Removal of an infected pulmonary artery fibroelastoma disguised as a presentation of pulmonary embolism using a percutaneous suction thrombectomy device

Daemar H. Jones, MS,[a] Benjamin Lin, BA,[a] David Brabham, DO,[b] *and* Bradley Trinidad, MD,[c,d] *Lubbock and Amarillo, TX*

ABSTRACT

Papillary fibroelastoma (PFE) is a rare, often benign, tumor originating typically in the endocardium and valves, with a preference for the left side of the heart. Although PFEs can appear asymptomatic, in the setting of embolization, they can lead to stroke, acute limb ischemia, and/or mesenteric ischemia. Rarely, PFEs can originate from the pulmonary valve, with the potential for embolic showering into the pulmonary artery, leading to potential right-sided heart outflow obstruction. Treatment has been open surgery in most cases, although treatment of right-sided heart masses with extracorporeal circulatory support extraction systems have been described. Recently, large bore suction thrombectomy devices have become available, typically used for cases of venous thromboembolism. In the present report, we describe a case of a symptomatic infected PFE treated by percutaneous suction thrombectomy using the Inari FlowTriever system (Inari Medical). (J Vasc Surg Cases Innov Tech 2023;9:101346.)

Keywords: Fibroelastoma; Mechanical thrombectomy; Percutaneous endovascular technology

Papillary fibroelastoma (PFE) is a benign tumor and the second most common primary cardiac tumor in adults.[1] This cardiac tumor prefers the endocardium, with >80% localizing to heart valve leaflets, predominantly in the left side of the heart.[1,2] In the setting of a patent foramen ovale, this can lead to devastating embolic complications.[2] Tumor localized to the right side of the heart is uncommon. Advancement into the pulmonary arteries is even more rare and can mimic the signs common to pulmonary embolism (PE), including shortness of breath, hypoxia, and global right-sided heart outflow obstruction and right-sided heart failure. We present a case of an infected PFE in a patient presenting with an initial suspicion of PE. This patient was treated using the Inari FlowTriever device (Inari Medical) for tumor removal. The patient provided written informed consent for the report of her case details and imaging studies.

CASE REPORT

A 76-year-old woman with osteoporosis, hypothyroidism, and dyslipidemia presented to the emergency room with generalized weakness, recurrent syncope, and shortness of breath of 2 weeks' duration. The patient denied any history of clots, smoking, or surgery. On presentation, she was febrile, tachycardic, mildly hypertensive, and hypoxic with an oxygen saturation of 86% on room air and needing 5 L of oxygen to maintain saturation. The pulmonary and cardiovascular examination findings were unremarkable. Her laboratory findings were significant for leukocytosis and elevated brain natriuretic peptide and D-dimer levels. Blood cultures were performed due to concerns for infection, and computed tomography angiography (CTA; Fig 1) was performed to rule out PE. CTA of the chest confirmed an intraluminal filling defect in the pulmonary artery that was concerning for thrombus vs a mass. Therapeutic anticoagulation was initiated. On careful investigation of the images, contrast was seen infiltrating within the filling defect, an unusual finding for a PE. Transesophageal echocardiography (TEE) performed by our cardiology colleagues revealed right-sided heart strain with associated tricuspid regurgitation. It also revealed a 2.3-cm heterogenous mobile mass in the pulmonary artery without valve involvement. The findings were unequivocal, and the patient remained symptomatic. Vascular surgery was consulted, and the patient was scheduled for percutaneous transluminal mechanical thrombectomy with the goals of symptom resolution and resection of a tissue specimen for pathologic examination.

Percutaneous access of the right common femoral vein under ultrasound guidance was performed and a 9F short sheath placed, followed by an Advantage glidewire (Terumo Interventional Systems). A long-angled pigtail catheter was used to cannulate the right side of the heart. The wire was exchanged for a 0.035-in. Amplatz stiff wire (Boston Scientific), which was

From the Department of Surgery, Texas Tech University Health Sciences Center School of Medicine, Lubbock[a]; the Department of Cardiology, Cardiology Center of Amarillo, Amarillo[b]; the Department of Surgery, Texas Tech University Health Sciences Center School of Medicine, Amarillo[c]; and the Department of Vascular Surgery, Northwest Texas Hospital, Amarillo.[d]

Correspondence: Daemar Jones, MS, Texas Tech University Health Sciences Center School of Medicine, 3601 4th St, Lubbock, TX 79430 (e-mail: jones.daemar28@gmail.com).

