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Non-Malignant Cardiac Tumors

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Abstract

Cardiac tumors represent an unusual clinical problem in that they are often discovered as an incidental finding during a routine echocardiogram or in the course of a work-up for a source of embolism. Malignant tumors of the heart are either defined as primary or metastatic from an extra-cardiac primary source—regardless, the prognosis is poor. However, there are several cardiac tumors that are characterized as being non-malignant with regard to their tumor biology, but their tendencies to cause embolic or obstructive complications can be just as catastrophic despite a lack of invasiveness or potential to metastasize. The purpose of this chapter is to review the common types of non-malignant cardiac tumors with regard to their incidence, presentation, potential for complications, and management—with emphasis on surgical indications and techniques.

Keywords: cardiac tumor, myxoma, fibroelastoma, cardiac surgery, benign tumors, cardiac lipomas, heart disease

1. Introduction

Embolic strokes are one of the most devastating medical conditions with regard to the overall impact on quality and quantity of life. Once an embolic complication occurs, management options are sometimes limited, but a critical aspect of appropriate disease management is searching for a source of embolism. The same concepts hold true with peripheral embolisms. Part of the rationale for the search for a source is to help determine optimal therapies with the goal of reducing additional embolic events and further complications. Despite substantial resources devoted to stroke (and embolic) prevention, it still remains a considerable problem.

A recent report by the American Heart Association illustrates the enormous burden that strokes represent to society. A cerebrovascular event (i.e., a stroke) occurs every 40 seconds in the United States with a related death occurring every 3.7 minutes [1]. While the causes of strokes are complex and often multi-factorial, cardiac sources represent a common etiology. Atrial fibrillation and associated left atrial appendage thrombi are one of the more frequently encountered sources [2]. Even though mechanical left atrial appendage closure or systemic anticoagulation remain the standard of care for treatment [3], it is important to consider that there are a variety of other cardiac-related causes of embolism and stroke. The most common non-thrombotic causes of cardiac embolism are infectious and non-infectious endocarditis—a topic that is the focus of other chapters [4].

The focus of this chapter is non-malignant and non-infectious cardiac masses—with an emphasis on diagnosis and management. Cardiac tumors are often

delineated as malignant and non-malignant with malignant tumors being either primary (i.e., cardiac sarcomas) or metastatic (i.e., breast carcinoma). They are distinguished from non-malignant tumors, such as myxomas and fibroelastomas, in that the latter, despite the pathologic implications of growth (i.e., valvular obstruction) and systemic embolism, lack true metastatic potential. Nevertheless, non-malignant cardiac tumors can be clinically devastating (i.e., malignant) by their tendency to cause potentially devastating, and occasionally fatal, embolic complications [5].

2. Methods

The focus of this review is on non-malignant tumors. The review methods consisted of Google Scholar (<https://scholar.google.com>) and PubMed (<https://www.ncbi.nlm.nih.gov/pmc/>) searches with emphasis on the following key words: cardiac tumors, benign cardiac masses, myxomas, fibroelastomas, and fibromas. Additional associated search terms included: surgery, diagnosis, imaging, and management. Selected references, including manuscript abstracts and full texts, were reviewed for relevance in the context of this review.

3. Myxomas

Cardiac tumors are rare, occurring at a frequency of 0.0017–0.33% [6]. Cardiac myxomas are one of the most common, comprising 77% of surgically excised tumors in autopsy series [7]. Myxomas affect females predominantly with an incidence 1.5–2 times that of males [8]. The average age at presentation is 53 [8]. The majority of myxomas are sporadic. Inherited forms are less common, seen in 7% of myxomas [9]. Initially reported by Carney in 1985, cardiac myxomas seen in association with pigmented skin lesions and endocrine tumors are collectively known as Carney complex, an autosomal dominant genetic disorder. Familial myxomas tend to affect younger patients and have a higher prevalence among females. In addition, they are more often multicentric with higher rates of embolism and recurrence following resection [9].

Myxomas tend to be rare tumors of mesenchymal origin. They are comprised of stromal cells and are characterized as being benign (**Figure 1**). Biochemically, they

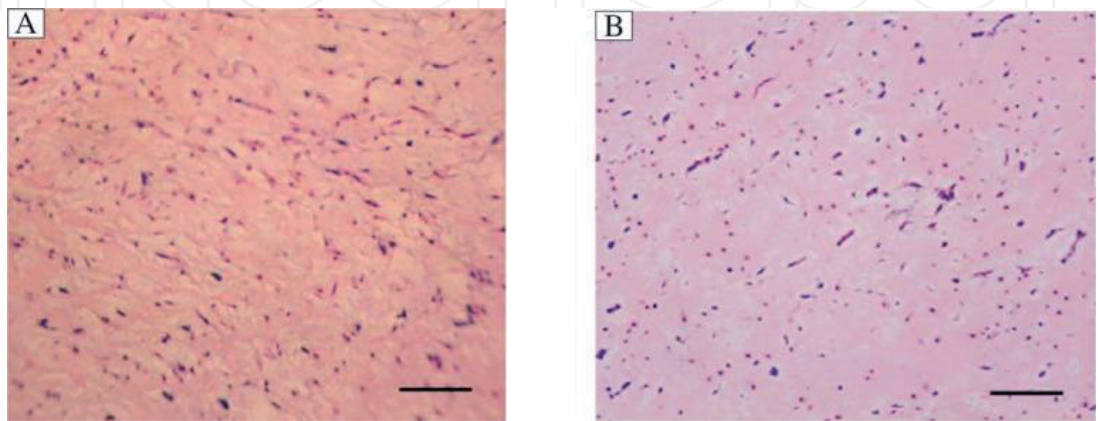


Figure 1. Histology of cardiac myxoma. Representative histology of cardiac myxoma tissue in a 26-year-old woman with multiple recurrences of cardiac myxoma (HE, 100 \times). A: Tumor in 2005. B: Tumor in 2010. Similar histologic appearance of A and B with irregular and papillary proliferations in the myxoid stroma. Bar = 30 μ m [10].

have been correlated with increased production of interleukin 6 (IL-6), however, the significance of this is unclear [11]. While ocular, cutaneous, intramuscular, and juxta-articular involvement has been described, the most common presentation is intra-cardiac with most (85%) involving the left atrium [12].

Cardiac myxomas are typically solitary lesions and most commonly arise from the septal endocardium near the fossa ovalis [8]. 85% arise from the left atrial septum and 11% arise from the right atrial septum [7]. The clinical presentation of myxomas is dependent on tumor location. When confined to the left atrium, myxomas present with symptoms of mitral valve stenosis [13]. Dyspnea and orthopnea result from pulmonary edema and left-sided heart failure. Conversely, right atrial myxomas cause tricuspid valve stenosis, leading to symptoms of right-sided heart failure [13]. In 22% of patients, embolism may occur, leading to symptoms of peripheral ischemia or stroke [8]. 20% of patients develop systemic symptoms, which are attributed to production of IL-6 by tumor cells [14]. Rarely, myxomas may become infected with symptoms similar to those of infective endocarditis [13].

