and no hypertrophy, chamber dilatation or kinetic anomalies were seen. Therefore, immediate surgical treatment was considered. Coronary angiography was performed to evaluate for the presence of coronary artery disease, and vascular characterization of the mass. It found no significant atherosclerotic lesions, and, interestingly, no signs of neovascularization were detected at the site of

the aforementioned mass. The patient further underwent cardiac surgery and surgical resection of the tumor.

Upon macroscopic examination there was a solid, elastic, brown translucent, polypoid mass of 50/30/20 mm, with an implantation base of 13 mm. The microscopic examination with Hematoxylin–Eosin (HE) staining (Figure 4, A–C) found a proliferation of oval-round cells and spindle cells, with a very low cell density, arranged in an abundant myxoid stroma associated with hemorrhagic and fibrino-hemorrhagic areas, and ectatic blood vessels. These HP findings were compatible with the diagnosis of a RV myxoma.

Postoperative course was favorable, except for repetitive episodes of paroxysmal atrial fibrillation interrupted by the administration of Amiodarone. The patient was discharged home on day 10 after surgery.

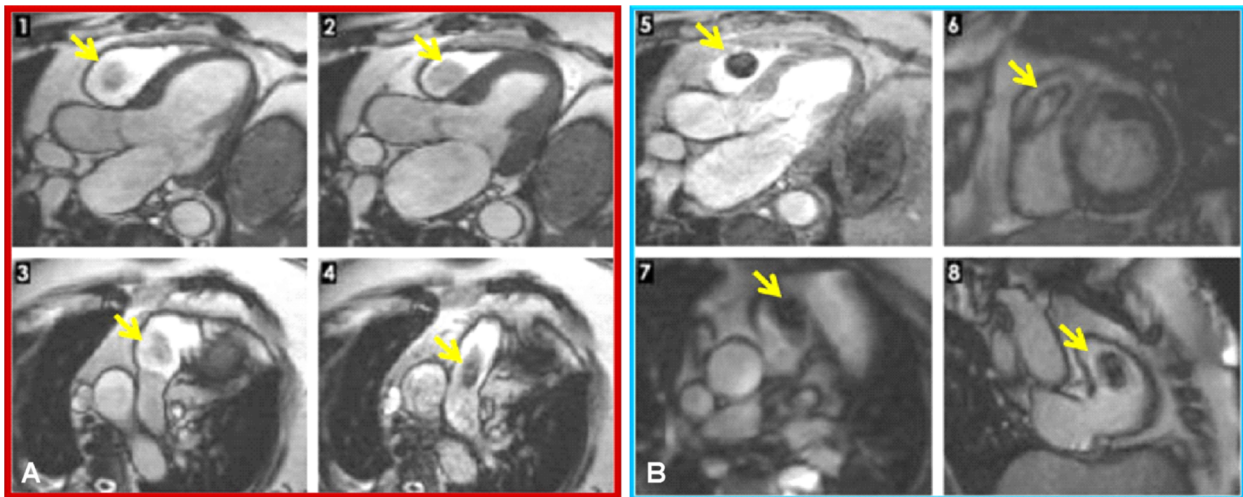


Figure 3 – (A and B) Cardiac magnetic resonance imaging (MRI). Images 1–4 (A) represent unenhanced imaging, and images 5–8 (B) a contrast-enhanced imaging. An ovoid mass of 4.7/1.6 cm is depicted in the right ventricle outflow tract, “clinging” to the apical trabeculae, and swinging during the cardiac cycle, causing partial obstruction of the pulmonary valve during systole. On contrast-enhanced imaging, the mass remains dark because of no contrast uptake.

