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CASE REPORT

CLINICAL CASE

Novel Use of a Catheter-Based Device Retrieval System for Percutaneous Treatment of Recurrent Atrial Myxoma

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ABSTRACT

Surgical resection is standard of care for the treatment of atrial myxoma. However, the optimal management strategy for recurrent cardiac tumors is less clear. Here we report the novel use of a catheter-based device retrieval system for the removal of a recurrent cardiac myxoma. (JACC Case Rep. 2024;■:102737) © 2024 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

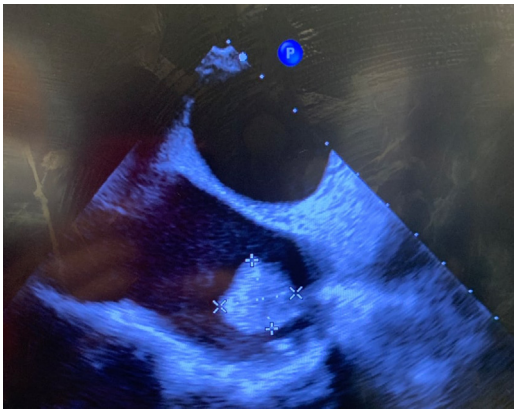
A 51-year-old man with no significant past medical history originally presented with left lower extremity pain. His vital signs were stable, and the physical examination was significant for no murmur on auscultation. He was found to have a superficial thrombus of the left midcalf. Given concern for endocarditis, he underwent transthoracic echocardiography (TTE), which showed a left atrial mass that measured 4.0 cm x 3.0 cm. The mass originated from the interatrial septum near the fossa ovalis, with diastolic imaging showing that the mass impinged on the anterior mitral leaflet with partial obstruction of left ventricular filling (Transesophageal echo **Figures 1 and 2**).

TAKE-HOME MESSAGES

- This case demonstrates the viability of a catheter-based approach for the minimally invasive management of recurrent cardiac myxoma.

The patient subsequently underwent surgical resection of the left atrial mass with septal patch closure using bovine pericardium and had an unremarkable postoperative course. Pathologic findings were consistent with atrial myxoma.

FIGURE 1 Transesophageal Echocardiogram Visualizing the Right Atrial Mass



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ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

CNC = Carney complex

CT = computed tomography

TEE = transesophageal echocardiography

TTE = transthoracic echocardiography

Approximately 6 years after surgical resection, the patient presented with peripheral emboli to his upper extremities and underwent TTE, showing a right atrial mass. Cardiac magnetic resonance (CMR) showed that the mass was attached to the posterior wall of the superior right atrium (Figures 3 and 4).

PAST MEDICAL HISTORY

The patient had no significant past medical history.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included endocarditis, a recurrent cardiac mass, and hypercoagulable disorder.

INVESTIGATIONS

There was concern regarding repeat cardiac surgery given the need for repeat sternotomy and an unquantifiable risk of another recurrence. Therefore, alternative minimally invasive options were investigated. Ultimately, after evaluation by an interventional cardiologist who specializes in adult congenital cardiac disease, it was determined to pursue minimally invasive excision of the right atrial intracardiac mass by using a novel catheter-based device retrieval system (ONO, ONOCOR Vascular).

MANAGEMENT

Minimally invasive catheter-based removal of the right atrial cardiac mass was successful, with the same-day pathology report confirming recurrence of myxoma (Figures 5 and 6). The procedure was well tolerated, and the patient went home on the same day.

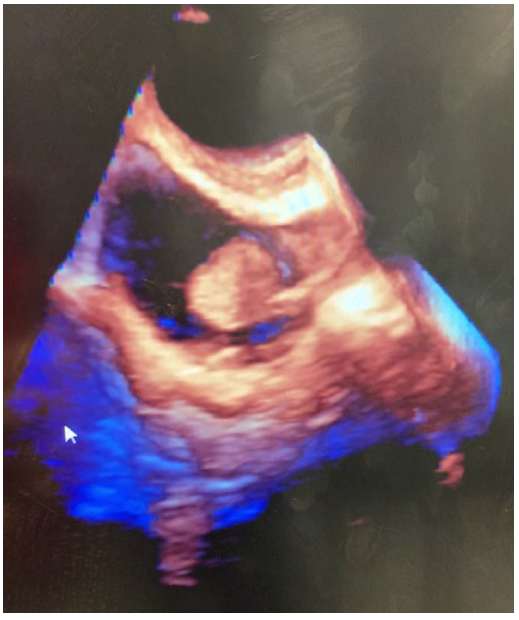
DISCUSSION

Cardiac myxomas are most commonly found in the left atrium. However, myxomas can also be found in the right atrium and, even less frequently, in the left ventricle.[1] Recurrence of myxoma has been observed in 3% of patients. A literature review that included 42 cases of left atrial myxoma reported that the most frequent site of recurrence was the left atrium (83%), with the next most frequent being the right atrium (14%).[2] The timing for recurrence is usually within 10 years from the initial diagnosis, although cases have been reported to recur as early as 10 months after the initial diagnosis.[3]