Echocardiography is the primary diagnostic modality for cardiac myxomas [15]. Typical echocardiographic findings are of a mobile mass arising from the septal endocardium, attached by a narrow stalk. Echocardiography is preferred to MRI and CT imaging due to enhanced spatial and temporal resolution. If echocardiography is non-diagnostic, MRI findings of a heterogeneous mass bright on T2-weighted imaging and CT findings of a heterogeneous mass with low attenuation are consistent with myxoma (**Figures 2 and 3**) [15]. Advanced imaging is often performed to help differentiate “benign” myxomas from more aggressive or potentially malignant cardiac tumors that might require a more comprehensive oncologic management strategy (i.e., aggressive debulking, adjuvant chemotherapy, or even palliative care for advanced tumors). While the imaging characteristics, as described above, of myxomas are often diagnostic, unusual appearing masses might prompt further imaging to rule-out other tissue types. The differential diagnosis for such masses includes teratomas (rare), lipomas, angiosarcomas, rhabdomyomas, and rhabdomyosarcomas [18]. Distinguishing characteristics often consist of intramyocardial tumor invasion on imaging. Tissue biopsy is rarely indicated as the risk of embolism from endomyocardial biopsies will typically prompt definitive surgical resection for primary diagnosis and therapeutic intervention. Tumor location and imaging characteristics, along with a detailed history and physical, can also help distinguish cardiac tumors from other types of intra-cardiac pathology, such as endocarditis or thrombus. A recent history of acute myocardial infarction or known left ventricular

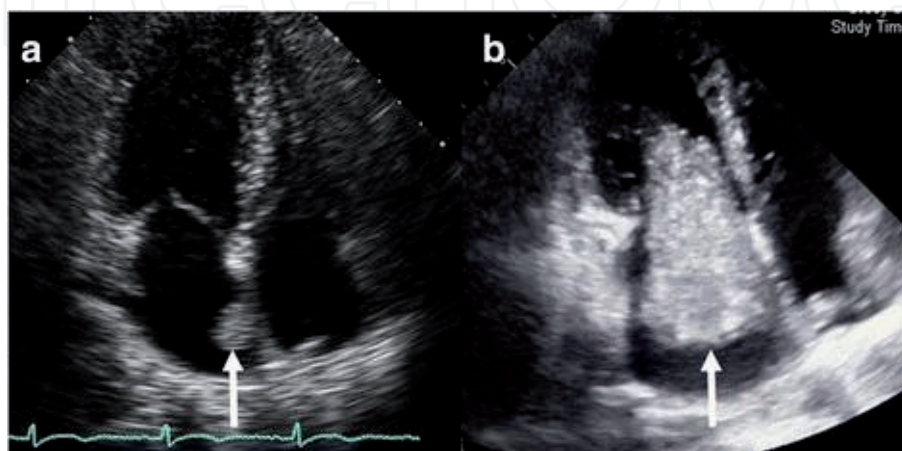


Figure 2.
Representative echocardiographic images of a large left atrial myxoma. From: Surgical resection of cardiac myxoma. Images of a rapidly growing myxoma. (a): A small myxoma is attached to the left atrial side of the fossa ovalis. (b): An enlarged myxoma passes in and out of the mitral valve according to the cardiac cycle [16].

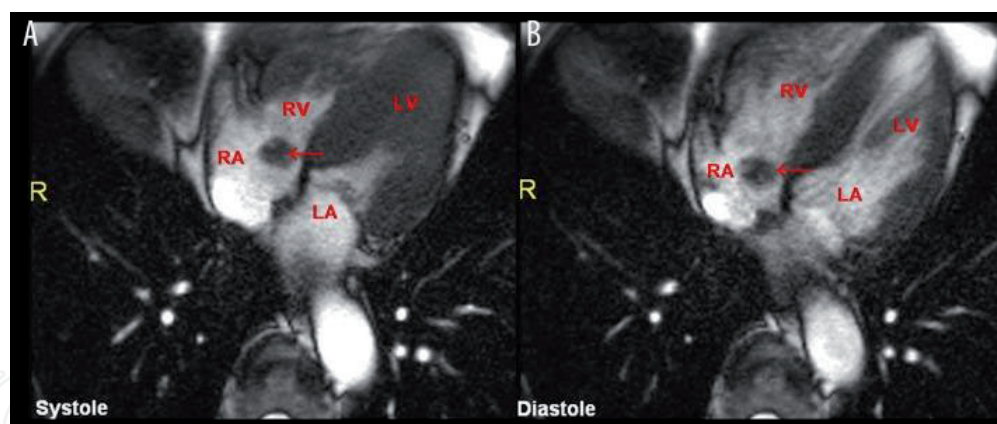


Figure 3. Representative cardiac MRI demonstrating a right atrial myxoma. Right atrial myxoma causing syncope. A: Cardiac MRI showing atrial myxoma during systole. B: Cardiac MRI showing atrial myxoma during diastole. RA, Right atrium; RV, Right ventricle; LA, Left atrium; LV, Left ventricle; arrow—Myxoma [17].

dysfunction might predispose to apical thrombus just as a history of atrial fibrillation is known to predispose to left atrial or left atrial appendage thrombus [19, 20].

Histologically, myxomas are composed of stellate mesenchymal cells in a background of myxoid stroma. Myxoma cells are variably positive for S-100, CD31, and CD34 [13]. In addition, 73.9% of cardiac myxomas express calretinin [21]. Inactivating mutations in the PRKAR1A gene are observed in both sporadic and non-sporadic myxomas. For this reason, routine immunohistochemical staining for PRKAR1A is recommended [22].

Recommended treatment for suspected cardiac myxomas, regardless of size, is immediate surgical resection due to embolic risk [23]. Tumor size ≥ 4.5 cm and soft tumors have been identified as independent risk factors for embolism. Prognosis is favorable with a 92.7% 10-year survival rate following surgical resection. Tumor recurrence is extremely rare. Multicentricity, observed with Carney complex, is an independent risk factor for recurrence following surgical resection [23]. Recurrence has been associated with incomplete resection and family history of complex or multiple myxomas [24]. In general, recurrence rates are typically less than 3%, often in complex or unusual cases [25].