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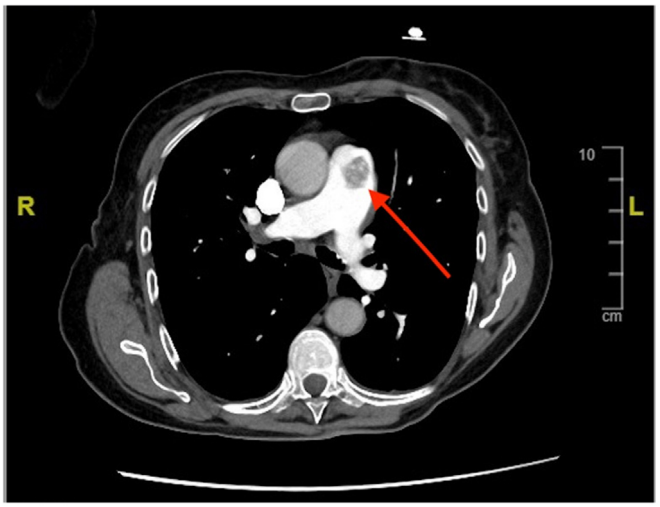

Fig 1. Computed tomography angiogram of the main pulmonary artery showing vegetation (*arrow*).

taken over the level of the left subsegmental pulmonary artery. Angiography revealed evidence of a mass in the main pulmonary artery (Fig 2, *a*). Intravascular ultrasound was used to confirm the finding. Next, the Amplatz wire was exchanged for a 24F long DrySeal sheath (W.L. Gore & Associates). The FlowTriever 20 (Inari Medical) was then advanced into the main pulmonary artery and suction thrombectomy performed. After one pass, the mass was removed in total (Fig 2, *b*). Filtered blood was not returned to the patient to reduce the risk of reentrance of the tumor burden into the circulatory system. Repeat intravascular ultrasound confirmed no further mass burden. The blood loss was 50 mL, the operative time was 1 hour, and the fluoroscopic time was 12 minutes. The vein access closure sites closed with 10 minutes of manual pressure. The specimen was sent to pathology for tumor identification (Fig 3).

Gross examination of the pulmonary artery mass showed a 2.0 × 1.7 × 0.4-mm tan, white, and irregular exophytic portion of soft tissue. Microscopic examination included an immunohistochemical stain for CD34, which demonstrated endothelial cells lining papillary fronds of the tumor, and elastin positively stained the acellular papillary cores. The findings were consistent with PFE. Tissue pathology cultures were sent and were positive for *Haemophilus parainfluenzae*. The infection was well controlled with ceftriaxone. Blood cultures were also taken at this time, which were positive for methicillin-sensitive *Staphylococcus aureus*. For this, she received a nafcillin infusion. She continued the ceftriaxone and nafcillin for 4 weeks after the last negative culture. Postoperative transthoracic echocardiography (TTE) showed mild pulmonary hypertension, with a right ventricular (RV) systolic pressure (RVSP) of 40 to 45 mm Hg, an improvement from her preoperative report (RVSP, 80-85 mm Hg). Her luminal filling defect improved after mass removal, but anticoagulation was recommended for 3 months, with follow-up TEE to rule out any residual tissue in the pulmonary artery.

DISCUSSION

PFE can present asymptomatically in many patients and might never be symptomatic. Approximately 30% are diagnosed incidentally on echocardiography or autopsy.[3] PFE can also be quite symptomatic, whether from pulmonary outflow obstruction or in the setting of a patent foramen ovale with dangerous embolic showering. Embolization can progress into the systemic circulation. This can cause stroke or mesenteric ischemia. Cardiac tumors of the right side of the heart are typically asymptomatic.[4] Problems arise depending on the size and location. Embolization into the pulmonary circulation can lead to PE, manifesting as hypoxemia and pulmonary hypertension.

The workup for PFE usually includes echocardiography and, in the setting of symptoms, often CTA. The features of PFE typically include a smaller size, a highly mobile appearing mass with a peduncle or stalk attached to the valve, and a frond-like appearance.[5] CTA allows for accurate and quick visualization for patients who might require emergent or urgent surgical treatment. The present patient had symptoms concerning for PE. CTA revealed the intraluminal filling defect vs thrombus. The mass was noted to have contrast infiltrate into the filling defect, which would be unusual for PE.

