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Myocardial Infarction Secondary to Inflammatory Myofibroblastic Tumor Obstruction of the Left Main: Treated With Primary PCI

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Abstract

Introduction: Cardiac inflammatory myofibroblastic tumor is a rare tumor that can cause potentially fatal outcomes.
Case Presentation: We describe a case where the tumor originated on the mitral valve and extended through the left ventricular outflow tract and aortic valve and into the left main artery obstructing it. Due to the hemodynamic instability of the patient, we proceeded for cardiac catheterization with the intention to aspirate the mass but were forced to do percutaneous intervention to stabilize the patient and bridge him to surgery.
Conclusions: The patient underwent surgery several days later with complete resection of the tumor and coronary stent retrieval but his left ventricular ejection fraction remained poor after several months and he was referred for cardiac transplantation.

Keywords: ST Elevated Myocardial Infarction, Cardiac Tumor, Left Main Artery Disease

1. Introduction

The prevalence of primary cardiac tumors in childhood is reported at 0.08% with the most frequent tumor being rhabdomyomas (1). Inflammatory myofibroblastic tumor (IMT) most commonly occurs in the gastrointestinal tract and lung and rarely originates within the cardiac chambers. However, several case reports described potentially fatal outcomes with cardiac IMT due to sudden death. This case report describes the management of an 11-year-old male initially presenting with an ST elevated myocardial infarction of the left main artery and the outcomes of performing percutaneous intervention to alleviate the obstructive tumor.

2. Case Presentation

An 11-year-old male presented with chest pain after wrestling with his friends. He complained of flu like symptoms with subjective fevers 2 - 3 weeks earlier. When he presented to the emergency room, his EKG showed 9mm ST-segment elevation in the anterior and anterior-lateral leads Figure 1A. Echocardiography revealed a 1.0 × 1.4 mm pedunculated mass attached to the anterior mitral valve flowing through the left ventricular outflow tract (LVOT) with an ejection fraction of 20% Figure 1B. The initial diagnosis considered was endocarditis with embolization. The patient was intubated for respiratory distress secondary to significant pulmonary edema and had several episodes of non-sustained ventricular tachycardia on telemetry. He was brought emergently to the catheterization lab to evaluate his coronary arteries and fluoroscopy showed a filling defect in the distal left main artery (LM) extending into the bifurcation Figure 1C. Heparin and eptifibitide were started and an intra-aortic balloon pump (IABP) was placed. A BMW wire was advanced into the left circumflex (LCx) and a Choice PT wire was advanced into the left anterior descending artery (LAD). Alleviation of the filling defect using aspiration thrombectomy (Pronto LP, Vascular Solutions, Inc., Minneapolis, MN and Angiojet, Medrad, Inc., Warrendale, PA) was unsuccessful Figure 1D. Furthermore, when the catheters were advanced across the mass, thrombus embolized into the LAD and LCx causing acute no-reflow.

Due to the critical presentation of the patient, it was decided to proceed with percutaneous intervention to establish coronary flow and stabilize the patient in anticipation for possible surgery. Kissing balloon angioplasty was performed with two Emerge 2.0 × 12 mm balloons inflated to 6 ATM at the LM bifurcation with some improvement in blood flow. A Vision 2.5 × 12 mm bare metal stent (BMS) was advanced into the proximal LCx and a Vision 3.0 × 15 mm BMS was advanced into the distal LM to proximal LAD and deployed using the mini-crush technique and had simultaneous kissing balloon angioplasty post dilation Figure 1E. The filling defect propagated into the proximal to mid LM

and another Vision 4.0 × 15 mm BMS was deployed in the ostial to mid LM with final angiography revealing patent coronary TIMI-3 flow Figure 1F and G.

The patient was monitored in the ICU with continuous eptifibitide and maintained IABP placement. He had open-heart surgery 4 days later, which revealed an elongated mass that attached to the chordae and anterior mitral valve and extended through the LVOT and aortic valve into the LM. The mass was resected with preservation of the mitral valve and the coronary stents were removed from the LM Figure 1H and I. Pathology reported the vegetation likely due to inflammatory myofibroblastic tumor (IMT). After 3 months, the patient did not have any recovery of his left ventricular function and was referred for cardiac transplant.

3. Discussion

A previous publication of this case was reported in World Journal of Pediatric Congential Heart Surgery describing the surgical perspective while this publication was focused on the coronary intervention aspect (2). When the patient presented with STEMI and a large mass was seen in the LVOT on echocardiogram, the initial diagnosis was embolized vegetation. The patient was hypotensive on multiple IV pressors and not a candidate for immediate surgery. He was brought to the cardiac catheterization laboratory with the intention to treat his embolized vegetation with aspiration thrombectomy but there was little success. Surprisingly, the mass was highly thrombogenic and embolized multiple clots into the LAD and LCx causing no reflow. Due to the patient's hemodynamic instability, it was decided to proceed for percutaneous intervention with stent deployment to maintain coronary patency and to stabilize the patient. Coronary revascularization was successful after a total of 4 stent placements and the patient was transferred to the intensive care unit. He underwent surgery several days later and had successful resection of the tumor as well as retrieval of the coronary stents.