4. Papillary fibroelastomas

Papillary fibroelastomas comprise less than 10% of primary cardiac tumors and are the most common primary tumor of cardiac valves (**Figure 4**) [14]. Men and women are affected equally at an average age of 60. Fibroelastomas are now thought to be more prevalent than myxomas, contradicting previous autopsy series in which myxomas were the most common primary intracardiac tumor [26]. Their etiology is unclear with development related to organizing thrombi, hamartoma proliferation, chronic viral endocarditis, and repeated hemodynamic trauma [27]. Iatrogenic cases of fibroelastomas following thoracic radiation and cardiac surgery have also been described, though these are typically non-valvular [28]. Fibroelastomas are valvular in 90% of cases, most commonly involving the aortic and mitral valves [29]. Less commonly, the left ventricular endocardium and tricuspid valves are affected [15]. Diseased valves are affected in 69.5% of cases, specifically post-rheumatic valves in 37.8% and fibrotic calcified valves in 62.2%. This finding has led some to speculate that a contributing factor to their development is repeated trauma to the cardiac valve surface from abnormal intra-cardiac blood flow and turbulence [13].

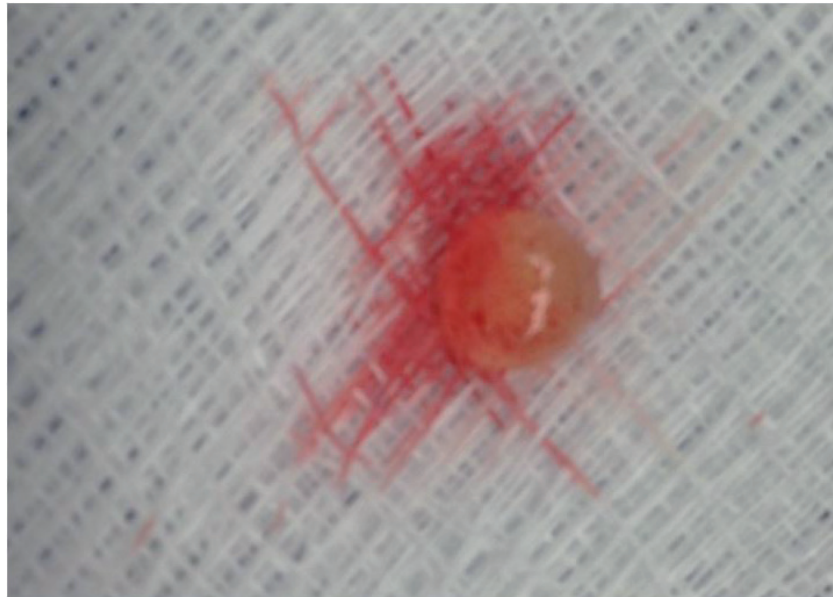


Figure 4.
Surgical specimen of a fibroelastoma. Surgical specimen of an incidental 5 mm fibroelastoma found during routine intra-operative transesophageal echocardiography in a patient undergoing routine coronary artery bypass surgery.

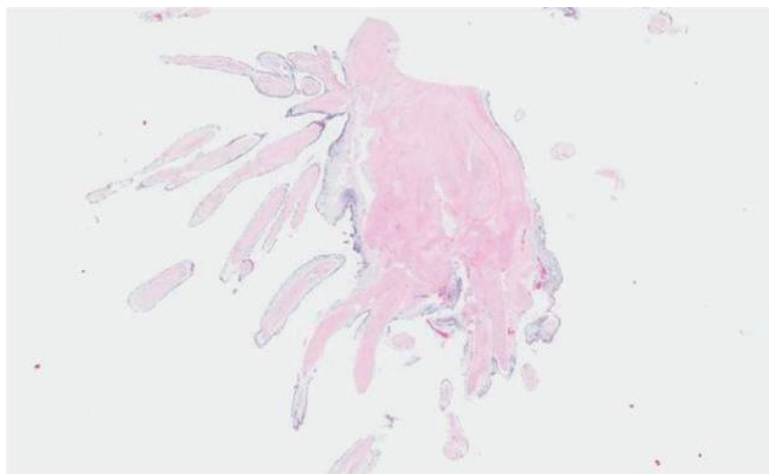


Figure 5.
Photomicrograph of a fibroelastoma. Photomicrograph of papillary fibroelastoma. From: Robotic excision of aortic valve papillary fibroelastoma and concomitant maze procedure [30].

Clinically, fibroelastomas may present with acute embolism following platelet and fibrin aggregation [8]. Alternatively, prolapse of fibroelastomas adjacent to coronary ostia may lead to angina, syncope, and sudden death. The diagnosis is made by echocardiography, which demonstrates a small, homogenous mobile mass attached to a valve by a short pedicle [15]. They have characteristic papillary fronds and resemble sea anemones (**Figure 5**) [31]. These papillary projections give fibroelastomas characteristic stippled edges on echocardiographic imaging [32]. Fibroelastomas may be mistaken for Lambl's excrescences, which are mobile frond-like lesions that occur along lines of valve closure [33]. Interestingly, there have been some anecdotal reports of spontaneous regression. For this reason, intraoperative transesophageal echocardiography is clearly indicated prior to surgical intervention [34]. However, these reports should not be used to advocate non-operative management except in very high-risk patients. In addition, there is no evidence to suggest a role for anti-platelet or anti-coagulation therapy as a means of treatment or secondary prevention once an embolic complication occurs. Nevertheless, in

patients who are not surgical candidates, such medical therapy might be reasonable. In theory, very small tumors could be drawn into the cardiopulmonary bypass circuit or surgical suction prior to excision precluding pathologic evaluation. In such cases, pre- and post-bypass imaging is critical to not only confirm the diagnosis, but also to demonstrate resection.

Although transthoracic echocardiography can be used to screen for papillary fibroelastomas, transesophageal echocardiography is preferred due to higher resolution and enhanced imaging capability (**Figure 6**) [36]. Multiplanar transesophageal echocardiography identifies the exact point of endocardial attachment, which facilitates operative planning. One of the limitations of echocardiography is its inability to stratify risk of embolization based on lesion characteristics [37]. Fibroelastomas are usually not visualized on MRI and CT imaging [15]. Histologically, fibroelastomas have a central core of dense connective tissue, which is surrounded by loose connective tissue and lined with hyperplastic endothelial cells [31]. These surface endothelial cells express vimentin and CD34. These findings have unclear clinical significance, but are helpful in establishing a pathologic diagnosis [13].

Surgical resection sparing underlying valve tissue is recommended in cases of papillary fibroelastoma. In a study of 511 cases over a 15-year period at the Mayo Clinic, 185 patients (36.2%) underwent surgical resection [37]. Primary excision was performed in 51% while excision as an adjunct to other cardiac surgery was performed in 49%. The aortic valve was most commonly affected and in 98% of cases, the native valve was preserved. Three hundred and twenty-six patients (63.8%) with echocardiographic findings of papillary fibroelastoma were managed non-operatively. Patients with papillary fibroelastoma suspected on echocardiography who did not undergo surgical resection had higher rates of stroke and mortality [37].

