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IMAGING VIGNETTE

ADVANCED

CLINICAL VIGNETTE

Attempted Cardiopulmonary Bypass Venous Cannula Extraction Catheter Extraction of a Rare Intracardiac Myoepithelioma

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ABSTRACT

Myoepithelioma of the soft tissue is a rare entity that can mimic myxoma when presenting within the heart. We present a case where cardiopulmonary bypass venous cannula extraction catheter removal of an intracardiac myoepithelioma was attempted with minimal debulking and subsequently required minimally invasive open-heart surgery with cardiopulmonary bypass. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;7:101698) © 2023 The Authors. 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 76-year-old man with hypertension, diabetes mellitus, persistent atrial fibrillation, coronary artery disease, severe thrombocytopenia attributed to immune thrombocytopenia, and recent resection of a right shoulder myoepithelioma, presented to an outside hospital after multiple mechanical falls. Transthoracic echocardiography showed a large mass adherent to the tricuspid valve. The patient was transferred to our institution for consideration of cardiopulmonary bypass venous cannula extraction catheter removal (AngioVac, AngioDynamics) of the right-sided mass.

Computed tomography imaging of the chest demonstrated bilateral pulmonary emboli and large filling defects in the right internal jugular vein, subclavian vein, and superior vena cava, consistent with thrombus (**Figure 1A**). The patient was taken to the hybrid cardiac catheterization laboratory for extraction with a cardiopulmonary bypass venous cannula extraction catheter. Intraprocedural transesophageal echocardiography demonstrated a complex mass in the right atrium, prolapsing into the right ventricle during diastole (**Figure 1B**, Video 1). Cardiopulmonary bypass venous cannula extraction catheter debulking of the mass (**Figure 1C**, Video 2) retrieved only a small amount of tissue within the filter (**Figure 1D**), which was sent for pathologic evaluation (**Figure 1E**). Pathology results were consistent with myxoma, and because the mass was unable to be extracted with the cardiopulmonary bypass venous cannula extraction catheter, the decision was made to proceed to minimally invasive open-heart surgery in the same setting. The mass was found to be

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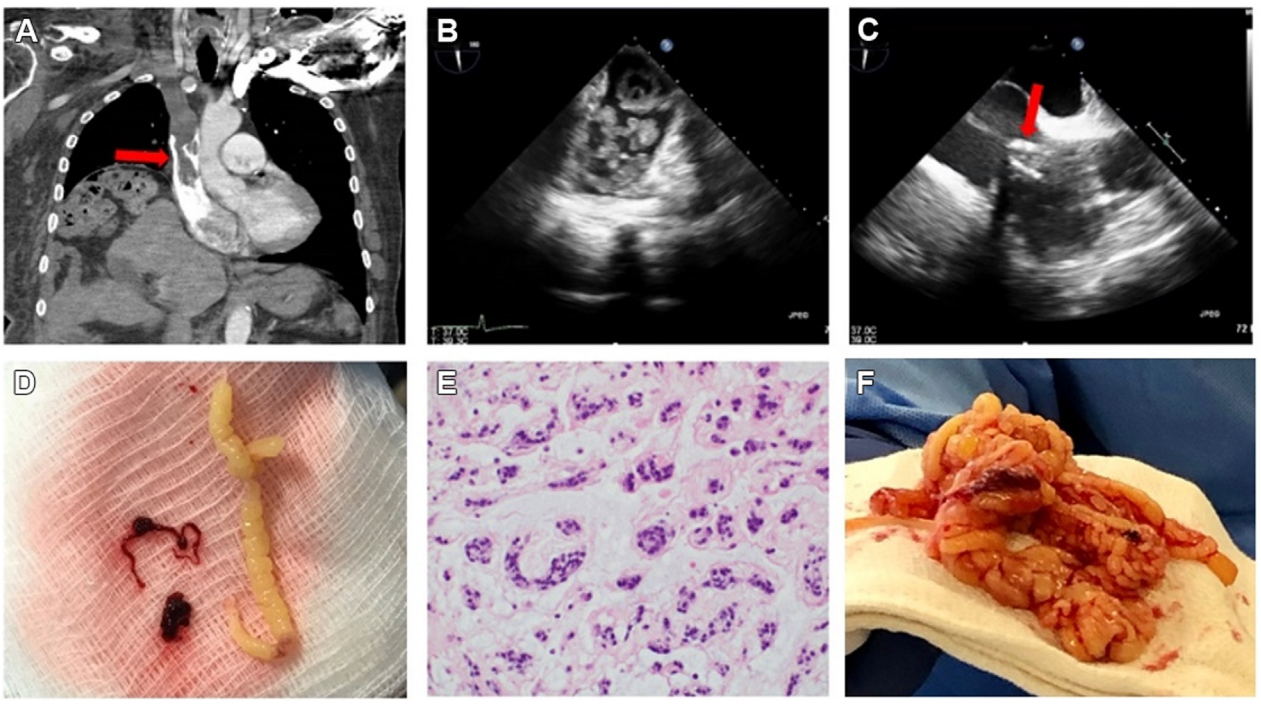
adherent to the walls of the right atrium and superior vena cava, resulting in incomplete removal at superior vena cava, with approximately 5% remaining. The specimen (**Figure 1F**) was sent for outside pathology consultation resulting in the diagnosis of myoepithelioma. There were no overt malignant histopathologic features, but given the context, it was thought to be metastatic spread from the recently resected right shoulder myoepithelioma. Given the patient's poor prognosis and progressive renal failure, the patient's family elected for hospice.

