

CASE REPORT

ADVANCED

CLINICAL CASE

Severe Functional Tricuspid Stenosis Due to Phosphoglyceride Crystal Deposition Disease in Right Atrium



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ABSTRACT

This case report concerns a 72-year-old-female with severe functional tricuspid stenosis due to phosphoglyceride crystal deposition disease and a history of atrial septum closure and tricuspid valvuloplasty. Phosphoglyceride crystal deposition disease is extremely rare, and percutaneous transcatheter biopsy under intracardiac echocardiographic guidance proved to be useful for its diagnosis. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:230-235)
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HISTORY OF PRESENTATION

A 72-year-old-female was admitted to the local hospital because of exertional dyspnea and lower-extremity edema. The patient had a chest x-ray and an increase in cardiothoracic index was described therefore a transthoracic echocardiogram (TTE) was requested. The patient was then transferred to our institution for further examination of the mass. Physical examination showed blood pressure of

134/62 mm Hg and a regular pulse of 84 beats/min. An electrocardiogram performed at our institution showed a normal sinus rhythm and inverted T-wave in V₁ to V₂. Laboratory data revealed elevated brain natriuretic peptide of 109 pg/mL, and TTE revealed a normal left ventricular (LV) ejection fraction of 61% and an LV end-diastolic volume of 62 mL and LV end-systolic volume of 24 mL, both normal. The iso-echoic solid mass measuring 35 × 40 mm was detected in the right atrium, which was attached to the interatrial septum and tricuspid annulus (**Figure 1, Videos 1 and 2**). The mass had also extended to the anterior and posterior leaflet of the tricuspid valve and aortic root (**Figure 1, Videos 1 and 2**). Furthermore, severe functional tricuspid stenosis caused by this mass was observed with a pressure half-time of 237 ms and mean pressure gradient of 9.2 mm Hg (**Figure 2, Video 3**). Transesophageal

LEARNING OBJECTIVES

- To understand the usefulness of differential diagnosis of intracardiac mass for patients with a previous history of cardiac surgery.
- To gain a better understanding of the pathogenesis of phosphoglyceride crystal deposition disease.

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echocardiography revealed similar findings to those obtained with TTE (Figure 3, Video 4). Three-dimensional transesophageal echocardiography clearly showed narrowing of the right ventricular inflow tract as a result of the mass (Figure 4, Video 5). Contrast-enhanced computed tomography showed a solid enhanced mass in the right atrium which was attached to the interatrial septum to the tricuspid annulus (Figure 5, Video 6). Cardiac magnetic resonance imaging showed a mass in the right atrium with isointense on T1-weighted image and a mixture of isointense and hypointense areas on T2-weighted image (Figure 6). ¹⁸F-fluorodeoxy glucose-positron emission tomography imaging showed a significantly increased uptake only by the mass, which was suspected of being a primary malignant cardiac tumor (Figure 7).

PAST MEDICAL HISTORY

The patient had a previous history of atrial septum closure for an atrial septal defect and tricuspid valvuloplasty for tricuspid regurgitation, respectively, 26 years previously. The patient has also

received methotrexate, salazosulfapyridine, famotidine, and risedronate sodium for rheumatoid arthritis for the previous 20 years.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of the mass in the right atrium based on the patient's presentation included myxoma or lipoma if benign, and malignant lymphoma or sarcoma if malignant. Because the mass extended to the normal surrounding tissue including tricuspid valve, tricuspid annulus, and aortic root, complete surgical resection of mass seemed to be impossible. Thus, we decided that a percutaneous transcatheter biopsy of the mass was required for a correct diagnosis.

INVESTIGATIONS

A 9-F sheath was inserted into the right internal jugular vein for the percutaneous transcatheter biopsy of the right atrial mass. Ten sequential samples were taken under the guidance of intracardiac

ABBREVIATIONS AND ACRONYMS

- LV = left ventricular
- PGDD = phosphoglyceride crystal deposition disease
- TTE = transthoracic echocardiogram