In the above described Mayo Clinic operative series, there was a 98% native valve preservation rate and 1.6% recurrence rate. Most importantly were their neurologic embolic outcomes. For the surgical population, the stroke risk was 2% at 1 year and 8% at 5 years. This rate was approximately 2.5x age-matched controlled. For the medically managed patients, the 1- and 5-year stroke risk was significantly higher than the operative group at 6 and 13%, respectively. There were 29 observed strokes versus 8.4 expected. Obviously, it was difficult to determine the impact of confounding risk factors that might have increased the stroke risk in patients who were otherwise poor surgical candidates. Regardless, the incidence was still nearly 3.5x that of matched controls. Furthermore, in the non-operative group, medical management with anti-thrombotic therapy (i.e., anti-platelet and anti-coagulant therapy, including dual therapy) had no impact on the stroke risk at 5 years when compared

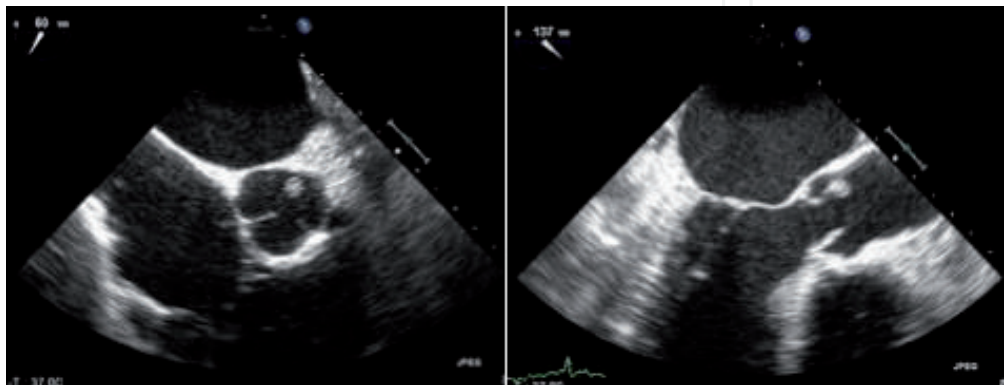


Figure 6. Echocardiographic imaging of a fibroelastoma. Fibroelastoma of the aortic valve. Short and long axis view transesophageal echocardiography. From: Surgery for cardiac papillary fibroelastoma: A 12-year single institution experience [35].

to those without anti-thrombotic therapy [37]. Overall, the authors did not find any echocardiographic or clinical variables that helped stratify patients into high- or low-risk groups for embolic complications. The significant increased stroke risk in the non-operative group (even when matched for age-adjusted controls) and the lack of benefit of medical therapies serve as contemporary evidence to justify surgical management as first-line therapy in appropriate risk-stratified patients (**Figure 7**).

Surgical intervention for papillary fibroelastoma should be considered in patients who are symptomatic, undergoing cardiac surgery for other reasons, or have large, highly mobile lesions [38]. Further study through randomized controlled, multicenter trials is needed to determine if the potential benefit of surgical resection outweighs risk in asymptomatic patients. In asymptomatic patients who are otherwise good surgical candidates, the decision for non-operative management should be well-documented as part of shared decision-making with the patients with emphasis on the theoretical risks and benefits of surgery versus the risks of embolic complications.

In another single-center review, there was a tendency for occurrence in elderly males (71% males and 57% older than 61 years of age). Most (72%) occurred on a cardiac valve. Rarely was more than 1 lesion encountered and rarely were they >1.5 cm in size (27.8%). Surgical management was uncomplicated in all cases, even though some patients required concomitant surgery (i.e., coronary artery bypass grafting), and 30-day survival was 100%. No recurrences were reported at 1-year follow-up [39]. Similar outcomes were reported from another large-volume program in which Mkalaluh and colleagues reported 0 peri-operative mortalities in 11 patients (7 of whom had valvular involvement) with 100% 1-year survival and 91% survival at a mean follow-up of 4.2 years [40].

The indications for surgical resection are often a function of presentation and whether the patient is an appropriate surgical candidate. Since many fibroelastomas are incidental findings and hence asymptomatic, the natural history is unclear even though their tendency to embolize as pedunculated masses is unpredictable. For this reason, in appropriate surgical candidates, the presence of a presumed fibroelastoma is often an indication for surgical management, especially with left-sided

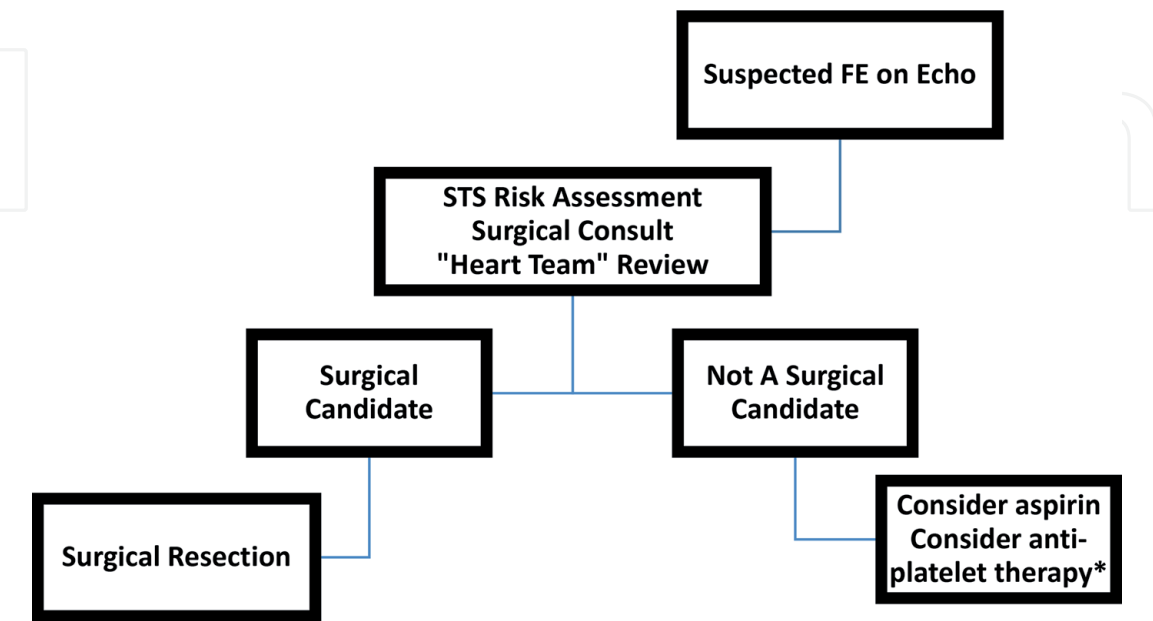


Figure 7.
Proposed algorithm for left-sided possible fibroelastoma. Legend: Adopted from Tamin et al. [37].
FE: Fibroelastoma. Note: there is little evidence to support the overall recommendation for anti-platelet or anti-coagulant therapy in this population.

lesions. While tumor size has not been correlated with embolic risk [41], there is some evidence to suggest that in medically managed patients, the risk of neurologic events was as high as 22% [42]. Nevertheless, the risks of non-operative management are poorly understood. Advocating for surgical intervention must be individualized as part of shared decision-making [43].