After the presumptive diagnosis of PFE, surgery is recommended for all patients because of the embolization risk and associated morbidity.[4] Complications related to tumor mobility result in the risk of stroke, PE, pulmonary hypertension, congestive heart failure, myocardial infarction, and mesenteric and limb ischemia.[6,7] Patients with suspected tumor mobility and increased size have greatly improved morbidity after surgery. The literature shows a surgical approach using tumor resection with possible valve replacement. Others require open surgery and bypass to remove the PFE.[8] More minimally invasive approaches to right-sided heart vegetations have been described using the Angiovac system (Angiodynamics). One report describes its use specifically for the of a PFE.[7] The Angiovac system requires at least dual access from the jugular and femoral veins, a 26F sheath, and initial extracorporeal circulation of right-sided heart bypass. Blood loss has also been an issue in the past with this device. The Inari FlowTriever (Inari Medical) was used in the present case. The Inari FlowTriever system is a percutaneous mechanical thrombectomy device and was the first Food and Drug Administration-approved device for the treatment of PE.[9] It has been shown to be more effective at reducing RV/left ventricular dilatation compared with anticoagulation therapy.[9,10] It can be used through a single venous access, with a smaller sheath, and allows for the option to return or not return filtrated blood. This was important for our patient, because we did not want to risk recirculating tumor cells into the bloodstream. The Inari

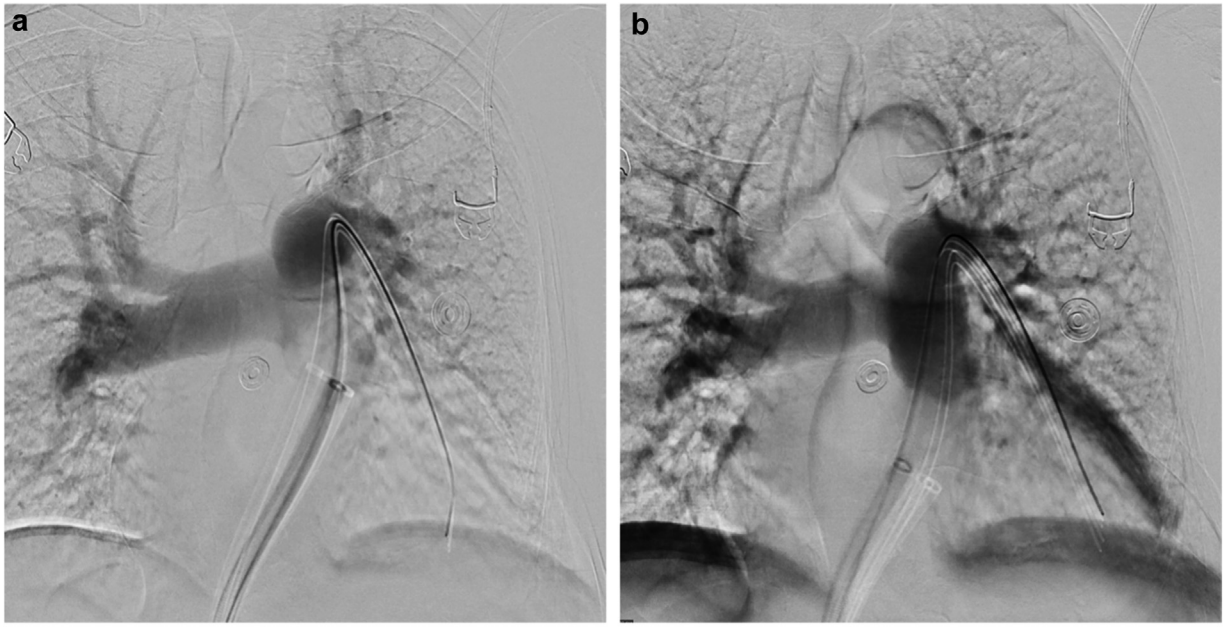

Fig 2. a, Digital subtraction angiography pulmonary arteriography showing a mass defect in the main pulmonary artery restricting blood to the lungs. **b,** Digital subtraction angiography pulmonary arteriography showing gross resolution of the main pulmonary artery that was free of the foreign mass.

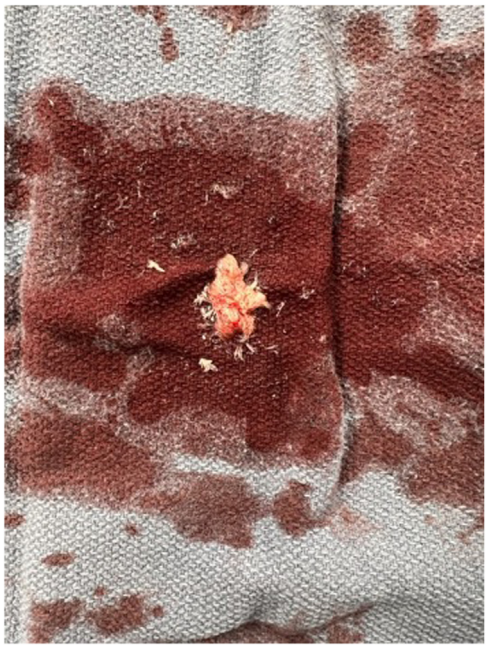

Fig 3. Photograph of the foreign mass removed by suction thrombectomy.