Inflammatory myofibroblastic tumor (IMT) was initially described in the lung in 1939 but has been reported to occur in multiple organs including the heart since then (3). The tumor is classified as low-grade neoplasm but when it originates within the heart, it may cause symptoms of shortness of breath, transient ischemic attacks, syncope, myocardial infarction, and sudden death. The histopathology of IMT is thought to be due to an exaggerated immunologic response to injury, inflammation, and infection by a proliferation of myofibroblastic mesenchymal spindle cells. Moreover, several reports revealed fibrin to be covering the surface of IMT, which may have explained why there were multiple thrombus embolizations during coronary intervention (4). Clinical presentation of these patients may include fever, growth impairment, iron-deficiency anemia, thrombocytosis, and hypergammaglobulinemia (5). Most often, these tumors are discovered incidentally during radiological studies and there are no established criteria to determine definitive diagnosis. Biopsies are often not enough for diagnosis and a whole specimen is usually needed. After resection of the tumor, there may be recurrence which can present as being much more rapid and aggressive than before leading to catastrophic outcomes (6).

This is the first known case report describing percutaneous intervention of a cardiac IMT obstruction of the left main artery. Surgery remains the definitive treatment and patients who are stable should proceed. For those who are hemodynamically unstable, coronary stenting can be considered as an option for temporary coronary revascularization and stabilization while bridging to surgery. Operators performing the catheterization should be aware that the tumor is highly thrombogenic and the patient should be on anticoagulation therapy. LM intervention is a complex high-risk procedure with high mortality but we report a successful outcome.

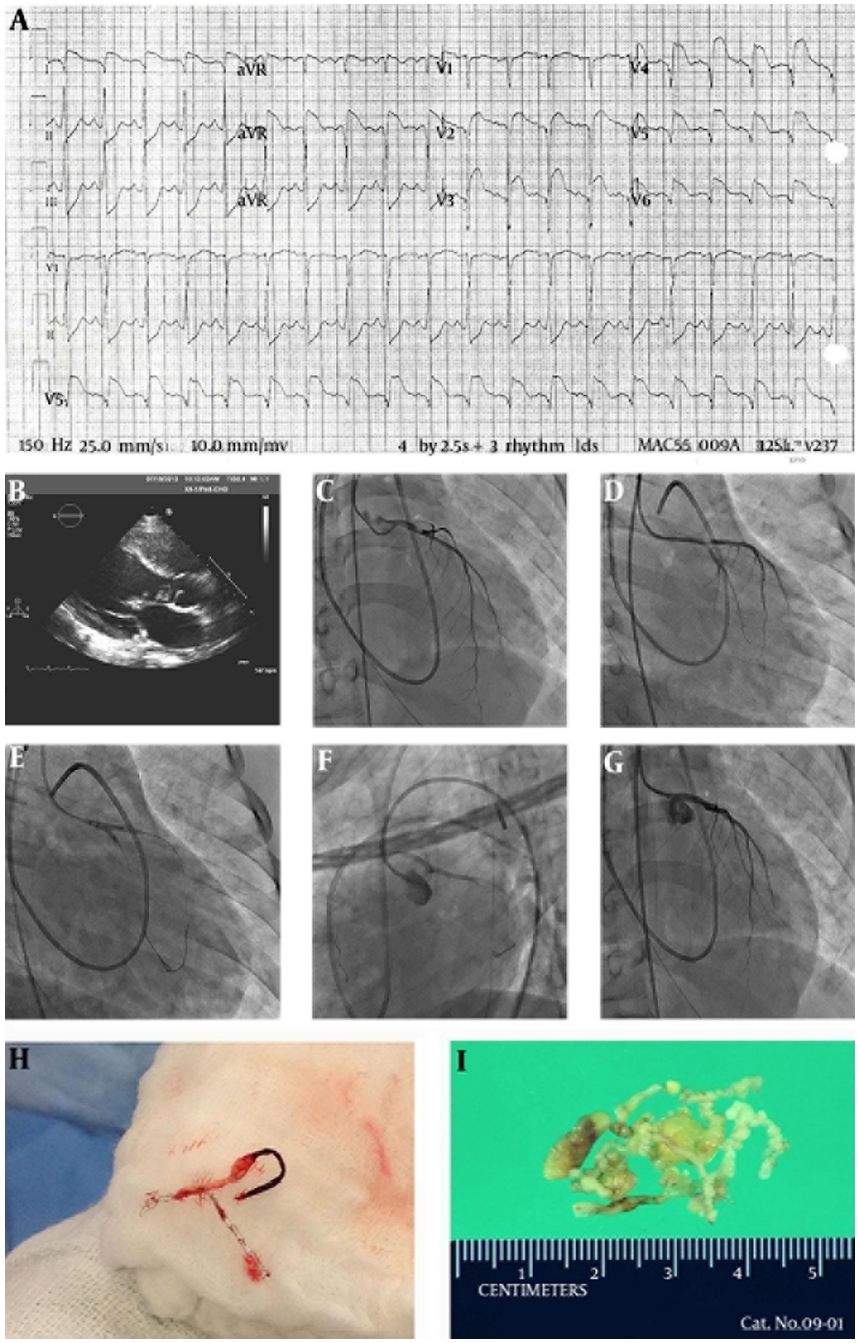

Figure 1. A, EKG demonstrating 9 mm ST-segment elevation in the anterior leads; B, pedunculated mass attached to the mitral leaflet protruding into the LVOT; C, filling defect at the distal left main extending into the bifurcation; D, unsuccessful aspiration thrombectomy; E, kissing balloon at the left main bifurcation; F, filling defect propagated into the prox to mid left main; G, final angiography demonstrating alleviation of the filling defect and TIMI-3 flow; H, resected elongated vegetation that extended from the mitral valve through the LVOT into the left main with retrieved coronary stents; I, resected elongated vegetation that extended from the mitral valve through the LVOT into the left main.

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