5. Cardiac lipomas

Cardiac lipomas, much like lipomas encountered elsewhere in the body, are typically composed of mature adipose (fat) cells and are well-encapsulated. The majority of cardiac lipomas are subendocardial (50%), while the remainder are myocardial (25%) or subpericardial (25%) (**Figure 8**). They are typically found in the left ventricle or right atrium. Embolic complications are extremely rare unless the tumor is coated with thrombus from abnormal flow patterns. Typically, the presentation is characterized by obstructive symptoms [46, 47]. Surgical resection is typically reserved for symptomatic patients and consistent of removal of the entire capsule with pericardial reconstruction of the residual defect if necessary [48]. Asymptomatic patients can be managed expectantly. It is important to note that lipomas must be distinguished from lipomatous hypertrophy of the intra-atrial septum. Lipomatous hypertrophy is considered a benign infiltrative process of the adipose septal tissue. However, obstructive symptoms and even complex atrial arrhythmias can develop from cellular proliferation. In such symptomatic cases, surgical debulking and reconstruction may be considered [49].

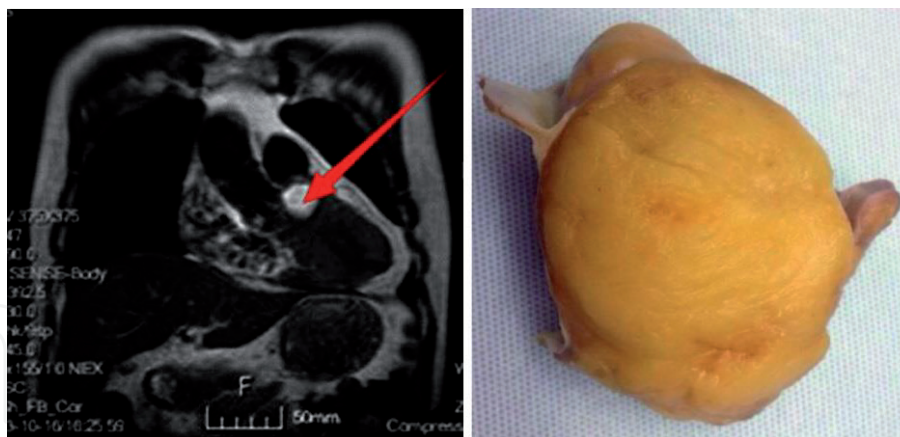


Figure 8.

MRI and surgical specimen of a cardiac lipoma. Left figure: Cardiac lipoma in the interventricular septum. Cardiac MRI showed a round-like signal, measuring 36 × 20 mm, in the upper portion of IVS, anterior to the right coronary sinus of aorta [44]. Right figure: Cardiac lipoma. The surface of the specimen is lined by smooth endocardial and epicardial tissue. The cut surface displays a yellow, lobulated appearance without hemorrhage, necrosis, fleshy change, or calcification [45]. Note: The surgical specimen is from the same patient as the MRI (both cc: Figures with this marker are used under the terms of Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms).*

6. Unusual/rare non-malignant cardiac tumors

While myxomas and fibroelastomas are the most common forms of non-malignant cardiac tumors, other cellular types have been encountered as intra-cardiac masses. Most are considered extremely rare and limited to cases

reports or small series from large institutions. These masses include teratomas, hamartomas, fibromas, hemangiomas, paragangliomas, and mesotheliomas of the atrioventricular node [50, 51]. Management is often similar to other cardiac masses and based on presenting symptoms, concern for embolic risk or obstructive physiology, and clinical risk of surgical intervention [52]. Because of the rarity of these types of tumors, little is known about pre-operative incidence. A definitive tissue diagnosis is often made at the time of surgical resection [53]. Unlike myxomas and fibroelastomas, embolic complications of these unusual tumors are rare. Patients typically present with obstructive heart failure symptoms. As most are asymptomatic, they are often only encountered at autopsy and are rarely considered the cause of death [54]. Resection or debulking is typically indicated based on presentation, tumor location (especially with regard to surrounding cardiac structures), and size. However, some patients with large tumors, presumed to be benign or potentially curative with resection, might need cardiac transplantation if safe resection is not possible [55]. Most ventricular fibromas can undergo safe resection, even if the tumor involvement is extensive, with good short- and long-term results and trivial risk of recurrence [56]. In general, symptoms of heart failure and arrhythmias resolve with resection. Asymptomatic tumors that fit into this category can be managed expectantly with serial imaging.

7. Surgical management of benign cardiac tumors

The surgical management of non-malignant cardiac tumors varies depending on the location of the tumor and the potential involvement of intra-cardiac structures [57]. Not only is the oncologic principle of wide excisional margins is not always possible due to the critical nature of certain cardiac structures, it is not necessary. The key principle is complex tumor excision if possible. Unless the diagnosis is obvious based on presentation, location, and clinical appearance, non-malignant tumors can often be confused with subacute endocarditis or intra-cardiac thrombus. Nevertheless, complex excision is necessary. Fibroelastomas can often be the easiest to remove, typically by sharp excision from their adherent structures. Even with valvular involvement, it would be uncommon to require leaflet reconstruction or valve replacement.

Intra-cardiac excisions, almost by definition, require cardiopulmonary bypass, aortic cross-clamping, and cardiac arrest. The specific techniques are beyond the scope of this chapter, as are the advantages and disadvantages of choice of incision (i.e., conventional full sternotomy, minimally-invasive sternal or right thoracotomy, or robotic-assisted techniques) and myocardial protection. The approach is probably best left to the individual comfort level and skill of the surgeon [58]. Nevertheless, because focused resection compared to wide debridement is typically the surgical goal, minimally-invasive approaches may be reasonable in appropriately selected patients. Cannulation techniques will vary depending on the tumor location. However, there should be a low threshold for bi-caval cannulation to assist in access to the atrial chambers and possible resection and reconstruction of the intra-atrial septum. In addition, root venting should always be considered, even with primary right-sided structures, as communication or left-sided involvement might introduce intra-cardiac air that would require appropriate de-airing upon weaning from cardiopulmonary bypass. In patients with concomitant cardiovascular pathology (i.e., coronary artery disease, aortic aneurysms, or separate valvular pathology), appropriate surgical intervention should be performed. The decision to perform a preoperative cardiac

catheterization should be based on an appropriate multi-disciplinary assessment of the risks of underlying coronary artery disease, risks of the procedures (i.e., catheter-induced dislodgement of the tumor), and the age and co-morbidities of the patient as suggested by clinical guidelines [59]. In situations in which cardiac catheterization is either relatively contraindicated (i.e., presence of an aortic valve mass) or perceived to be of low clinical benefit, coronary computed tomography might be considered and potentially helpful in guiding therapy [60]. Intra-operative transesophageal echocardiography should be used routinely to ensure an accurate diagnosis, clear identification of involved structures, and complete resection prior to surgical closure [61].

