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Case Report

Mechanical pulmonic valve thrombosis: expanding role of cardiac CT and multimodality imaging ☆,☆☆,★

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ABSTRACT

We report a case of a 44-year-old man with a clinical history of Tetralogy of Fallot status post staged surgical correction with mechanical pulmonic valve replacement who presented with progressive exertional dyspnea in the setting of non-compliance with anticoagulation. In the context of this suggestive clinical presentation, the diagnosis of mechanical pulmonic valve thrombosis (MPVT) was made possible via multimodality imaging, including transthoracic echocardiogram and cardiac computed tomography angiography. Due to the uncommon nature of the condition, the patient was treated with systemic thrombolysis and anticoagulation using evidence-based guidelines, largely extrapolated from left-sided mechanical valve thrombosis. Our case underscores the importance of anticoagulation in MPVT and recognizing the features of MPVT on clinical history, physical examination, and multimodality imaging. It is essential to understand the pivotal role of multimodality imaging in the assessment of MPVT and realize the limitations of available data regarding the management of MPVT in the current era.

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Introduction

Mechanical pulmonic valve thrombosis (MPVT) is an uncommon, and hence, a challenging condition with only a few case reports and series reported in the literature [1]. Our case herein demonstrates the successful adoption and implementation of societal guidelines in a case of MPVT that is largely extrapolated from left-sided mechanical valve thrombosis management.

Case presentation

A 44-year-old man with past medical history of Tetralogy of Fallot status post staged surgical correction with mechanical pulmonic valve replacement in the 1980s presented with progressive exertional dyspnea and decline in his exercise tolerance for 1 year as well as intermittent lower extremity swelling. He was on lifelong anticoagulation with Coumadin until about 1.5 years earlier when he was lost to follow-up with no available labs or medication data in our system.

Differential diagnosis included MPTV, pannus formation, pulmonary hypertension, systolic and/or right-sided heart failure, among other possibilities. His general examination was unremarkable. He was afebrile with normal vital signs. Lungs were clear to auscultation bilaterally. His cardiovascular examination was notable for a harsh ejection systolic murmur with an early diastolic component, best heard over the pulmonary area with no audible "mechanical click."

A transthoracic echocardiogram (TTE) was obtained and revealed severe pulmonic valve stenosis and moderate regurgitation, with peak systolic velocity of 5 m/sec, peak systolic gradient of 100 mm Hg, and peak regurgitant gradient of 31 mm Hg (Fig. 1). Cardiac computed tomography angiography (CCTA), focusing the contrast timing to the right heart, was subsequently obtained for better characterization of the valve motion. CCTA depicted a bi-leaflet mechanical prosthesis with severely restricted leaflet mobility and multiple hypodense areas consistent with thrombus (Fig. 2, Panels A-C and Supplementary Video 1). A diagnosis of MPVT was made and he was managed per most recent American Heart Association/American College of Cardiology (AHA/ACC) guidelines for management of valvular heart disease, as outlined below.

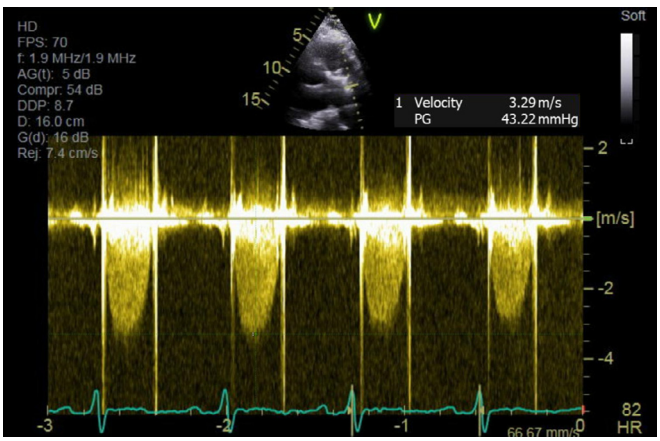

Fig. 1 – Continuous wave Doppler interrogation of the pulmonic valve. Spectral Doppler depicting significant pulmonic valve regurgitation and severe stenosis.