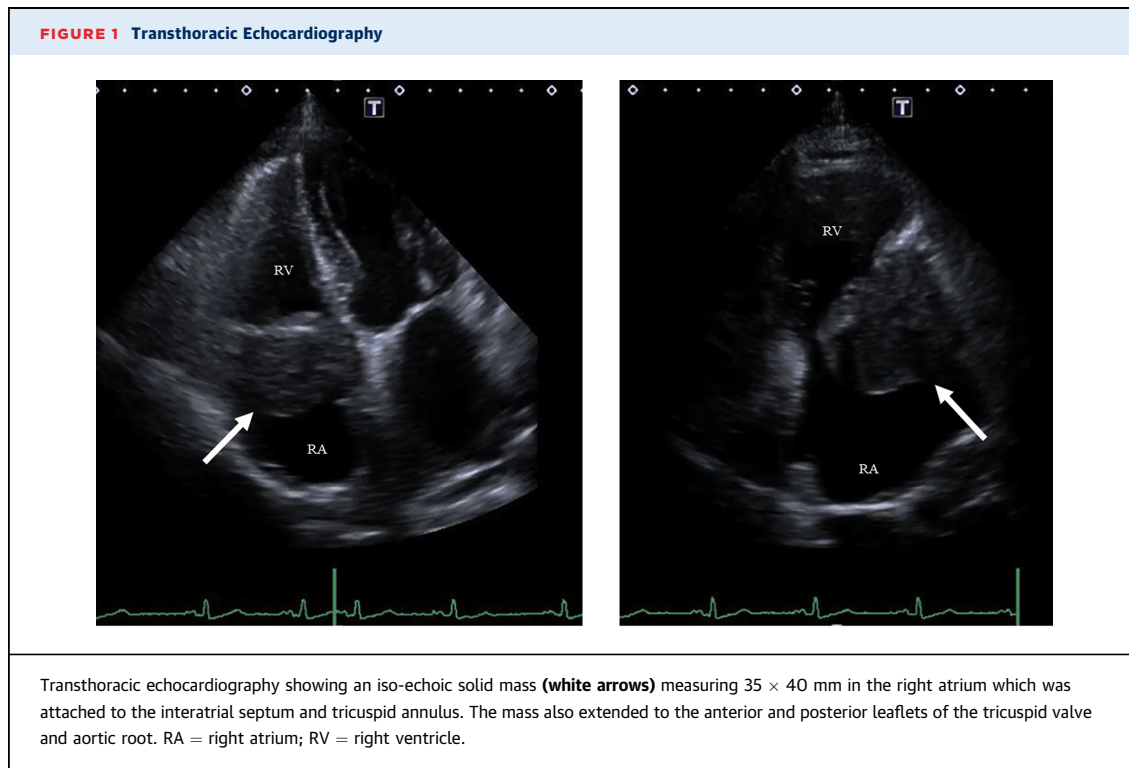
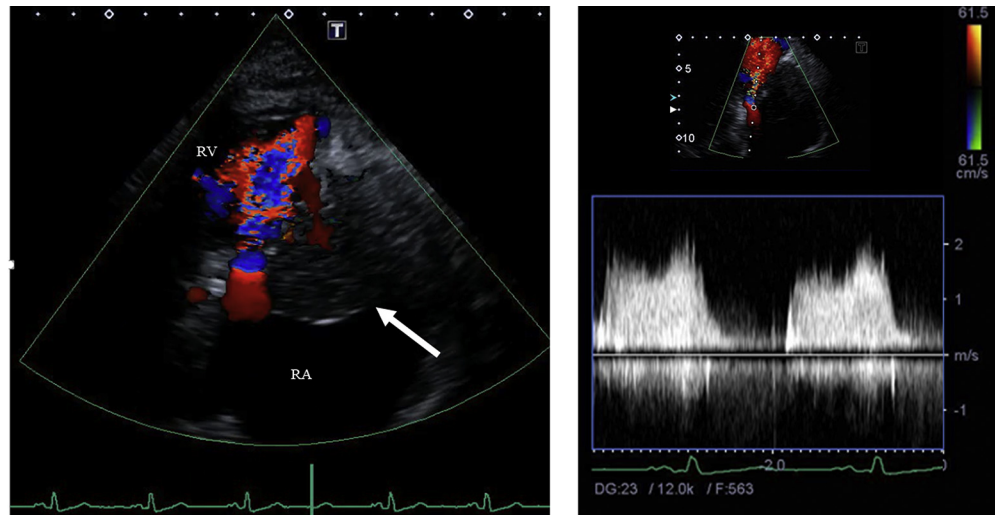
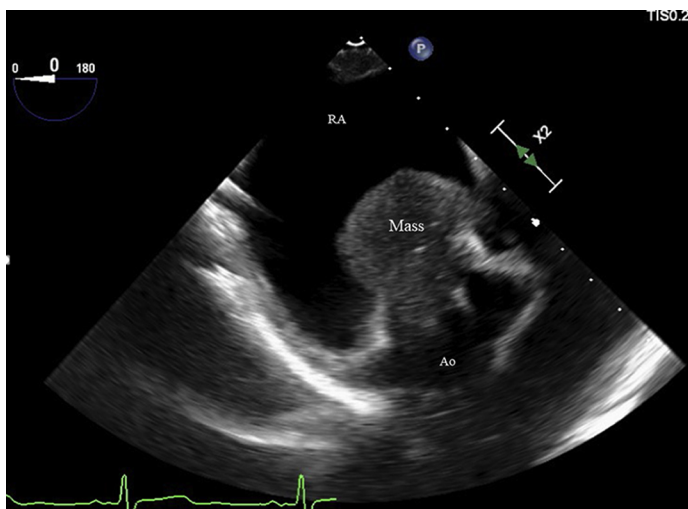
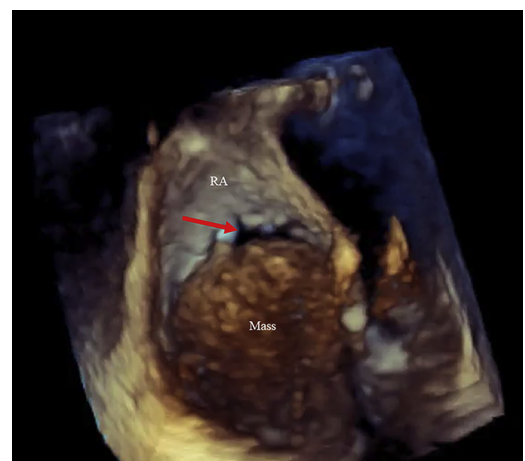


