

Three dimensional echocardiographic imaging of multiple recurrent myxomas

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Abstract

We report a case of a recurrence of 5 cardiac myxomas in both atria with atypical anatomical features difficult to image. Although a multimodality imaging was performed, three-dimensional

echocardiography (3DE) was the only technique able to correctly identify all the recurrences and the anatomical characteristics of the myxomas. MRI detected the blood supply of the mass but even after careful review was able to identify only 4 of the 5 lesions. Even though it was already reported the usefulness of 3DE to better delineate the site of attachment of cardiac tumors, it was never reported its sensibility in the setting of multiple myxomas; this case highlights the ability of the 3DE in this challenging scenario and its potential for being considered the key adjunctive modality for the anatomy when advanced surgical plan is required.

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Key words: Three-dimensional echocardiography; cardiac myxomas; magnetic resonance imaging; two-dimensional echocardiography.

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Introduction

Cardiac myxomas are the most common primary cardiac tumors, with an estimated incidence of 0.5 per million individuals annually [1-3]. The majority of cardiac myxomas manifest as endocavitary gelatinous masses that arise adjacent to the fossa ovalis in the left atrium. However, some of them have atypical localization such as the right atrium (18% of cases) and, more rarely, the aorta, pulmonary artery, ventricles and vena cava; they are also, though rarely, found in the roof of the left atrium [1-5]; they exhibit a strong preference for women aged 40-60 years [1-5]. They are typically pedunculated but can also arise in a sessile fashion [1-5]. The detection by imaging of the typical anatomical findings easily lead to the diagnosis [6,7]; however, being the myxomas mimicker of other diseases both from an imaging and clinical point of view [6,8,9], in atypical and unusual cases, their diagnosis is very challenging. After surgery, recurrences have been observed in up to 3% of cases, except when myxomas occur as part of an underlying genetic syndrome [10,11].

The role of two-dimensional (2D) echocardiography (E) and magnetic resonance imaging (MRI), mainstay in the diagnosis and clinical decision making in cardiac myxomas, has been extensively described in medical literature [6,7]; few reports have been published suggesting an incremental usefulness of the three-dimensional (3D) echocardiography in this scenario [12-14].

We report a case of multiple biatrial recurrences of a cardiac myxoma with rare localizations where 3DE was the only technique able to correctly identify all the recurrent lesions.

Case Report

A 33-year-old lady who underwent left atrial myxoma resection was found to have on 3 years follow up transthoracic (TT) echocar-

diography a large multilobular mass in the right atrium (RA). The RA mass was partially protruding into the tricuspid valve during diastole with no significant obstruction to flow. It was not possible to correctly detect the type of the insertion of the tumor; no other masses were found; except for the above findings, the study was unremarkable. The patient was not aware of other occurrences of cardiac myxomas in family members. Further 2D transesophageal echocardiography (TEE) showed two multi-lobulated masses in RA: one bigger (measuring around 4 x 3 cm, difficult to correctly size for its asymmetric morphology) attached by a peduncle to the superior lateral RA wall (Figure 1A, green arrow) and one smaller attached by a stalk to the inferior RA wall; a remnant likely a suture was seen on the right side of the fossa ovalis; two additional masses were detected in the left atrium: a small (size 0.7 x 1.4 cm) sessile mass on the left atrial (LA) side of the interatrial septum (IAS) (Figure 1A, white arrow) close to the scar of the previous surgery; and a small sessile (size 0.6 x 0.8 cm) mass attached to the mitral annulus close to the posteromedial commissure and P3 scallop of the posterior leaflet of the mitral valve. 3D TEE allowed an anatomical imaging able to identify two pedunculated right atrial masses (Figure 1C, green arrow: peduncle attached to superior RA wall) and three sessile LA masses: one on the left side of the interatrial septum close to the previous resection area (Figure 2C, white arrow; Figure 2A, white arrow); one at the opening of the (left atrial appendage (LAA) (Figure 2A, blue arrow); and one on the mitral annulus in the area of the posterior commissure of mitral valve (Figure 2B, yellow arrow). MRI multifunctional assessment CINE, tissue characterization with T2 weighted edema imaging with fat saturation and first pass Gadolinium perfusion imaging and late enhancement imaging identified and showed perfusion of: two masses in the RA with the pedicles (Figure 1B, green arrow point at the stalk); one mass on the LA side of interatrial septum (Figure 1B, white arrow); one mass on the

mitral annulus close to mitral valve posterior commissure (Figure 1B, yellow arrow); however, it was not able to detect the small mass close to the LA appendage (A) even after careful review by an expert reader.

