Unusual presentation of an obstructing cardiac myxoma

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Video clip is available online.

CLINICAL SUMMARY
A 16-year-old previously healthy female patient presented to the emergency department (ED) after suffering a syncopal episode 6 hours after donating blood. At the ED, she was diagnosed with anemia and a mild concussion. Cardiac auscultation revealed a systolic murmur. The rest of her physical examination was normal. Her family history was significant for 4 relatives who died prematurely from nonspecified heart disease and 5 additional distant relatives who suffered unexplained sudden cardiac death between their fourth and fifth decades of life. She was discharged home and followed-up with her primary care physician, who ordered an outpatient echocardiogram for evaluation of her murmur. The echocardiogram findings were notable for a large mass attached to the antero-superior portion of the right ventricle (RV), flailing in and out the pulmonary valve, and causing right ventricle outflow tract (RVOT) obstruction with an RVOT velocity of 3.9 m/s (Figure 1). Accordingly, the patient was referred urgently to our institution for further management.

On admission, computed tomography angiography of the chest confirmed the presence of a RV mass and no signs of distal pulmonary embolism. Cardiac magnetic resonance imaging (MRI) followed, which demonstrated a 5 × 3-cm mass in the RVOT (Video 1). Due to our concern for pulmonary embolism, the patient was taken urgently to the operating room from the MRI suite. The surgery was performed via median sternotomy, under cardiopulmonary bypass support. After opening the right atrium, a previously undetected patent foramen ovale was found and closed. The mass was attached to the RV free wall by a narrow stalk. Through the right atrium and pulmonary artery, the stalk of the mass was resected and the mass liberated en-bloc from the RV (Video 2). Perioperative frozen section showed no concern for malignancy. After confirming tricuspid valve competency, we closed the heart, and the patient was weaned off cardiopulmonary bypass. She was successfully extubated in the operating room and transferred to the cardiovascular intensive care unit. Postoperative recovery was unremarkable, and the patient was discharged home on postoperative day 4. The final pathology report indicated paucicellular myxoid spindle cell formation, consistent with cardiac myxoma.1

DISCUSSION
Pediatric cardiac tumors are rare. Their clinical presentation varies with the size and location of the tumor.1 In this case, the patient presented after donating blood and developing a syncopal episode several hours afterwards. Her symptoms were most likely secondary to RVOT

CENTRAL MESSAGE
An altruistic act of blood donation may lead to the finding and treatment of a rare and threatening right ventricle myxoma.
obstruction by the mass, combined with a relative hypovolemic state following the blood donation. Light-headedness and syncopal events related partly to vasovagal reactions and hypovolemia are the most common adverse events associated with blood donation, especially in female patients.\(^2\) Although regularly seen in the clinical practice, complete patient evaluation is warranted in selected patients, particularly if they have an extensive family history.

It is remarkable that a cardiac auscultation prompted an echocardiogram and the final diagnosis of a large intracardiac mass. Without the careful auscultatory examination in the ED, it is likely that this mass would have continued to go undiagnosed with a potential much worse outcome.

Echocardiogram is the gold standard for initial diagnosis of cardiac tumors.\(^3\) Other imaging modalities, including cardiac MRI, provide superior structural definition and tissue characterization, allowing to accurately plan a surgical approach.\(^4\)

Familial cardiac myxomas are rare, but due to the patient’s family history of unexplained sudden cardiac death, we decided to perform an echocardiogram on the patient’s siblings, finding no lesions. Although further investigation may be necessary, we recommend surveillance echocardiography on first-degree relatives of patients suffering from myxomas as an initial screening method.

The treatment of choice for cardiac myxomas is complete surgical resection,\(^2\) which was successfully achieved in this case, where an altruistic act of blood donation led to her diagnosis and opportune treatment. Oral and written informed consent for publication of this case was obtained from the patient and her mother.

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FIGURE 1. Transthoracic echocardiography showing a large, mobile mass (arrow) attached to the RV free wall. RV, Right ventricle; LV, left ventricle; Ao, aorta; LA, left atrium.

VIDEO 1. Cardiac MRI right 3-chamber view showing large mass flailing through the pulmonary valve and obstructing the RVOT. Video available at: https://www.jtcvs.org/article/S2666-2507(20)30305-9/fulltext.

VIDEO 2. En-bloc extraction of the RV cardiac myxoma through the tricuspid valve. Video available at: https://www.jtcvs.org/article/S2666-2507(20)30305-9/fulltext.