The patient was started on an unfractionated Heparin (UFH) drip which was later discontinued until his activated partial thromboplastin time was less than 50 seconds. This was followed by thrombolysis with intravenous alteplase infusion of 25 mg over 25 hours with no bolus. This process was repeated twice with each time being followed by a 6-hour infusion of UFH (70 unit/kg bolus and 16 unit/kg/hour with a target activated partial thromboplastin time of 1.5-2.0 times the control value in our laboratory). The following day, TTE demonstrated reduced transvalvular gradient (52 mm Hg) and velocity (3.6 m/sec; Fig. 3). His cardiovascular exam was now notable for an audible "mechanical click" appreciated on auscultation. Repeat CCTA was obtained which showed significantly improved leaflet mobility and reduction in thrombus burden (Fig. 4, Panels A-C and Supplementary Video 2).

Discussion

The risk of mechanical valve thrombosis is higher on right-sided mechanical valves as opposed to those on the left side, largely due to the slower blood flow in right-sided cardiac chambers [2]. With the improved survival of patients with congenital heart disease into adulthood and the need for re-do surgeries, there has been an increase in the use of mechanical valves in the pulmonic position [3]. This patient population presents unique challenges owing to their complex past medical and surgical histories. Moreover, there is paucity of data on the optimal diagnostic and therapeutic approaches in this patient population.

Patients with MPVT most commonly present with signs and symptoms of right-sided heart failure [2]. Less commonly, they may present with pulmonary embolism or paradoxical systemic embolism [4]. There will often be a history of non-compliance with anticoagulation, as with our patient [1]. When MPVT is suspected, the first-line imaging modality is TTE [5]. Increased transvalvular gradients should raise the index of suspicion [4,6]. If the initial evaluation is suggestive of MPVT, the AHA/ACC guidelines now recommend further evaluation with multi-modality imaging, such as CCTA [7]. This recommendation was promoted from "class II-a, level of evidence (LOE)-C" in the 2014 guidelines to "class I, LOE-B" in the 2020 guideline update [8]. CCTA enables anatomic and dynamic leaflet assessment, which supported its choice for the assessment of MPVT in our case [4]. Alternative imaging modalities include fluoroscopy and transesophageal echocardiography, the latter being more useful in left-sided valves and chambers [1].

MPVT is a life-threatening condition and requires prompt treatment. Fibrinolytic therapy should be considered as first-line treatment, provided there are no absolute contraindications (class II-a recommendation, LOE-B) [1,6]. Low-dose slow infusion of tissue plasminogen activator (eg, alteplase) is con-

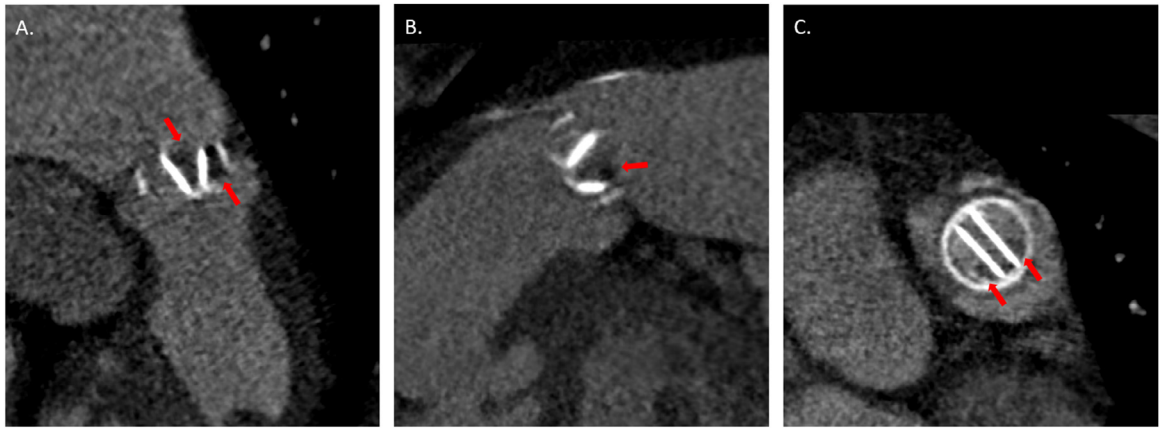

Fig. 2 – Computed tomographic multiplanar reformats of the mechanical pulmonic valve before thrombolysis. Long axis (A and B) and short axis (C) views demonstrating the mechanical prosthetic pulmonic valve with large thrombus burden (red arrows) in a fixed open position.