There are several theories to explain cardiac myxoma recurrence, including the following: 1) inadequate surgical removal; 2) totipotent multicentricity (multifocal disease); and 3) genetic predisposition.

Inadequate surgical resection is a known concern and is typically addressed with surgical approaches that use wide resection fields, often incorporating prophylactic resection of the atrial septum with

FIGURE 2 3-Dimensional Imaging of the Right Atrial Mass



The arrow indicates the right atrial mass.

FIGURE 3 Axial View Cardiac Magnetic Resonance Visualizing the Right Atrial Mass

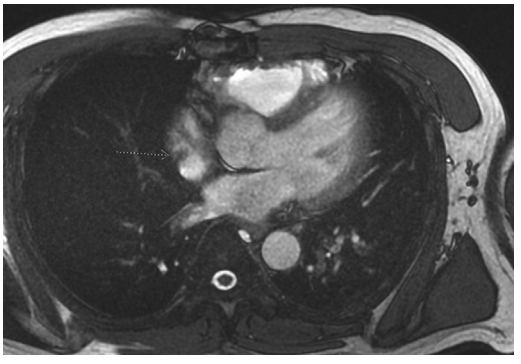


FIGURE 4 Coronal View Cardiac Magnetic Resonance Visualizing the Right Atrial Mass

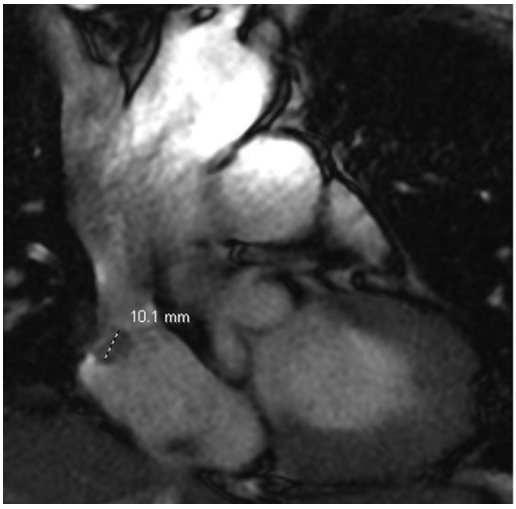
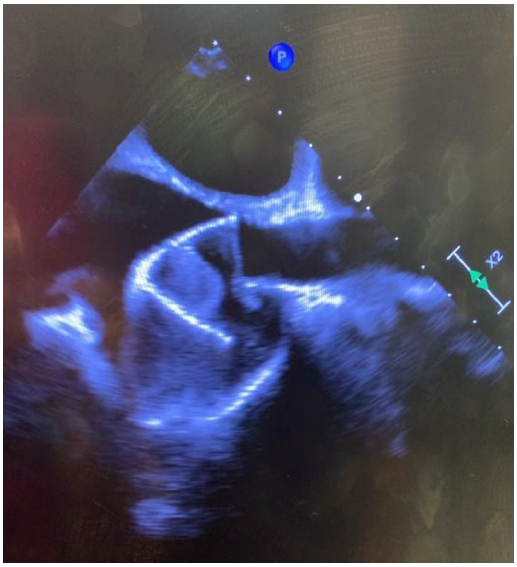


FIGURE 5 Transesophageal Echocardiogram Visualizing the Snare Device Capturing the Right Atrial Mass



subsequent patching.[2] Inadequate resection is believed to be a likely cause when recurrence is at or near the same site of the original primary tumor.