FIGURE 2 Transthoracic Echocardiography (Color Doppler)

Transthoracic echocardiography showing severe functional tricuspid stenosis due to this mass (**white arrow**) with a pressure-half time of 237 ms, and mean pressure gradient of 9.2 mm Hg. Abbreviations as in [Figure 1](#).

FIGURE 3 Transesophageal Echocardiography

Ao = aortic route; RA = right atrium.

FIGURE 4 3-Dimensional Transesophageal Echocardiography

Three-dimensional transesophageal echocardiography showing narrowing of the right ventricular inflow tract (**red arrow**) caused by the mass. RA = right atrium.

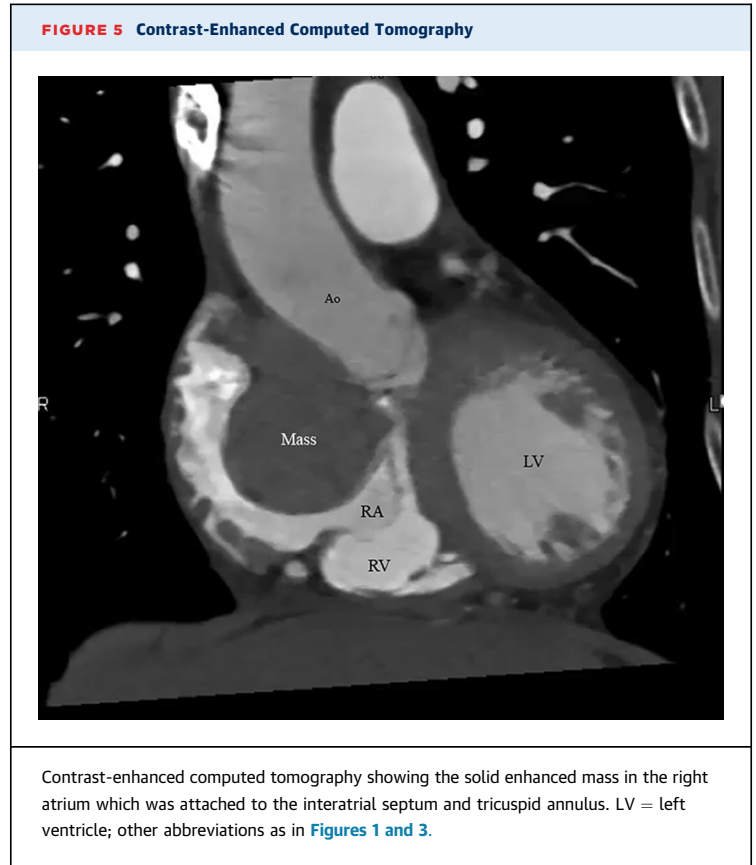
echocardiography without any procedural complications (Figure 8, Video 7).

MANAGEMENT

Pathological examination using hematoxylin-eosin staining revealed that a granular substance 50 μ m in size was surrounded by multinucleated giant cells (Figure 9A). In addition, it revealed that foreign body granulomas consisting of crystal deposition which characteristically showed an eosinophilic fibrinous rim and string-like crystals under polarized light (Figure 9B). Furthermore, this deposition showed positive when the gold hydroxamic acid method was used for screening for phosphoglyceride (Figure 9C). These findings led to a diagnosis of phosphoglyceride crystal deposition disease (PGDD). Our heart team came to the conclusion that complete surgical resection of the mass was impossible because of widespread invasion of the right-sided heart; therefore, careful periodic TTE will be performed.