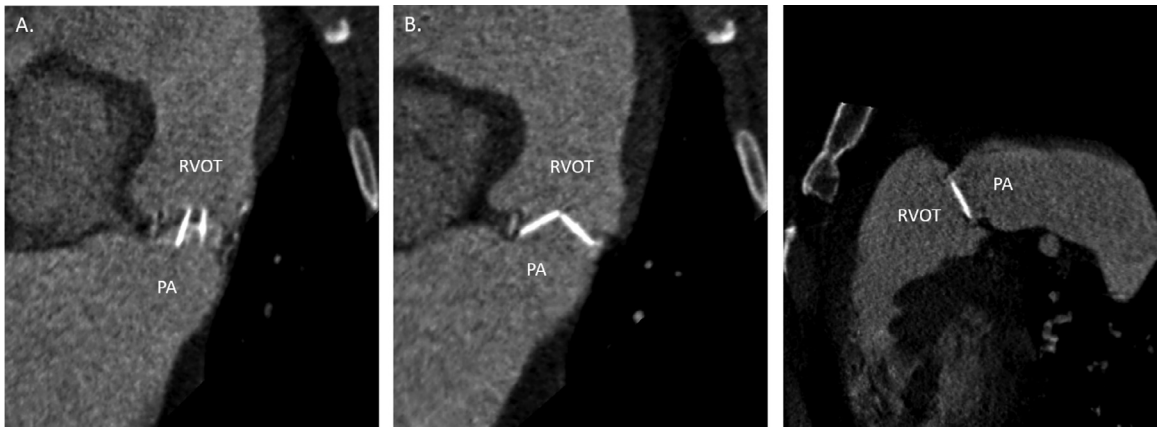

Fig. 3 – Computed tomographic multiplanar reformats of mechanical pulmonic valve after thrombolysis. Long axis during systole (A) and diastole (B and C) demonstrating improved leaflet motion.

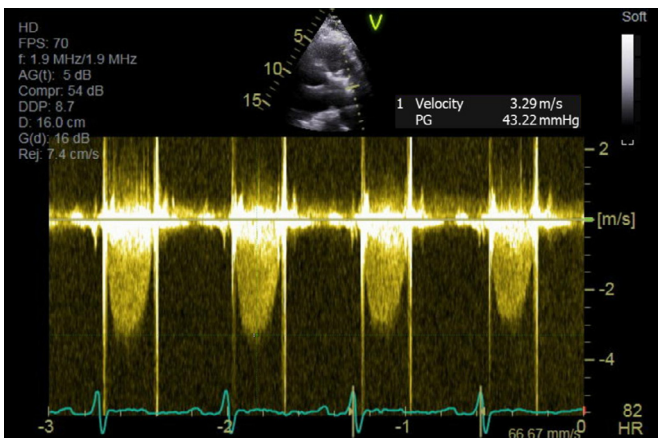

Fig. 4 – Continuous wave Doppler interrogation of the pulmonic valve after thrombolysis. Spectral Doppler depicting no pulmonic valve regurgitation, reduction in gradient, and restoration of "metallic click" Doppler signals at the beginning and end of systole.

sidered safe and effective [9]. This should be followed by a 6-hour infusion of UFH and repeat TTE [9]. The low-dose infusion protocol can be repeated if there is no improvement in transvalvular gradients, to a maximum cumulative alteplase dose of 200 mg [10]. Surgical valve replacement or thrombectomy are alternative treatment options in patients who do not respond to medical therapy, but are associated with high postoperative mortality. Hence, surgery is usually reserved for select cases with left-sided mechanical valve thrombosis [1,6].

Patients who undergo successful fibrinolysis should be treated with low-dose aspirin (75-100 mg daily) plus warfarin, if they had not been on aspirin [4,6]. Given the high risk for recurrent thrombosis, especially if the international normalized ratio (INR) had been in the therapeutic range, it is recommended to aim for a higher INR goal (eg, 3.0-4.0). This goal may be further increased to (3.5-4.5) if patients develop MPVT while on aspirin and had a therapeutic INR [1,6].

Our patient was successfully bridged to warfarin and started on aspirin, with close follow-up for further assessment with TTE and/or CCTA [6].

Conclusions

MPVT may present a clinical conundrum. We highlight the role of fibrinolytic therapy as a safe and effective first-line treatment modality. It would be prudent to report more cases as such when they are encountered in the clinical setting to advance our knowledge and the current state of evidence. This may further validate the described treatment protocol in similar cases.

Our case underscores the importance of anticoagulation in MPVT and recognizing the features of MPVT on clinical history, physical examination, and multimodality imaging. It is essential to understand the pivotal role of multimodality imaging in the assessment of MPVT and realize the limitations of available data regarding the management of MPVT in the current era.

Patient consent statement

Patient's consent was obtained for publishing this de-identified case.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2022.05.079.

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