The patient underwent reoperation through a biatrial approach: two lesions were found in the right atrium, one multilobular with two consistencies, one solid and one myxomatous, both connected by a thick pedicle to the right atrial wall superiorly and laterally. This one was resected with the underlying attachment to the right atrium; another separate lesion with a separate pedicle and attachment to the right atrial wall more inferiorly was also detected and it was also resected with the adjoining atrial tissue. Three lesions were found in the left atrium: one, about 1 cm of diameter, close to the scar for the last surgery and also other two, smaller in size; one close to the left atrial appendage (Figure 2C, blue arrow) and one close to the posterior commissure of the mitral valve (Figure 2D, yellow arrow). The mitral valve was slightly involved during the resection of the lesion from the valvular annulus and was repaired by 4-0 Prolene sutures. Five specimens were sent to pathology; all of them, upon histopathologic study, were consistent with benign cardiac myxoma. No major clinical criteria suggestive of Carney Syndrome (skin, conjunctiva and lips lentiginos, subclinical hypercortisolism and nodular thyroid changes) except than cardiac myxoma and no mutations of PRKARIA gene were found. After 18 months follow up, no recurrences have been detected.

Discussion

Multiple recurrences of myxomas are rare [1-3,6,10-11]. Several large case series of patients with non-familial cardiac myx-