For tumors that involve either the aortic valve or left ventricular outflow track, the surgical approach should be trans-aortic—essentially a similar approach as is used for traditional aortic valve replacement surgery. Aortic valve replacement is rarely necessary. Careful trans-aortic exposure to the left ventricular outflow track can provide access to tumors in the LVOT or on the left ventricular side of the anterior leaflet of the mitral valve. Even residual aortic or mitral insufficiency, either primary insufficiency or as a consequence of resection, can be well tolerated for many years. The indications for valve replacement in such cases should be limited to those patients in whom residual regurgitation would otherwise require repair or replacement based upon current guidelines for valvular dysfunction management [62].

For tumors that involve an isolated valve, a standard surgical approach to the specific valve is typically employed based on surgeon preference, i.e., trans-right atrial for tricuspid pathology and right ventricular masses. For masses such as myxomas that involve the intra-atrial septum, a variety of approaches can be used. The most common approach is through the right atrium (even if the tumor is on the left atrial side of the septum) with excision of the fossa ovalis or the involved intra-atrial septum to remove not only the tumor, but the stalk and the potential “tumor roots” in the septal tissue (**Figure 9**). Even with large tumors, primary reconstruction of the intra-atrial septum can be often performed as the size (width and length) of the associated stalk rarely correlates with the actual extent of septal tissue involvement. For large septal defects, reconstruction with bovine pericardium can easily be performed [16]. Depending on surgeon experience, preference, and tumor location, a right thoracotomy approach to either the left or right atrium can be considered [63]. While a left atrial approach has been described for tumors on the left atrial side of the intra-atrial septum, if such an approach is chosen, it is critical that the principles of complex tumor excision, including the stalk and septal roots (with reconstruction of the intra-atrial septum if necessary), be maintained [64]. Merely shaving the tumor off the intra-atrial septum without resecting the septal tissue is inappropriate as it might leave residual tumor behind and predispose to tumor recurrence.

All surgical excisions should be sent for pathology and microbiology. A comprehensive pathological evaluation is critical as many tumors have extensive thrombotic material covering them that might confound a true diagnosis. Occasionally, tumors can become infected with presentations similar to endocarditis. When encountered, a prolonged course of targeted antibiotic therapy is recommended, as with any form of endocarditis. Likewise, microbiologic assessment of the mass is necessary to rule out a potentially infectious etiology, especially in the absence of a clear preoperative diagnosis. Concomitant infected tumors, while part of any differential diagnosis, are rare [65].

Post-operative management should be consistent with that of any other post-cardiotomy patient. Anti-coagulation should only be considered if indicated for other reasons, such as if recommended by neurologic consultants for the



Figure 9.
Surgical specimen of a left atrial myxoma. Approximately 3 cm left atrial cardiac myxoma surgical specimen. Note the resected intra-atrial septal tissue.

treatment of embolic strokes. For patients whose tumors appear to have a large thrombus burden, a hypercoagulable state work-up and appropriate treatment should be considered. Guidelines for post-resection imaging surveillance are lacking and should be symptom-based unless there is concern for incomplete resection or recurrence.

8. Summary

Intra-cardiac masses represent a challenging clinical problem. Patients often present with embolic complications or obstructive heart failure symptoms. Alternatively, they may be asymptomatic with the mass discovered as an incidental finding in work-up of an alternative diagnosis or in preparation for other therapies (i.e., coronary artery bypass surgery). As discussed, such tumors are rare and must be distinguished from other cardiac masses, specifically endocarditis and intra-cardiac thrombus for which the management strategies are well-established. The overriding principle of management is prevention of potentially catastrophic embolic complications, specifically neurologic events. However, the data to support this approach is either limited or not based on high-quality randomized or controlled trials [3, 29]. As such, when encountered in appropriately risk-stratified patients, surgical removal is often curative and should be considered first-line therapy. While STS risk scoring is often used to evaluate these patients, a specific risk-model for treatment is not part of the STS calculator. However, it most closely matches the risks for patients undergoing valve repair or replacement (<http://riskcalc.sts.org/stswebriskcalc/calculate>), Risk for recurrence is low and post-operative survival is excellent. While medical management of these masses is considered in high-risk patients and those who refuse surgery, it is important to consider there is little data to support this approach and some evidence to suggest an increased stroke risk. Medical management, specifically anti-thrombotic therapies, have little role and can potentially delay a diagnosis until a catastrophic neurologic event occurs. It should only be considered in unusual cases. Furthermore, while the mere presence of an intra-cardiac mass is considered an indication for surgical resection, if there are concerns about the diagnosis based on echocardiographic characteristics or if there is concern for an invasive primary or metastatic malignant tumor, CT or MRI imaging can be useful. Given the rarity of these tumors, if there are any concerns about the diagnosis or management, referral to a tertiary care center should be considered.

9. Conclusions

Intra-cardiac masses are rare, but are occasionally found during work-up for a source of embolism or encountered as an incidental finding. Tumor location and echocardiographic characteristics often suggest a diagnosis. However, definitive surgical resection for both diagnostic and therapeutic reasons should be considered first-line therapy. Patients managed non-operatively have increased risk for embolic complications. Medical therapies have not been shown to be effective although definitive data is lacking and controlled trials may be difficult to perform.