FlowTriever for thrombectomy has been shown to have minimal outcomes in distal embolization. Compared with anticoagulation alone to treat PE, the FlowTriever has been shown to result in less clinical deterioration.[11] Bleeding remains the top concern with using this device. There is a risk of systemic embolization by not conducting surgery. If concerned for distal embolization, one should monitor the patient's symptoms and hospital progress to determine the next plan of action in their treatment. In the present case, the luminal filling defect improved after mass removal. She did not have symptoms concerning for stroke, acute myocardial infarction, or acute limb ischemia throughout her hospital stay. She was free of severe symptoms and was discharged when she was ready. The preoperative TTE showed an RVSP of 80 to 85 mm Hg. The postoperative TTE showed an RVSP of 40 to 45 mm Hg with relief of symptoms. The patient received postoperative anticoagulation therapy for 3 months with frequent follow-up visits. At 3 months after discharge, she was scheduled for TEE, which did not show any remnant tumor. She denied all symptoms concerning for distal embolization. It was recommended that she undergo TEE every 3 months for follow-up. Other reports have shown that cardiac magnetic resonance imaging is better for assessing the endocardium and offers better resolution.[12] We had a multidisciplinary discussion with the cardiologists and patient and concluded that it would be best to continue TEE. Although no consensus has been reached regarding appropriate treatment of cardiac vegetations and tumors, minimally invasive options will continue to become more prominent in any treatment algorithm. The present case demonstrates the functionality of the Inari FlowTriever device in offering another minimally invasive approach for these patients.

CONCLUSIONS

This is a novel case using the Inari FlowTriever device for removal of an isolated infected PFE. Percutaneous removal of cardiac tumors using mechanical

thrombectomy can be safely performed in the right patients. The use of minimally invasive options is reasonable in the setting of right-sided heart tumors and will continue to be used as advances in technology continue.

DISCLOSURES

None.

REFERENCES

1. Hakim FA, Aryal MR, Pandit A, et al. Papillary fibroelastoma of the pulmonary valve–a systematic review. Echocardiography 2014;31:234-40.
2. Trombetti D, Nardi P, Asta L, Pisano C, Altieri C, Ruvolo G. A rare localization of papillary fibroelastoma. Kardiochir Torakochirurgia Pol 2022;19:103-4.
3. Jullian L, Harrogate SR, Harky A, Roberts N. Presentation of papillary fibroelastoma of the aortic valve with atypical symptoms. BMJ Case Rep 2018;2018:bcr-2018-224367.
4. Uehara H, Uchiyama M, Hori T, Iida M, Imazuru T, Shimokawa T. Surgical treatment of papillary fibroelastoma of the pulmonary valve: a case report. J Cardiothorac Surg 2022;17:149.
5. Klarich KW, Enriquez-Sarano M, Gura GM, Edwards WD, Tajik AJ, Seward JB. Papillary fibroelastoma: echocardiographic characteristics for diagnosis and pathologic correlation. J Am Coll Cardiol 1997;30:784-90.
6. Sastre-Garriga J, Molina C, Montaner J, et al. Mitral papillary fibroelastoma as a cause of cardiogenic embolic stroke: report of two cases and review of the literature. Eur J Neurol 2000;7:449-53.
7. Grolla E, Dalla Vestra M, Zoffoli G, et al. Papillary fibroelastoma, unusual cause of stroke in a young man: a case report. J Cardiothorac Surg 2017;12:33.
8. Bicer M, Cikirikcioglu M, Pektok E, Müller H, Dettwiler S, Kalangos A. Papillary fibroelastoma of the left atrial wall: a case report. J Cardiothorac Surg 2009;4:28.
9. Toma C, Bunte MC, Cho KH, et al. Percutaneous mechanical thrombectomy in a real-world pulmonary embolism population: interim results of the FLASH registry. Catheter Cardiovasc Interv 2022;99:1345-55.
10. Kucher N, Boekstegers P, Müller OJ, et al. Randomized, controlled trial of ultrasound-assisted catheter-directed thrombolysis for acute intermediate-risk pulmonary embolism. Circulation 2014;129:479-86.
11. Tu T, Toma C, Tapson VF, et al. A prospective, single-arm, multicenter trial of catheter-directed mechanical thrombectomy for intermediate-risk acute pulmonary embolism: the FLARE study. JACC Cardiovasc Interv 2019;12:859-69.
12. Zamolo F, Calvagna C, D'Oria M, Sgorlon G, Zandonà L, Adovasio R. Neoplastic embolization from cardiac papillary fibroelastoma with unusual presentation: a case report. Ann Vasc Surg 2017;38:317.e1-4.

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