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DOI: 10.5603/FM.a2022.0040

Article type: Case report

Submitted: 2022-01-07

Accepted: 2022-03-07

Published online: 2022-04-05

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Abstract

Cardiac myxomas are the most common primary cardiac tumors in adults. They usually present as a solitary, solid mass in the left atrium. Their most common radiographic appearance is that of a hypodense lesion on computed tomography (CT) and inhomogeneous lesion (hypo to isointense on T1 sequences and hyperintense on T2 sequences) on magnetic resonance (MR) with some contrast enhancement. However, different patterns are recognized due to secondary changes within the tumor. We present a case of a 60-year-old man with a hypervascular myxoma. The lesion was a sessile mass located in the left atrium and rigidly attached to the interatrial septum. On CT and MR, it showed vivid contrast enhancement due to intratumoral flush of arterial blood form branches of dominant left circumflex artery and a possible fistula to the left atrium. Furthermore, we review the literature for different atypical radiographic appearances of myxomas.

Key words: cardiac tumor, radiology, computed tomography, magnetic resonance

CASE REPORT
A 60-year-old man was referred to our institution with a diagnosis of a cardiac tumor.

The patient complained of itching chest pain after exertion. Otherwise, his past medical history was unremarkable. The primary work-up was performed outside our institution. As far as diagnostic imaging is concerned, these investigations consisted of coronary angiogram, transthoracic and transesophageal echocardiography. The coronary angiogram showed no significant stenotic lesions, but demonstrated atypical branches of the dominant left circumflex artery converging to the interatrial septum. Transthoracic and transesophageal echocardiography revealed a sessile mass located in the left atrium and attached to the interatrial septum. The lesion contained calcifications, had a visible blood flow on Color Doppler imaging, and measured 35x45mm. No other significant abnormalities were noted. Chest radiograph showed no abnormalities.

On admission the patient was asymptomatic, and his vital signs (heart rate, blood pressure, blood oxygen saturation level, temperature) were within normal limits. Physical examination revealed systolic murmur in the 2nd right intercostal space. Basic metabolic panel showed slight elevation of ALT level (95U/l, normal range 7-56U/l). Otherwise, his physical, laboratory and electrocardiographic examinations were unremarkable.

Cardiac computed tomography (CT) and magnetic resonance (MR) were performed to better characterize the mass. CT was performed using a 320-row dynamic volume CT scanner (Aquilion One, Toshiba Medical Systems, Ottawara, Japan) with administration of contrast agent (Iomeron-400; Bracco, Milan, Italy). The protocol consisted of ECG-gated non-contrast, arterial and delayed phases. Arterial phase was triggered when a contrast enhancement threshold of 180HU was exceeded in the descending aorta followed by a 5 seconds delay. Delayed phase was aquired 60 seconds after contrast administration. A four-phasic intravenous injection regimen was used: 5ml of saline + 60ml of contrast agent + 30ml of 50% contrast agent and 50% saline + 40ml saline chaser at a flow rate of 6ml/s. The detailed parameters of CT imaging are presented in Table I. MR was performed using a 1.5T clinical whole-body MR system (MAGNETOM Avanto; Siemens AG, Erlangen, Germany) with administration of contrast agent as a bolus dose of 0.1 mmol/kg of Gadobutrol (Gadovist, Bayer Schering, Berlin, Germany) followed by a 20ml saline flush at a flow rate of 3ml/s. The cardiac MRI protocol contained HASTE, SSFP CINE, T2-TIRM, T1, T1FS, perfusion and DE-PSIR sequences in several planes (typical cardiac
2CH, 3CH, 4CH and SA as well as specifically adjusted to transect the lesion). The detailed parameters of MRI sequences are summarized in Table II.