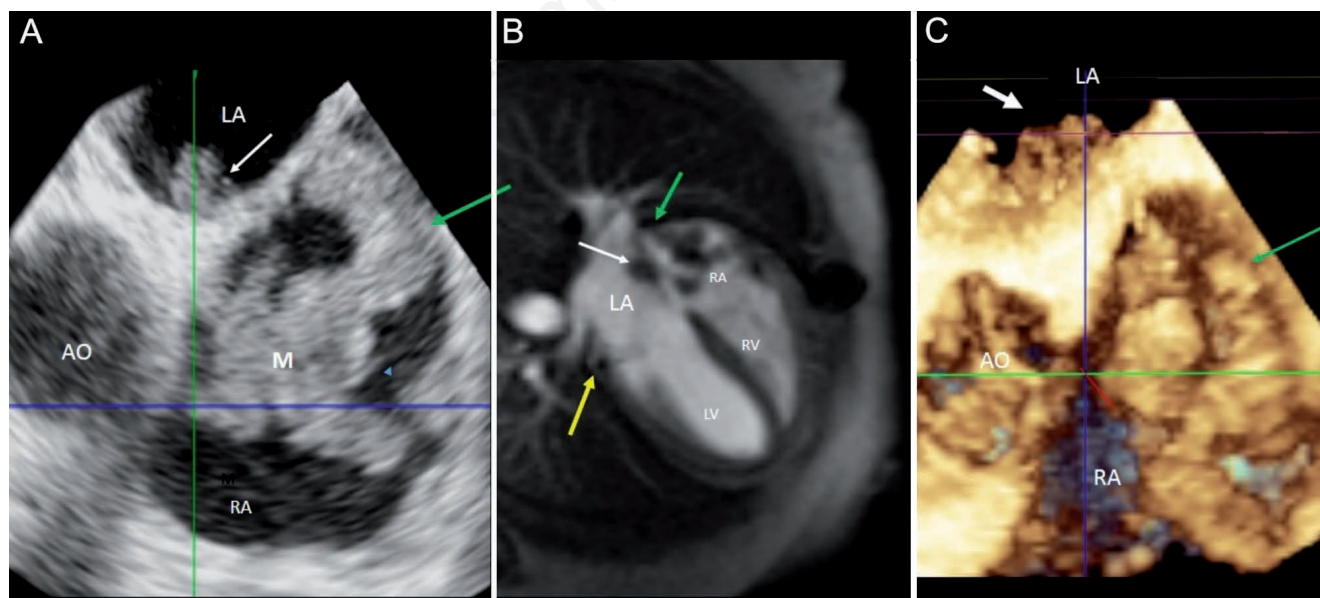


Figure 1. A) 2D TEE: view at the level of the atria; green arrow point at the peduncle of the multilobular mass attached to the superior lateral wall; white arrow points at left atrium mass on the left side of the interatrial septum (IAS); M, myxoma. B) MRI with first pass perfusion; 4 chamber view: white arrow points at left atrium mass on the left side of the IAS; green arrow points at the stalk attached to the superior lateral RA wall; yellow arrow points at the left atrium mass close to the posterior commissure. C) 3DE: view of the two atria after cropping of atrial wall; green arrow points at the stalk attached to the superior lateral RA wall; white arrow points at LA mass on the left side of the interatrial septum (IAS); M, mass; RA, right atrium; LA, left atrium; AO, aorta; RV, right ventricle; LV, left ventricle.

omas have demonstrated recurrence rates of up to 3% almost in one chamber and very rarely in multiple chambers [10,11]. Possible causes of multiple recurrences include incomplete excision of the original tumor, growth from secondary “pre-tumorous” foci and malignant transformation. Multifocality has been reported to be the reason for recurrence in most cases; in the present case, several recurrent myxomas arose concurrently at different sites, which suggests multifocal disease [10,11]. A multimodality imaging for detecting the anatomical features and the perfusion of the lesions is mainstay in considering myxomas in the differential diagnosis and excluding other tumors like metastasis or sarcomas [6,7]. Very rare is the localization in our case; at the best of our knowledge, the localization at the LAA opening in recurrent myxomas was detected in two cases [14,15] and the one close to the mitral valve in one report only [3]. 2D TEE was able to identify 4 of the 5 recurrences and the attachment by a stalk of the two RA masses; MRI confirmed the TEE findings and the perfusion of the masses found at surgery. 3D TEE was the only technique able to identify 5 lesions. In particular 3D TEE arose the suspicion of a lesion close to left atrial appendage by imaging an “en face view” of the left atrium from above and by allowing its electronic sectioning by 2D cross sectional planes; this ability of sectioning in

unconventional views allowed also a clearer identification of the peduncles of the masses in the right atrium.

Even though MRI is considered the gold standard for detecting cardiac tumors [7], after careful review, it was not able to identify the presence of the mass close to LAA. This may be dependent on technical limitations as artifact due to poor breath hold technique. MRI provides excellent morphologic and functional evaluation of the heart and overcomes the limited field of view. MRI has superior tissue characterization to either CT and echocardiography with multiple parameters can be evaluated that include tissue characterization edema, iron content, perfusion, enhancement, fat saturation/suppression and has higher tissue to contrast resolution. Spatial and temporal resolution MRI is less than that of CT or echocardiography which limits the evaluation of small masses or valvular lesions. Scan time is more prolonged with required breath hold technique [7]. CT has excellent detection of fluid and fat and can be used for identification of calcification which currently is not possible with MRI.

Such imaging features made the surgeons aware to better examine the left atrium for discovering any recurrences. In fact, the missing one mass and leaving a residual recurrence would have affected and completely modified the outcome. Cardiac CT that