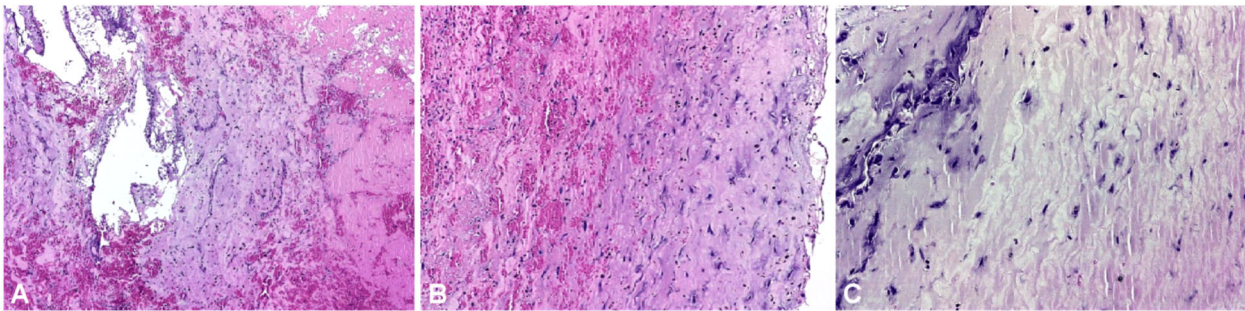


Figure 4 – (A–C) Fusiform and stellate cells, and capillary blood vessels, all disposed in an abundant, rich, myxoid stroma that associates hemorrhagic areas. Hematoxylin–Eosin (HE) staining: (A) 40×; (B) 100×; (C) 200×.

☒ Discussions

Despite de Senac’s statement in 1783 that “the heart is an organ too noble to be attacked by a primary tumour”, the heart has no specific immunity from neoplasia, first recognition of a heart tumor being attributed to Columbus, in 1559 [6]. Czapek, in 1891, was among the first to provide a pathological description of a RV myxoma. Almost 70 years passed before Kishimoto & Sakaibori described another similar tumor, which was recognized at autopsy [7]. In adults, the primary differential diagnosis for a solitary, intracavitary cardiac mass includes thrombus, myxoma, lipoma and non-myxomatous neoplasms, most of which are malignant [8]. In our case, the possibility of metastatic tumor was ruled out by the absence of signs and symptoms of the primary tumor and absence of visceral metastasis on CT scanning.

Myxomas are neoplasms of endocardial origin. The tumor typically projects from the endocardium into the cardiac chamber. It is believed that cardiac myxomas are derived from multipotential mesenchymal cells that persist as embryonal residues during septation of the heart [9]. They can be of variable size, shape, and mobility. On macroscopic appearance, these masses may have smooth or lobulated, polypoid features. The latter tends to associate more with thromboembolic phenomenon, either from friable tumor, or adherent surface thrombus forming on the frond-like areas [10].

The most common differential diagnosis of myxoma is the development of a thrombus. Thrombi account for the most commonly encountered intracardiac masses. They can occur in any of the cardiac cavities, though they most often involve the left-sided heart. Thrombus formation is frequently caused by hypercoagulable states, systolic dysfunction with wall motion abnormalities, atrial fibrillation, or artificial valves. A thrombus typically appears as a hypodense, low attenuation filling defect in a contrast pool within a cardiac chamber and may be differentiated from primary and secondary tumors by considering predisposing risk factors, attachment location, shape, and lack of mobility. They are frequently crescent-shaped filling defects with broad based attachments. Nevertheless, a pedunculated appearance has been observed and can mimic myxomas. RV thrombus is a rare finding and is usually associated with intravenous catheters, arrhythmogenic RV dysplasia, Behçet’s disease, metastatic disease, and trauma [2, 11]. Clinical symptoms may be similar in myxoma and RV thrombus, particularly with respect to intracardiac obstruction and peripheral embolization. However, the precise determination of the nature of the mass is important because treatment modalities vary depending on etiology [7].

Right-sided tumors may present with repeated episodes of syncope, shortness of breath, fatigue, night sweats and right heart failure, and may progress rapidly despite medical treatment. Most patients though have a variety of different and atypical symptoms. That is why some authors call

myxomas “the great masquerader” [10]. Asymptomatic cases of large tumors are extremely rare [12]. Our patient was asymptomatic for a long time before mild dyspnea occurred. The diagnosis was made incidentally and was suggested initially by echocardiography.

Routine laboratory findings are usually non-specific and non-diagnostic. If present, abnormalities may include: increased erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), leukocytosis, thrombocytopenia, normochromic or hypochromic anemia, polycythemia, and increased interleukin (IL)-6 serum level [1, 6]. Radiographic findings are usually non-specific, as was also seen in our case.