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References

- [1] Benjamin EJ, Virani SS, Callaway CW, Chamberlain AM, Chang AR, Cheng S, et al. Heart disease and stroke statistics—2018 update: A report from the American Heart Association. *Circulation* 2018;137(12):e67-492
- [2] Khurram IM, Dewire J, Mager M, Maqbool F, Zimmerman SL, Zipunnikov V, et al. Relationship between left atrial appendage morphology and stroke in patients with atrial fibrillation. *Heart Rhythm*. 2013;10(12):1843-1849
- [3] Holmes DR, Doshi SK, Kar S, Price MJ, Sanchez JM, Sievert H, et al. Left atrial appendage closure as an alternative to warfarin for stroke prevention in atrial fibrillation: A patient-level meta-analysis. *Journal of the American College of Cardiology*. 2015;65(24):2614-2623
- [4] Firstenberg MS. Introductory chapter: Introduction to advanced concepts in endocarditis. In: Firstenberg MS, editor. *Advanced Concepts in Endocarditis*. London: IntechOpen; 2018. p. 79883. DOI: 10.5772/intechopen. Available from: <https://www.intechopen.com/books/advanced-concepts-in-endocarditis/introductory-chapter-introduction-to-advanced-concepts-in-endocarditis>
- [5] Butany J, Nair V, Naseemuddin A, Nair GM, Catton C, Yau T. Cardiac tumours: Diagnosis and management. *The Lancet Oncology*. 2005;6(4):219-228
- [6] Wold LE, Lie JT. Cardiac myxomas: A clinicopathologic profile. *The American Journal of Pathology*. 1980;101:219-240
- [7] Burke AP, Virmani R. Tumors of the Heart and Great Vessels. *Atlas of Tumor Pathology*. Washington, DC: Armed Forces Institute of Pathology; 1996. pp. 1-11
- [8] Burke A, Tavora F. The 2015 WHO classification of tumors of the heart and pericardium. *Journal of Thoracic Oncology*. 2016;11(4):441-452
- [9] Wei K, Guo HW, Fan SY, Sun XG, Hu SS. Clinical features and surgical results of cardiac myxoma in carney complex. *Journal of Cardiac Surgery*. 2019;34:14-19
- [10] Cao H et al. *Journal of Biomedical Research*. 2011;25(5):368-372
- [11] Seino Y, Ikeda U, Shimada K. Increased expression of interleukin 6 mRNA in cardiac myxomas. *British Heart Journal*. 1993;69(6):565-567. DOI: 10.1136/hrt.69.6.565. PMC 1025174. PMID 8343326
- [12] Knepper LE, Biller J, Adams HP, Bruno A. Neurologic manifestations of atrial myxoma. A 12-year experience and review. *Stroke*. 1988;19(11):1435-1440. DOI: 10.1161/01.str.19.11.1435. PMID 3188128
- [13] Burke AP, Tazellar H, Gomez-Roman J, et al. WHO Classification of Tumours. Pathology and Genetics of Tumors of the Lung, Pleura, Thymus and Heart. Lyon: IARC Press; 2004. pp. 260-265
- [14] Wang JG, Li YJ, Liu H, et al. Clinicopathologic analysis of cardiac myxomas: Seven years' experience with 61 patients. *Journal of Thoracic Disease*. 2012;4(3):272-283
- [15] Araoz PA, Mulvagh SL, Tazelaar HD, Julsrud PR, Breen JF. CT and MR imaging of benign primary cardiac neoplasms with echocardiographic correlation. *Radiographics*. 2000;20(5):1303-1319
- [16] Lee KS, Kim GS, Jung Y, Jeong IS, Na KJ, Oh BS, et al. Surgical resection of cardiac myxoma—A 30-year single institutional experience. *Journal of Cardiothoracic Surgery*. 2017;12(1):18

- [17] Animashaun IB, Akinseye OA, Akinseye LI, Akinboboye OO. Right atrial myxoma and syncope. *The American Journal of Case Reports*. 2015;**16**:645-647
- [18] Kassi M, Polsani V, Schutt RC, Wong S, Nabi F, Reardon MJ, et al. Differentiating benign from malignant cardiac tumors with cardiac magnetic resonance imaging. *The Journal of Thoracic and Cardiovascular Surgery*. 2019;**157**(5):1912-1922
- [19] McCarthy CP, Vaduganathan M, McCarthy KJ, Januzzi JL, Bhatt DL, McEvoy JW. Left ventricular thrombus after acute myocardial infarction: Screening, prevention, and treatment. *JAMA Cardiology*. 2018;**3**(7):642-649
- [20] Di Minno MN, Ambrosino P, Russo AD, Casella M, Tremoli E, Tondo C. Prevalence of left atrial thrombus in patients with non-valvular atrial fibrillation. *Thrombosis and Haemostasis*. 2016;**115**(03):663-677
- [21] Acebo E, Val-Bernal JF, Gomez-Roman JJ. Thrombomodulin, calretinin and c-kit (CD117) expression in cardiac myxoma. *Histology and Histopathology*. 2001;**16**(4):1031-1036
- [22] Maleszewski JJ, Larsen BT, Kip NS, et al. PRKAR1A in the development of cardiac myxoma: A study of 110 cases including isolated and syndromic tumors. *The American Journal of Surgical Pathology*. 2014;**38**(8):1079-1087
- [23] Wang Z, Chen S, Zhu M, et al. Risk prediction for emboli and recurrence of primary cardiac myxomas after resection. *Journal of Cardiothoracic Surgery*. 2016;**11**(22):1-8
- [24] Reynen K. Cardiac myxomas. *The New England Journal of Medicine*. 1995;**333**(24):1610-1617
- [25] Shinfeld A, Katsumata T, Westaby S. Recurrent cardiac myxoma: Seeding or multifocal disease? *The Annals of Thoracic Surgery*. 1998;**66**(1):285-288
- [26] Fleischmann KE, Schiller NB. Papillary fibroelastoma: Move over myxoma. *Journal of the American College of Cardiology*. 2015;**65**(22):2430-2432
- [27] Bicer M, Cikirikcioglu M, Pektok E, et al. Papillary fibroelastoma of the left atrial wall: A case report. *Journal of Cardiothoracic Surgery*. 2009;**4**(28):1-4
- [28] Kurup AN, Tazelaar HD, Edwards WD, et al. Iatrogenic cardiac papillary fibroelastoma: A study of 12 cases (1990 to 2000). *Human Pathology*. 2002;**33**(12):1165-1169
- [29] Remadi JP, Degandt A, Rakotoarivello Z. Cardiac papillary fibroelastoma of the mitral valve chordae. *Heart*. 2004;**90**:12
- [30] Murphy ET. Robotic excision of aortic valve papillary fibroelastoma and concomitant maze procedure. *Global Cardiology Science and Practice*. 1 Nov 2013;**2012**(2):93-100. DOI: 10.5339/gcsp.2012.27. PubMed PMID: 24688994; PubMed Central PMCID: PMC3963717
- [31] Grinda JM, Couetil JP, Chauvaud S, et al. Cardiac valve papillary fibroelastoma: Surgical excision for revealed or potential embolization. *The Journal of Thoracic and Cardiovascular Surgery*. 1999;**117**:106-110
- [32] Klarich KW, Enriquez-Sarano M, Gura GM, et al. Papillary fibroelastoma: Echocardiographic characteristics for diagnosis and pathologic correlation. *Journal of the American College of Cardiology*. 1997;**30**(3):784-790
- [33] Kamran H, Patel N, Singh G. Lambl's excrescences: A case report and review of the literature. *Clinical Case Reports and Reviews*. 2016;**2**(7):476-478