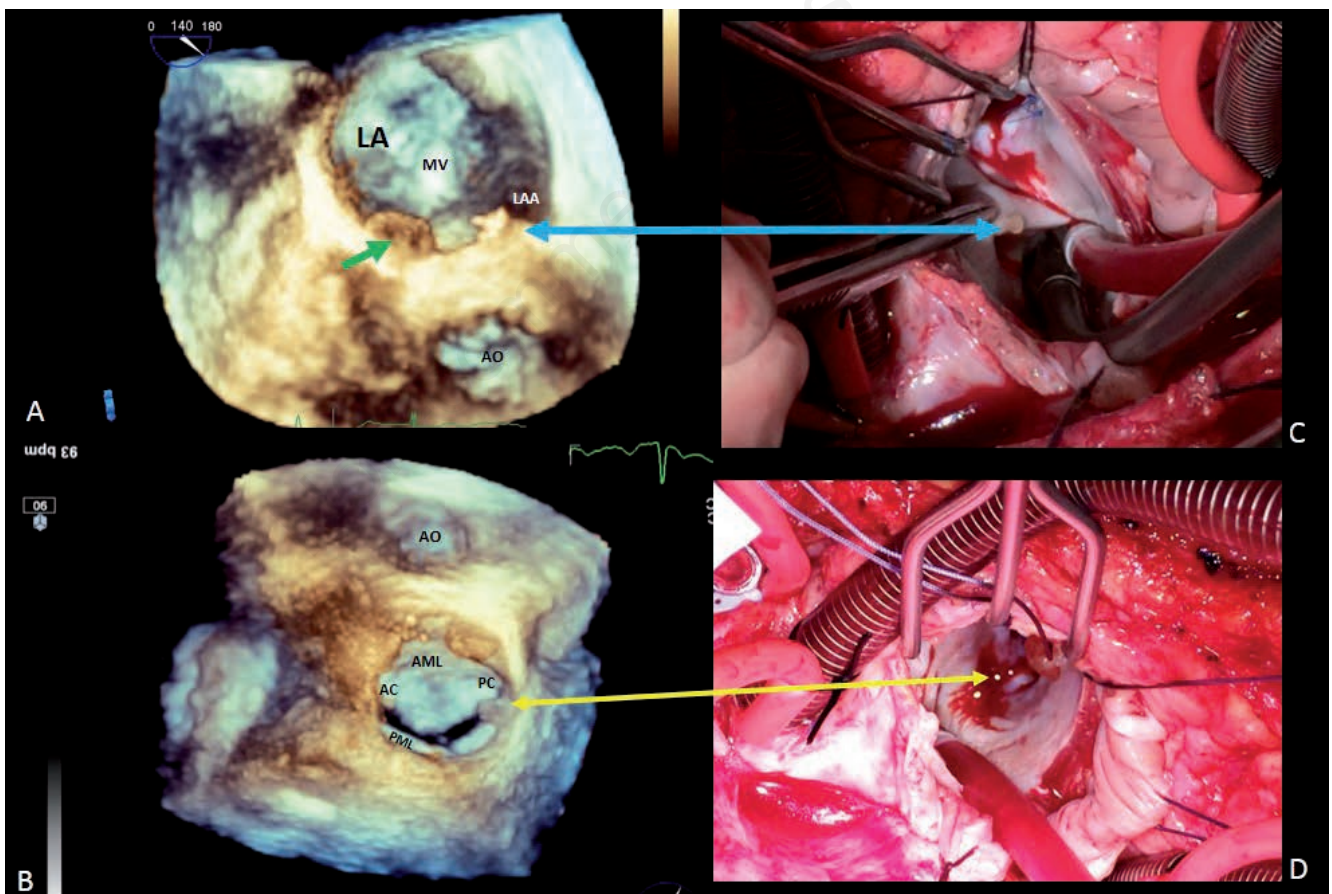


Figure 2. A) 3DE: view of the left atrium from above; green arrow points at the mass attached to the left side of the IAS; blue arrow points at the small mass at the opening of left atrial appendage; MV, mitral valve; AO, aorta; LAA, left atrial appendage opening. B) 3DE: view of the mitral valve from above “surgical view”; yellow arrow points at the mass close to the posterior commissure of the mitral valve (MV); AML, anterior mitral leaflet; PML, posterior mitral leaflet; AC, anterior commissure; PC, posterior commissure. C) Intraoperative picture: blue arrow points at the small mass at the opening of LAA. D) Intraoperative picture: yellow arrow points at the mass close to the posterior commissure of the MV.

has been reported to offer a more detailed spatial imaging compared to MRI and F-fluorodeoxyglucose positron emission tomography/computed tomography able useful information to CT or MRI alone have not been performed: this can be considered a minor study limitation.

Conclusions

A multimodality imaging is required to correctly detect recurrent myxomas by identifying the anatomical features and the perfusion; even though all imaging modalities were able to lead to the diagnosis by describing the anatomical features and the perfusion, 3DE was the only technique able to image all the recurrent myxomas making the surgeons aware to better examine the left chambers and its use has the potential for being considered the key adjunctive modality for the anatomy when advanced surgical plan is required.

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