Myoepithelioma is a rare entity that typically occurs in the salivary glands. Myoepithelioma of the soft tissue is less common and is usually found in the trunk and limbs, where there is no known normal cellular counterpart.[1,2] They are often embedded within a myxoid stroma, potentially accounting for the initial diagnosis of myxoma on the frozen sections in this case.[2] Standard of care for treatment is not clearly defined given the rarity, but most patients undergo surgical resection, often without the ability to obtain complete margins, with or without adjunctive chemotherapy.[1] As a result, recurrence is relatively common.[1] Intracardiac involvement is exceedingly rare, but has been reported as both local spread from the mediastinum and a primary tumor in the right ventricular outflow tract in an infant.[3,4]

CONCLUSIONS

Myoepithelioma of the soft tissue is a rare entity that can mimic myxoma when presenting in the heart because of histopathologically similar myxoid stroma. Cardiopulmonary bypass venous cannula extraction catheter debulking of this type of mass has not been reported and was successful in obtaining a small fragment that allowed for initial pathologic evaluation with subsequent minimally invasive open-heart surgery.

FIGURE 1 Extraction of an Intracardiac Myoepithelioma



(A) Computed tomography of the chest demonstrating a large filling defect in the superior vena cava **(red arrow). (B)** Transesophageal echocardiogram demonstrating a complex mass in the right atrium, prolapsing into the right ventricle during diastole. **(C)** Transesophageal echocardiogram-guided cardiopulmonary bypass venous cannula extraction catheter removal of the mass (cannula, **red arrow**). **(D)** Small fragment of mass extracted and sent for pathologic evaluation. **(E)** Pathology of mass, initially identified as myxoma, but later identified as myoepithelioma upon outside consultation. **(F)** Gross pathology of the surgically resected mass.

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REFERENCES

1. Rastrelli M, Del Fiore P, Damiani GB, et al. Myoepithelioma of the soft tissue: a systematic review of clinical reports. *Eur J Surg Oncol J*. 2019;45(9):1520–1526.

2. Jo VY, Fletcher CDM. Myoepithelial neoplasms of soft tissue: an updated review of the clinicopathologic, immunophenotypic, and genetic features. *Head Neck Pathol*. 2015;9(1):32–38.

3. Briasoulis A, Salem N, Siddiqui F, Rashed A, Othman M. Myoepithelial carcinoma in the mediastinum involving the left atrium. *Cardiovasc Pathol*. 2016;25(2):158–160.

4. Patel P, Roberts R, Cole J, et al. A rare case of an intracardiac myoepithelial carcinoma in an infant. *J Pediatr Hematol Oncol*. 2019;41(4):e206.

KEY WORDS cancer, echocardiography, imaging

APPENDIX For supplemental videos, see the online version of this paper.