Two-dimensional echocardiography is the investigational modality of choice in cases of cardiac masses, allowing the physician to identify the location, size, shape, attachment, and mobility of the tumor, with an accuracy within 1 mm to 3 mm [13]. The most important distinguishing feature of a myxoma is the characteristic narrow stalk, followed by tumor mobility and distensibility, although broad based, non-mobile myxomas can also occur. On microscopy, myxomas may be either homogeneous, or have central areas of hyperlucency, representing hemorrhage and necrosis. Calcification with echogenic foci may also be detected [8]. In the case of our patient, the RV mass had an atypical location, however showing features consistent with a possible primary cardiac tumor – large, ovoid, and pedunculated, an aspect suggestive for a myxoma.

Cardiac CT and MRI are often used synergistically with echocardiography in the evaluation and management of cardiac masses. On cardiac CT, approximately two-thirds of myxomas are ovoid, with a smooth or lobular shape, with the remainder having a villous appearance. When visualized on non-contrast CT, they typically appear hypodense, having the same intensity of blood, and may demonstrate calcifications. On contrast-enhanced cardiac CT, myxomas appear as intracavitary filling defects with heterogeneous contrast enhancement, though the intensity may be variable depending on their chronicity and whether necrosis or hemorrhage is present [2].

Cardiac MRI is often the preferred imaging modality for cardiac masses because it provides specific information about tissue characteristics that facilitate distinction between myxoma and thrombus. Hypointensity in T1-weighted images relative to the myocardium, and hyperintensity in T2-weighted images, demonstrating tissue with high extracellular water content are commonly observed in myxomas. Moreover, myxomas show a heterogeneous appearance in MRI, due to areas of necrosis, hemorrhage, or calcification. In gadolinium-enhanced MRI, myxomas high neovascularization typically occurs as a heterogeneous pattern of contrast enhancement. In contrast, atrial thrombi have a brighter appearance than myocardium in images with short inversion time, and a darker appearance in images with long inversion time, and almost never show contrast enhancement in terms of neovascularization [14]. In addition, MRI provides information with respect to localization, site of insertion and size of the mass. It is characterized by high temporal resolution and multiplane imaging possibilities [14]. In our case, cardiac MRI showed absence of contrast enhancement suggesting an avascular tumor such as cardiac thrombi.

Catheterization of the diseased ventricle is contraindicated due to the risk of tissue embolization. However,

angiographic imaging of the coronary arteries can be useful in differentiating between cardiac myxoma and thrombus, since tumors, such as myxomas, are considered highly vascularized, with supplies from branches of the left or right coronary arteries. Furthermore, coronary angiography can expose concomitant coronary artery disease (especially in patients over 40 years old) [6]. Rahmanian *et al.* reported, in a study including 23 patients with cardiac myxoma undergoing coronary angiography, that signs of neovascularization were only present in 12 (52%) cases [14]. In our case, no signs of neovascularization were revealed with coronarography.

Integrating findings of tissue characteristics, neovascularization, anatomical results, and clinical symptoms concludes the preoperative diagnosis of cardiac tumor. Assessing the nature of the mass facilitates the choice of the appropriate therapeutic approach. In patients with a diagnosis of myxoma, surgical intervention is the treatment of choice. In those with cardiac thrombus anticoagulation therapy is strongly recommended. Furthermore, the use of anticoagulation may potentially be harmful, increasing the risk of peripheral embolization in the presence of a cardiac tumor [14, 15]. On the other hand, cardiac mass biopsy prior to surgical removal may lead to catastrophic embolization when it proves to have actually been thrombus [3].

In our case, due to conflicting imaging results, surgery and mass HP analysis were mandatory both for diagnostic, as well as treatment purposes. HP analysis was the final diagnostic test that established the correct diagnosis when non-invasive tissue characterization with cardiac MRI was inconclusive.

The precise rate of growth of cardiac myxomas is unknown, although it is believed to be reasonably fast, which poses an increased risk for peripheral embolization, cardiac valvular obstruction, or sudden cardiac death. Surgical resection is curative. Interventions are generally considered urgent, especially if there is a history of embolism or syncope [10].