- [34] Marstrand P, Jensen MB, Ihlemann N. Valvular excrescences: A possible transient phenomenon. *Case Reports in Cardiology*. 2015;**2015**:380765. DOI: 10.1155/2015/380765. Epub 2015 Nov 16
- [35] Mkalaluh S, Szczechowicz M, Torabi S, Dib B, Sabashnikov A, Mashhour A, et al. Surgery for cardiac papillary fibroelastoma: A 12-year single institution experience. *Medical Science Monitor Basic Research*. 2017;**23**:258-263
- [36] Shelh M. Multiplane transesophageal echocardiography detection of papillary fibroelastomas of the aortic valve causing a stroke. *European Heart Journal*. 1997;**18**(4):702-703
- [37] Tamin SS, Maleszewski JJ, Scot CG, et al. Prognostic and bioepidemiologic implications of papillary fibroelastoma. *Journal of the American College of Cardiology*. 2015;**65**(22):2420-2429
- [38] Sun JP, Asher CR, Yang S, et al. Clinical and echocardiographic characteristics of papillary fibroelastomas. A retrospective and prospective study in 162 patients. *Circulation*. 2001;**103**:2687-2693
- [39] Abu Saleh WK, Al Jabbari O, Ramlawi B, Reardon MJ. Cardiac papillary fibroelastoma: Single-institution experience with 14 surgical patients. *Texas Heart Institute Journal*. 2016;**43**(2):148-151
- [40] Mkalaluh S, Szczechowicz M, Torabi S, Dib B, Sabashnikov A, Mashhour A, et al. Surgery for cardiac papillary fibroelastoma: A 12-year single institution experience. *Medical Science Monitor Basic Research*. 2017;**23**:258
- [41] Gowda RM, Khan IA, Nair CK, Mehta NJ, Vasavada BC, Sacchi TJ. Cardiac papillary fibroelastoma: A comprehensive analysis of 725 cases. *American Heart Journal*. 2003;**146**(3):404-410
- [42] Klarich KW, Enriquez-Sarano M, Gura GM, Edwards WD, Tajik AJ, Seward JB. Papillary fibroelastoma: Echocardiographic characteristics for diagnosis and pathologic correlation. *Journal of the American College of Cardiology*. 1997;**30**(3):784-790
- [43] Sabet A, Haghighiabyaneh M, Tazelaar H, Raisinghani A, DeMaria A. The clinical dilemma of cardiac fibroelastic papilloma. *Structural Heart*. 2018;**2**(4):274-280
- [44] Li D, Wang W, Zhu Z, Wang Y, Xu R, Liu K. Cardiac lipoma in the interventricular septum: A case report. *Journal of Cardiothoracic Surgery*. Dec 2015;**10**(1):69
- [45] Naseerullah FS, Javaiya H, Murthy A. Cardiac lipoma: An uncharacteristically large intra-atrial mass causing symptoms. *Case Reports in Cardiology*. 2018;**2018**:3531982
- [46] Laeeq R, Merchant O, Khalid UM, Lakkis NM. Large cardiac lipoma in a patient presenting with atrial tachycardia and systolic heart failure. *Journal of Cardiac Failure*. 2017;**23**(8):S94
- [47] Wijesurendra RS, Sheppard KA, Westaby S, Ormerod O, Myerson SG. The many faces of cardiac lipoma—An egg in the heart! *European Heart Journal Cardiovascular Imaging*. 2017;**18**(7):821
- [48] Kim YS, Lee KH, Choi SJ, Baek WK. Cardiac lipoma arising from left ventricular papillary muscle: Resect or not? *The Journal of Thoracic and Cardiovascular Surgery*. 2018;**156**(1):244-246
- [49] Dickerson JA, Smith M, Kalbfleisch S, Firstenberg MS. Lipomatous hypertrophy of the intraatrial septum resulting in right atrial inflow obstruction and atrial flutter. *The Annals of Thoracic Surgery*. 2010;**89**(5):1647-1649

- [50] Shah DJ, Reardon MJ. Case of a hamartoma or hamartoma of a case? The Journal of Thoracic and Cardiovascular Surgery. 2018;**155**(1):351-352
- [51] El-Ashry AA, Cerfolio RJ, Singh SP, McGiffin D. Cardiac paraganglioma. Journal of Cardiac Surgery. 2015;**30**(2):135-139
- [52] Tudor AA, Tschui J, Schmidli J, Schmid RA, Dorn P. Cardiac paraganglioma—A rare subset of a rare tumor. World Journal of Cardiovascular Diseases. 2017;**7**:1-9
- [53] Thompson KA. Managing benign cardiac tumors. MD Anderson Practices. Onco-Cardiology. 2016;**30**:30-35
- [54] Sarjeant JM, Butany J, Cusimano RJ. Cancer of the heart: Epidemiology and management of primary neoplasms and metastases. American Journal of Cardiovascular Drugs. 2003;**3**:407-421
- [55] Valente M, Cocco P, Thiene G, Casula R, Poletti A, Milanese O, et al. Cardiac fibroma and heart transplantation. The Journal of Thoracic and Cardiovascular Surgery. 1993;**106**(6):1208-1212
- [56] Cho JM, Danielson GK, Puga FJ, Dearani JA, McGregor CG, Tazelaar HD, et al. Surgical resection of ventricular cardiac fibromas: Early and late results. The Annals of Thoracic Surgery. 2003;**76**(6):1929-1934
- [57] Yanagawa B, Mazine A, Chan EY, Barker CM, Gritti M, Reul RM, et al. Surgery for tumors of the heart. Seminars in Thoracic and Cardiovascular Surgery. 2018;**30**(4):385-397
- [58] Luo C, Zhu J, Bao C, Ding F, Mei J. Minimally invasive and conventional surgical treatment of primary benign cardiac tumors. Journal of Cardiothoracic Surgery. 2019;**14**(1):76
- [59] Baumgartner H, Falk V, Bax JJ, De Bonis M, Hamm C, Holm PJ, et al. ESC/EACTS guidelines for the management of valvular heart disease. European Heart Journal. 2017;**38**(36):2739-2791
- [60] Opolski MP, Staruch AD, Jakubczyk M, Min JK, Gransar H, Staruch M, et al. CT angiography for the detection of coronary artery stenoses in patients referred for cardiac valve surgery: Systematic review and meta-analysis. JACC: Cardiovascular Imaging. 2016;**9**(9):1059-1070
- [61] Couture P, Denault AY, McKenty S, Boudreault D, Plante F, Perron R, et al. Impact of routine use of intraoperative transesophageal echocardiography during cardiac surgery. Canadian Journal of Anesthesia. 2000;**47**(1):20-26
- [62] Nishimura RA, Otto CM, Bonow RO, Carabello BA, Erwin JP, Fleisher LA, et al. AHA/ACC focused update of the 2014 AHA/ACC guideline for the management of patients with valvular heart disease: A report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. Journal of the American College of Cardiology. 2017;**70**(2):252-289
- [63] Ellouze M, Pellerin M, Jeanmart H, Lebon JS, Bouchard D. Mini right anterior thoracotomy approach versus sternotomy for resection of intracardiac myxoma. Innovations: Technology and Techniques in Cardiothoracic and Vascular Surgery. 2018;**13**(4):292-295
- [64] Jain S, Maleszewski JJ, Stephenson CR, Klarich KW. Current diagnosis and management of cardiac myxomas. Expert Review of Cardiovascular Therapy. 2015;**13**(4):369-375
- [65] Yuan SM. Infected cardiac myxoma: An updated review. Brazilian Journal of Cardiovascular Surgery. 2015;**30**(5):571-578