Recurrence at a site different from the original primary tumor site suggests that multifocal disease may be present, a theory termed totipotent multicentricity. This theory is supported by the presence of recurrence in new sites despite wide surgical resection and patching of the atrial septum.[2]

Syndromic myxoma resulting from genetic disease makes up approximately 10% of all myxomas.[1] The most clearly characterized genetic condition associated with cardiac myxoma is Carney complex (CNC). CNC is diagnosed by the presence of an endocrine tumor, spotty pigmentation, and a mesenchymal tumor (ie, myxoma) in the heart or skin.[1] CNC is inherited in an autosomal dominant pattern, with sequence variants in the *PRKAR1A* gene causing most cases.[1]

Cardiac myxoma can be diagnosed using echocardiography, computed tomography (CT), or CMR. Clinical examination findings such as a systolic or diastolic murmur may be present if the myxoma causes atrioventricular valve obstruction or regurgitation.[4] TTE is the most commonly used tool for diagnosing cardiac tumors and has a diagnostic accuracy of 90%, whereas transesophageal echocardiography (TEE) has an accuracy of 96%. TEE is often used preprocedurally to characterize tumor size, attachment site, relationship with other cardiac structures, and hemodynamic effects. Chest CT or magnetic resonance imaging is often used to evaluate for coexisting conditions in the thorax and help with operative planning. In patients aged <40 years, preoperative CT coronary angiography is usually performed, whereas patients aged >40 years typically undergo invasive coronary angiography.[5]

FIGURE 6 Excised Right Atrial Myxoma

FIGURE 7 ONOCORE Snare (ONOCOR Vascular) Looping Around the Mass

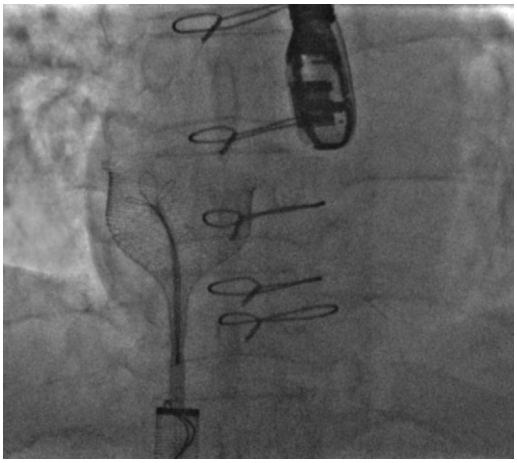
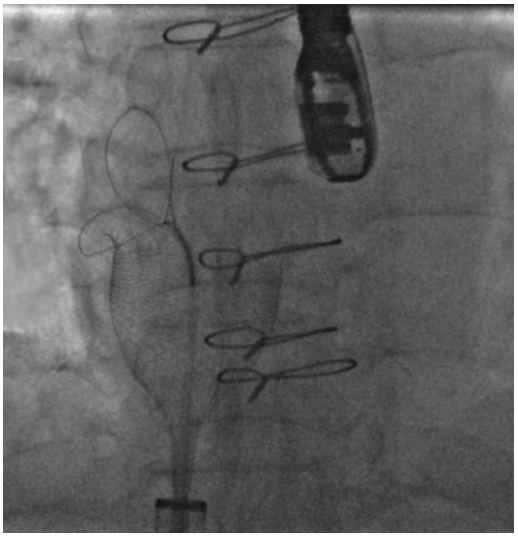


FIGURE 8 ONOCORE Snare (ONOCOR Vascular) Pulling the Mass Into the Basket



Surgical excision without delay is recommended as soon as the diagnosis is made. This recommendation is based on the risk of serious sequelae of myxoma such as heart valve obstruction—which can lead to sudden death—and the risk of embolization to coronary arteries or the systemic vasculature.

Cardiac surgery through a median sternotomy remains the most common approach for the management of atrial myxoma. Other techniques have also been reported, such as endoscopic robotic resection with percutaneous cannulation.[6]

Radical resection with wide margins is critical to prevent recurrence.[6] Specifically, full-thickness removal of the attachment base with patching of the interatrial septum to help prevent recurrence is used to increase the chances of having clear resection margins. Subendocardial resection is used only when full-thickness resection would lead to compromise of structural integrity of the heart.

There are limited data on the rates of recurrence specifically attributed to inadequate resection margins. A retrospective study that reported on the outcomes of 194 patients who underwent surgical resection for cardiac myxoma found no difference in recurrence whether endocardial resection was performed or full-thickness resection was performed.[7] It was found that when the primary myxoma was located in the ventricle, it was 7 times more likely to recur than an atrial myxoma. Additionally, smaller tumor size was associated with a higher likelihood of recurrence when compared with large tumor size. There was no clear rationale for the association of smaller size with higher rates of recurrence. Future microbiologic and molecular studies may help clarify this issue.