Prognosis in solitary myxomas after complete surgical resection is good, as evidenced by the low post-operative mortality rate of 1–3% [16, 17]. Due to their location, removal of the tumors may result in supraventricular arrhythmias or atrio-ventricular node dysfunction requiring treatment [18]. In our case, atrial fibrillation occurred soon after surgery, and antiarrhythmic treatment with Amiodarone was started. No recurrences at three months and 12 months on 24-hour Holter monitoring were detected. Late recurrences have been reported to occur in 0.4–5% of surgically treated patients from three months to 22 years after operation [19]. Recurrence is usually attributable to incomplete excision of the tumor, or intraoperative metastatic dissemination from the primary tumor. Thus, long-term clinical and echocardiographic follow-up is mandatory [6].

☐ Conclusions

This case illustrates the difficulty of establishing the correct etiological diagnosis of a cardiac mass especially when located in the right ventricle. Multimodality imaging remains the cornerstone of noninvasive tissue characterization of cardiac masses. However, definite diagnosis of

a myxoma is histopathological, particularly with conflicting imaging results and the rare localization in the right ventricle.

Conflict of interests

The authors declare that they have no conflict of interests.

Funding

This work was conducted on the MODERNIZE project platform (Modernization of infrastructure in the center of research – development in minimally invasive interventional medical techniques in internal medicine and gastroenterology), funded by the National Authority of Scientific Research and Innovation, in the name of the Ministry of European Funds, through the Operational Program Increase of Economic Competitiveness, Priority Axis 2 – Operation 2.2.1 (POSCCE-A2-0.2.2.1-2013-1), co-financed by the European Regional Development Fund.