On native CT the lesion was slightly hypodense, with some calcifications (fig. 1). Arterial phase showed flush of the contrast medium from at least two branches of dominant left circumflex artery to the center of the mass and a possible narrow connection with the left atrium cavity suggestive of a fistula (figs. 2 and 3). It also confirmed no significant coronary artery disease. On delayed images it was hard to depict any contrast enhancement of the mass beyond the vascular part (fig. 1). On MR the lesion was inhomogeneous, hypo to isointense on T1 sequences and hyperintense with some hypointense foci on T2 sequences (fig. 4). Perfusion images showed the already known flush of arterial blood in the center of the mass. Delayed images demonstrated weak, heterogeneous enhancement of its remaining part (fig. 5). Both studies were suggestive of a myxoma with an intratumoral fistula between branches of circumflex artery and left atrium.

The patient underwent successful resection of the tumor. The postoperative period was uneventful. The histopathological examination confirmed the diagnosis of a myxoma.

**DISCUSSION**

We present a case of an atypical radiographic appearance of a cardiac myxoma.

Myxomas are the most common primary cardiac tumors in adults accounting for about 50% of cases [1]. They are usually found in the middle-aged population (mean age of about 50) with female predominance [2].

The typical appearance is that of a solitary mass in the left atrium, attached to the interatrial septum, with narrow attachment base, lobulated contours, and a diameter of 3-4cm [1]. On CT myxomas present as hypodense lesions with a very weak contrast enhancement and calcifications in about 10-20% of cases. On MR they are inhomogeneous, hypo to isointense on T1 sequences, hyperintense on T2 sequences, with heterogeneous contrast enhancement. Myxoid matrix shows very high signal intensity on T2 sequences. Secondary changes within the tumor (e.g., calcifications, fibrosis, cysts, and various stages of hemorrhage) are responsible for their heterogeneity on both T1 and T2 sequences.

However, many atypical radiographic patterns are found in the literature.
As mentioned before, the vast majority of myxomas are solitary lesions located in the left atrium, with attachment to the interatrial septum. Multiple lesions occur very rarely and are usually associated with genetic syndromes. The best-known connection is Carney complex described in 1980s [3], a multiple endocrine neoplasia syndrome including skin pigmentation, cardiac and extracardiac myxomas, pituitary adenoma, psammomatous melanotic schwannoma, testicular tumors and osteochondromyxoma. Seldomly, multiple lesions occur without a known genetic predisposing factor [4]. As far as multiplicity of myxomas is concerned, they have a well-established potential to metastasize. The most common site of metastases is brain [5]. In the literature there are found cases of myxomas’ metastases to many other organs including pancreas, kidneys, stomach, bones and even skin [6]. Location of myxomas in other parts of left atrium or in other cardiac chambers is also very rare. It occurs more often in multiple lesions in genetic syndromes. There are published cases of myxomas in all cardiac chambers - right atrium, biatrial, left ventricle, right ventricle, right ventricle outflow tract [7] and even pulmonary artery [8].

Neither width of attachment to the interatrial septum nor contour of the tumor's surface is a discriminating factor. Some tumors are mobile, pedunculated, with a narrow attachment base, while others are non-mobile, sessile, and rigidly attached to the cavity wall. The length of the stalk in pedunculated lesions determines mobility of the mass. The longer the stalk, the more mobile the lesion and more probable to cause obstruction of the atrio-ventricular tract. Surface of the tumors vary from smooth, through lobulated, to even very irregular. Furthermore, the tumor may be covered with a thrombus, which most often occurs in the irregular type. Tumor size and its growth rate are variable. At presentation tumors measure usually 3-4cm (ranging from 1 up to 15cm). Their reported mean growth rate is 4-5mm per year. Some remain stable for several years, while others grow fast mimicking malignant tumors [1].