There is a low rate of postoperative complications following surgical resection, with the most common complication being atrial fibrillation.[5] Postoperative mortality rates at 30 days have been reported to range between 3% and 10%.[5] At 1 year after cardiac surgery, survival rates been reported to range between 85% and 96%.[5]

To date, the only management strategy reported for recurrent atrial myxoma is surgical excision.[6] However, repeat surgical excision involves repeat sternotomy and exposure to risks associated with cardiac surgery. Accordingly, a minimally invasive treatment option that circumvents risks associated with repeat cardiac surgery would be a highly favorable alternative.

The ONOCOR retrieval system enables precise removal of mispositioned structural heart devices percutaneously. It has been reported to be effective at retrieving erroneously placed devices such as MitraClips (Abbott).[8] Other applications have included intracardiac thrombectomy in high-risk pediatric patients.[9]

The ONOCOR retrieval system consists of a retrieval sheath, a loop snare catheter, and a 35-mm self-expanding nitinol basket. Typically, the retrieval sheath is placed by the operator in a location near the target to be removed to help facilitate positioning and alignment of the snare and basket. Once the retrieval sheath is in place, the snare catheter is

deployed to secure positioning around the target. Next, the basket is deployed to help further anchor the target. After the snare and basket are deployed around the target, the basket is collapsed and retrieved through the catheter for removal.

For this case, a retrieval sheath was loaded through a 12-F catheter and positioned in the right atrium by using venous access. Next, under direct visualization with fluoroscopy and TEE, the loop snare catheter was positioned around the stalk of the right atrial mass (**Figure 7**). Once the mass was entrapped with the snare catheter, the self-expanding basket was deployed to secure positioning (**Figure 8**). Subsequently, gentle tension was applied and delivered through the snare as the basket was crimped down and retrieved through the inner lumen of the retrieval catheter. Intraoperative TEE showed removal of the right atrial mass. Heparin was not used intraoperatively given that it was a right-sided mass. Although the loops forming the basket of the ONOCOR may theoretically help prevent debris from a mass from being embolized, known potential complications of using this device are embolization and stroke.

With consideration for the strong preference for investigating a viable, minimally invasive alternative to surgical excision for a recurrent cardiac mass, the ONOCOR retrieval system was used for percutaneous removal of the recurrent right atrial mass.

FOLLOW-UP

TTE at 3-month follow-up showed no signs of recurrence at the site of removal. Surveillance TTE is planned every 6 months for the first 2 years after the procedure.

CONCLUSIONS

Excluding this report, surgical resection has been the only reported management strategy for recurrent atrial myxoma.[6] To our knowledge, this is the first reported case of the use of a catheter-based device retrieval system to remove a recurrent atrial myxoma. Percutaneous removal of recurrent cardiac masses with a device retrieval system may be a viable therapeutic option that can be considered in the treatment algorithm for patients with these recurrent masses.

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REFERENCES

1. Imai Y, Taketani T, Maemura K, et al. Genetic analysis in a patient with recurrent cardiac myxoma and endocrinopathy. *Circulation*. 2005;69:994-995.

2. Shinfeld A, Katsumata T, Westaby S. Recurrent cardiac myxoma: seeding or multifocal disease. *Ann Thorac Surg*. 1998;66:285-288.

3. Zeybek R, Başaran Y, Öztop F. Recurrence of an atrial myxoma. *Int J Cardiol*. 1991;33:435-437.

4. Pucci A, Gagliardotto P, Zanini C, Pansini S, Di Summa M, Mollo F. Histopathologic and clinical characterization of cardiac myxoma: review of 53 cases from a single institution. *Am Heart J*. 2000;140:134-138.

5. Samanidis G, Khoury M, Balanika M, Perrea D. Current challenges in the diagnosis and treatment of cardiac myxoma. *Kardiol Pol*. 2020;78:269-277.

6. Rosen JL, Yost CC, Wong DH, et al. Routine endoscopic robotic cardiac tumor resection using an 8-mm working port and percutaneous cannulation. *J Card Surg*. 2022;37:4803-4807.

7. Shah IK, Dearani JA, Daly RC, et al. Cardiac myxomas: a 50-year experience with resection and analysis of risk factors for recurrence. *Ann Thorac Surg*. 2015;100(2):495-500.

8. Elison D, Aldea GS, Jelacic S, Chung C, Mackensen GB, McCabe JM. First-in-human percutaneous excision of a failed MitraClip followed by transcatheter mitral valve replacement. *JACC Cardiovasc Interv*. 2024;17(4):571-573.

9. Karunanandaa A, Patel ND, Wong PC, et al. Novel use of the ŌNŌ retrieval system for intracardiac thrombectomy in a high-risk pediatric patient. *J Soc Cardiovasc Angiogr Interv*. 2023;2:100976.

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