References

- [1] Nina VJS, Silva NAC, Gaspar SFD, Rapôso TL, Ferreira EC, Nina RVAH, Lages JS, Silva FACC, Filho NS. Atypical size and location of a right atrial myxoma: a case report. *J Med Case Rep*, 2012, 6:26. <https://doi.org/10.1186/1752-1947-6-26> PMID: 22269461 PMCID: PMC3277469
- [2] Kassop D, Donovan MS, Cheezum MK, Nguyen BT, Gambill NB, Blankstein R, Villines TC. Cardiac masses on cardiac CT: a review. *Curr Cardiovasc Imaging Rep*, 2014, 7(8):9281. <https://doi.org/10.1007/s12410-014-9281-1> PMID: 25018846 PMCID: PMC4090749
- [3] Knight TE, Shiramizu B, Ly P, Thompson KS, Reddy V. Paroxysmal nocturnal dyspnea secondary to right ventricular myxoma: a novel presentation of an unusual tumor. *Case Rep Pediatr*, 2018, 2018:4791379. <https://doi.org/10.1155/2018/4791379> PMID: 29682382 PMCID: PMC5851333
- [4] Gribaa R, Slim M, Kortas C, Kacem S, Ben Salem H, Ouali S, Neffati E, Remadi F, Boughzela E. Right ventricular myxoma obstructing the right ventricular outflow tract: a case report. *J Med Case Rep*, 2014, 8:435. <https://doi.org/10.1186/1752-1947-8-435> PMID: 25515693 PMCID: PMC4301803
- [5] Gopal AS, Arora NS, Messineo FC. Right ventricular myxoma. *N Engl J Med*, 2000, 342(4):295. <https://doi.org/10.1056/NEJM200001273420418> PMID: 10660393
- [6] Lone RA, Ahanger AG, Singh S, Mehmood W, Shah S, Lone G, Dar A, Bhat M, Sharma M, Lateef W. Atrial myxoma: trends in management. *Int J Health Sci (Qassim)*, 2008, 2(2):141–151. PMID: 21475496 PMCID: PMC3068734
- [7] Gajjar TP, Shah GB, Desai NB. Giant ventricular myxoma obstructing right ventricular outflow tract. *Rev Bras Cir Cardiovasc*, 2011, 26(4):663–666. <https://doi.org/10.5935/1678-9741.20110060> PMID: 22358285
- [8] van der Heusen FJ, Stratmann G, Russell IA. Right ventricular myxoma with partial right ventricular outflow tract obstruction. *Anesth Analg*, 2006, 103(2):305–306. <https://doi.org/10.1213/01.ane.0000226144.27041.67> PMID: 16861407
- [9] Hecser L, Siklodi KP, Matei D, Jung H, Csiki G. Right ventricular cardiac myxoma. Histopathology diagnosis in forensic autopsy case. *Rom J Leg Med*, 2011, 19(2):79–82. <https://doi.org/10.4323/rjlm.2011.79>
- [10] El Sabbagh A, Al-Hijji MA, Thaden JJ, Pislaru SV, Pislaru C, Pellikka PA, Arruda-Olson AM, Grogan M, Greason KL, Maleszewski JJ, Klarich KW, Nkomo VT. Cardiac myxoma: the great mimicker. *JACC Cardiovasc Imaging*, 2017, 10(2):203–206. <https://doi.org/10.1016/j.jcmg.2016.06.018> PMID: 28183439
- [11] Scheffel H, Baumüller S, Stolzmann P, Leschka S, Plass A, Alkadhi H, Schertler T. Atrial myxomas and thrombi: comparison of imaging features on CT. *AJR Am J Roentgenol*, 2009, 192(3):639–645. <https://doi.org/10.2214/AJR.08.1694> PMID: 19234259
- [12] Obrenović-Kirčanski B, Mikić A, Velinović M, Božić V, Kovacević-Kostić N, Karan R, Parapid B, Djukić P, Savić D, Vranes M. Right ventricular myxoma – a case report. *Vojnosanit Pregl*, 2013, 70(6):609–611. <https://doi.org/10.2298/vsp1306609o> PMID: 23885530
- [13] Vozzi CR, Pechacek LW, Garcia E, Mathur VS, De Castro CM, Hall RJ. Two-dimensional echocardiography in the diagnosis of unusual left atrial myxomas. *Cardiovasc Dis*, 1980, 7(3):246–256. PMID: 15216251 PMCID: PMC287861
- [14] Rahmanian PB, Castillo JG, Sanz J, Adams DH, Filsoufi F. Cardiac myxoma: preoperative diagnosis using a multimodal imaging approach and surgical outcome in a large contemporary series. *Interact Cardiovasc Thorac Surg*, 2007, 6(4):479–483. <https://doi.org/10.1510/icvts.2007.154096> PMID: 17669910
- [15] O'Rourke F, Dean N, Mouradian MS, Akhtar N, Shuaib A. Atrial myxoma as a cause of stroke: case report and discussion. *CMAJ*, 2003, 169(10):1049–1051. PMID: 14609975 PMCID: PMC236231
- [16] Kalra DK, Hemu M, Kyung S, Reddy V, Rao A, Volgman A. Atrial myxoma – the great masquerader. *QJM*, 2019, 112(5):363–364. <https://doi.org/10.1093/qjmed/hcz042> PMID: 30759245
- [17] Shah IK, Dearani JA, Daly RC, Suri RM, Park SJ, Joyce LD, Li Z, Schaff HV. Cardiac myxomas: a 50-year experience with resection and analysis of risk factors for recurrence. *Ann Thorac Surg*, 2015, 100(2):495–500. <https://doi.org/10.1016/j.athorac.2015.03.007> PMID: 26070596
- [18] Owers CE, Vaughan P, Braidley PC, Wilkinson GA, Locke TJ, Cooper GJ, Briffa NP, Hopkinson DN, Sarkar PK. Atrial myxomas: a single unit's experience in the modern era. *Heart Surg Forum*, 2011, 14(2):E105–E109. <https://doi.org/10.1532/HSF98.20101163> PMID: 21521672
- [19] Mahmoud HM, Moursi I. A rare case of a big left ventricular myxoma presenting with a cerebrovascular stroke. *Egypt Heart J*, 2014, 66(4):375–377. <https://doi.org/10.1016/j.ehj.2014.03.005>

Corresponding author

Elisabeta Bădilă, Associate Professor, MD, PhD, Clinical Department No. 5, Carol Davila University of Medicine and Pharmacy, Bucharest; Department of Internal Medicine, Clinical Emergency Hospital Bucharest, 8 Floreasca Road, Sector 1, 014461 Bucharest, Romania; Phone +4021–599 23 00 / 102, Fax +4021–317 01 79, e-mails: elisabeta.badila@gmail.com, elisabeta.badila@umfcd.ro

Received: February 19, 2020

Accepted: January 3, 2021