Myxomas may contain variable portions of postnecrotic and posthemorrhagic calcifications and cysts. Likewise, vascularization of these tumors is highly variable. Calcifications are present in about 10-20% of myxomas [1]. Some lesions are highly calcified with myxomatous tissue undetectable by means of diagnostic imaging [9]. In these cases, preoperative differential diagnosis with calcified amorphous tumor, thrombi, vegetations or caseous calcification of mitral annulus may be impossible. Cystic components are far less common. Multiplicity and size of the cysts are variable. Some
lesions resemble a single cyst with only a small solid nodule [10] and some have a 
multicystic appearance [11]. Differential diagnosis of cystic myxomas include hydatid cyst, 
bronchogenic cyst, interatrial septum aneurysm, varices, and foramen ovale cyst. The vast 
majority of myxomas are hypovascular and enhance weak after contrast agent 
administration. Some lesions may show no contrast uptake and simulate a thrombus. 
Differentiation between myxoma and a thrombus may be even more challenging as they 
may coexist. MR post-contrast T1 sequences with high inversion times (500-600ms) are 
helpful in the differential diagnosis. They are very specific for thrombi, which show no 
signal intensity. On the other hand, some myxomas are highly vascular or contain a fistula 
between coronary arteries and cardiac chambers. Stiver et al. [12] presented a case of a 
myxoma with a well-documented myocardial ischemia due to myxoma’s steal 
phenomenon. We present a similar case of a patient with an exertion chest pain and a 
highly vascular cardiac myxoma with a possible fistula between branches of the left 
circumflex artery and left atrium. The symptoms of our patient cannot be directly 
correlated with the presence of a highly vascular myxoma. These symptoms did not meet 
the criteria of typical angina and we have not performed a stress test to confirm the 
association. Follow-up studies will reveal whether these symptoms pass or remain, and the 
tumor was only an incidental finding.

CONCLUSIONS

We present a case of an atypical radiographic appearance of a highly vascular 
myxoma. Additionally, we review the literature for other atypical radiological patterns of 
these tumors. The summary is presented in Table III. When dealing with cardiac tumor one 
should keep in mind the possibility of atypically located, multiple and metastasizing 
myxomas. These tumors may contain variable portions of calcifications, cystic and 
vascular components and thus, simulate other masses.

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**Table I.** CT acquisition parameters

| Parameter                      | Non-contrast + contrast-enhanced phases |
|--------------------------------|-----------------------------------------|
| Tube voltage [kV]              | 120                                     |
| Tube current [mA]              | Smart mA                                |
| Rotation time [s]              | 0.275                                   |
| D-FOV [mm]                     | 220.0                                   |
| Focus                          | Small                                   |
| Slice thickness / interval [mm]| 0.5 / 0.25                              |

FOV – field of view, kV – kilovolts, mA – milliamperes, s – seconds, mm – millimeters

**Table II.** MR sequences’ parameters

| Sequence Parameter | HASTE | SSFP CINE | T2-TIRM | T1 + T1FS | PERFUSION | DE-PSIR |
|--------------------|-------|-----------|---------|-----------|-----------|---------|
| Plane              | Axial | 2CH, 3CH, 4CH, SA | SA, specific | Specific | Specific | SA, specific |
| Repetition time [ms] | 800       | 42.3 (2,3CH); 40.05 (4CH); 57.86 (SA) | 664       | 740       | 160.76   | 700     |
| Echo time [ms]     | 40     | 1.12      | 47       | 30        | 1.05     | 1.21    |
| Flip angle [deg.]  | 160    | 80 (2,3,4CH); 79 (SA) | 180       | 180       | 12       | 45      |
| iPAT factor        | 2      | 2         | -        | 2         | 2        | 2       |
| Number of signal averages | 1 | 1 | 1 | 1 | 1 | 1 |
| FOV [mm]           | 370    | 340 (2,3CH,SA); 380 (4CH) | 360       | 340       | 360      | 340     |
| FOV phase [%]      | 75     | 80.4 (2,3,4CH); 81.3 (SA) | 81.3      | 81.3      | 75       | 68.8    |
| Breath-hold        | No     | Yes       | Yes      | Yes       | Yes      | Yes     |
| Resolution [mm]    | 2.4x1.4x8 | 1.5x1.5x6 (2,3CH); 1.7x1.7x6 (4CH); | 1.9x1.4x8 | 2.2x1.3x5 | 2.8x2.3x10 | 2.5x1.8x8 |
2.5x1.4x8 (SA)

HASTE - half-Fourier acquisition single-shot turbo spin-echo, SSFP CINE – steady-state free-precession cine, TIRM -turbo inversion recovery magnitude, FS – fat saturation, DE-PSIR – delayed enhancement phase-sensitive inversion recovery, 2CH – two chamber plane, 3CH – three chamber plane, 4CH – four chamber plane, SA – short axis plane, iPAT – integrated parallel acquisition techniques, FOV – field of view, ms – milliseconds, deg – degrees, mm – millimeters

**Table III.** Atypical patterns of cardiac myxomas

| Feature                        | Atypical finding       | Report                      |
|--------------------------------|------------------------|-----------------------------|
| Multiplicity                   | Intracardiac Genetic   | Carney J. A., 1985 [3]      |
|                                | Non-genetic            | Kataoka S., 2016 [4]        |
| Extracardiac metastases        |                        | Wan Y., 2019 [5]            |
|                                |                        | Terata Y., 2000 [6]         |
| Location                       | Cardiac chambers besides left atrium | Katiyar G., 2020 [7]        |
|                                |                        | Baris V., 2016 [8]          |
| Secondary changes within the tumor | Large degree of calcifications | López-Marco A., 2014 [9]    |
| Cysts                          | Single                 | Park J. K., 2013 [10]       |
|                                | Multiple               | Singhal A., 2017 [11]       |
| Hypervascularity               |                        | Stiver K., 2015 [12]        |

**Figure 1.** A. Axial non-contrast CT image. B. Axial delayed phase contrast-enhanced CT image. Images demonstrate a hypodense mass measuring 35x45mm in the left atrium. The lesion contains calcifications (yellow arrow on image B) and contrast-enhancing vascular part (red asterisk on image B). Size of cardiac chambers is within normal limits. There are no other cardiovascular abnormalities. RA – right atrium, RV – right ventricle, LA – left atrium, LV – left ventricle.

**Figure 2.** A. Axial arterial phase CT image. B. MIP reconstruction of sagittal arterial phase CT images. Images demonstrate the mass measuring 35x45mm in the left atrium with its vascular and non-vascular components. They present enlarged, tortuous feeding vessels.
(yellow arrows on image B) and flush of arterial blood in the vascular nidus of the tumor (red asterisks on both images). The non-vascular part of the mass remains hypodense. Size of cardiac chambers is within normal limits. There are no other cardiovascular abnormalities. RA – right atrium, RV – right ventricle, LA – left atrium, LV – left ventricle, Ao – aorta.

**Figure 3.** Coronary CT reconstruction. Image shows ascending aorta and coronary tree. It presents domination of left coronary artery and separate origins of left anterior descending (LAD) and left circumflex (LCX) arteries from the left aortic sinus. Two enlarged branches of left circumflex artery (yellow arrows) feed the vascular part of the mass (red asterisk). No significant coronary artery disease is noted. Ao – aorta, LAD – left LCX – left circumflex artery.

**Figure 4.** A. 4-chamber T1 image. B. 4-chamber T1 fat-saturated image. C. Short axis T2 fat-saturated image. D. Axial HASTE image. Images demonstrate heterogeneity of the mass (red asterisk) on MR. Secondary changes within the tumor (i.e., calcifications and vascular component) are responsible for the hypointense foci on T2 weighted sequences (image C). The lesion contains no fat as it has similar signal intensity on T1 weighted sequences with and without fat saturation (images A and B). Size of cardiac chambers is within normal limits. There are no other cardiovascular abnormalities. RA – right atrium, RV – right ventricle, LA – left atrium, LV – left ventricle.

**Figure 5.** 4-chamber T1 delayed post-contrast image. Image reveals contrast enhancement of both myxoid and vascular parts of the mass (red asterisk). The vascular component follows signal intensity and contrast enhancement of blood on all MR sequences. The myxoid part shows some contrast enhancement (red arrow) and a non-enhancing cap (yellow arrow) on delayed imaging. Size of cardiac chambers is within normal limits. There are no other cardiovascular abnormalities. RA – right atrium, RV – right ventricle, LA – left atrium, LV – left ventricle.