DISCUSSION

PGDD is a very rare disease defined as phosphoglyceride crystals deposited as tumors in the soft tissue or bone with no relation to the joint (1-3). Phosphoglycerides are a class of phospholipids, which include lecithin and cephalin, and constitute a major component of cell membranes. Phosphoglyceride crystal deposition disease is considered a lipid metabolic disorder, occurring predominantly in injured soft tissues and forming foreign body



granulomas (1,4). PGDD sometimes results in the formation of large masses, which may be misdiagnosed as malignant tumors. Although little is known about the etiology and pathogenesis of PGDD, this deposition disease is apparently triggered by injury and subsequent macrophage aggregation, and

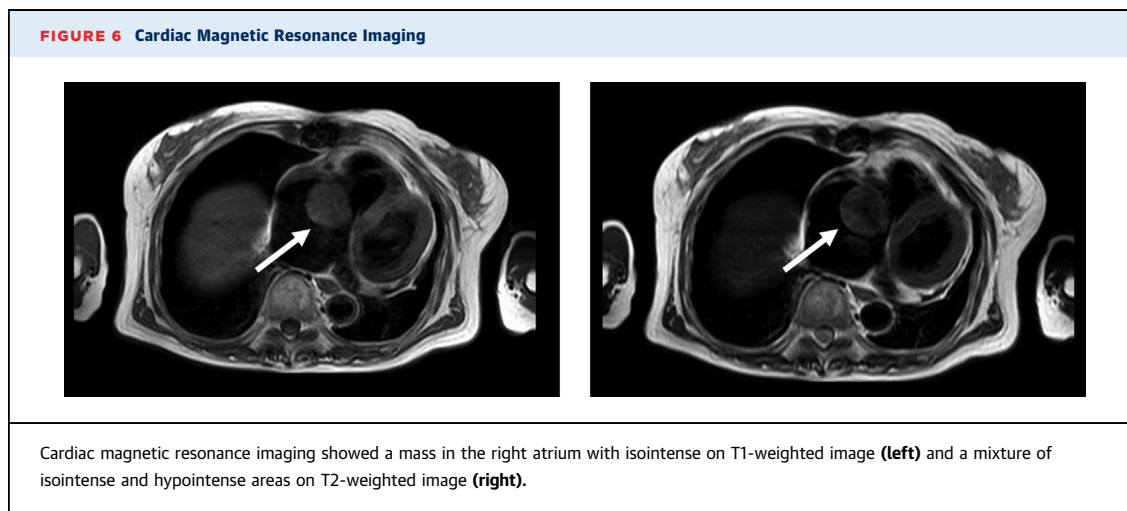
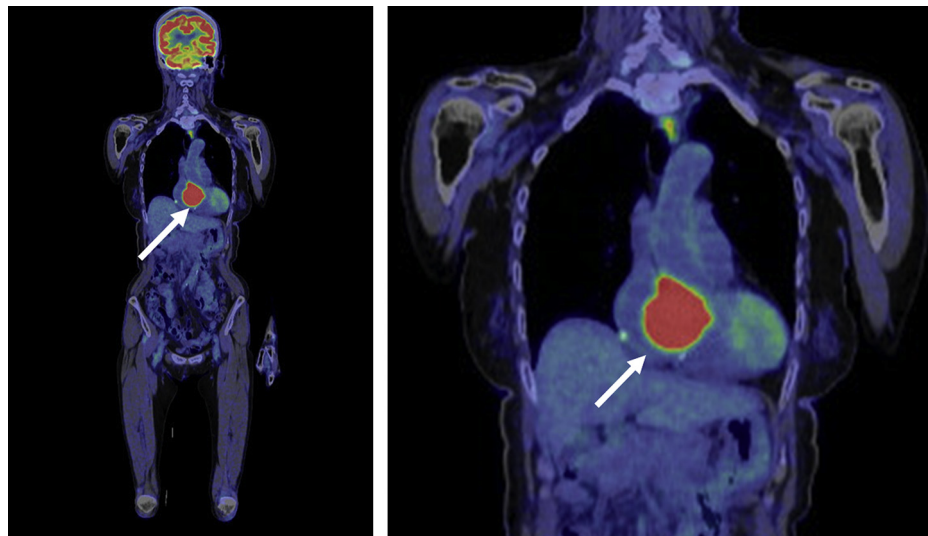


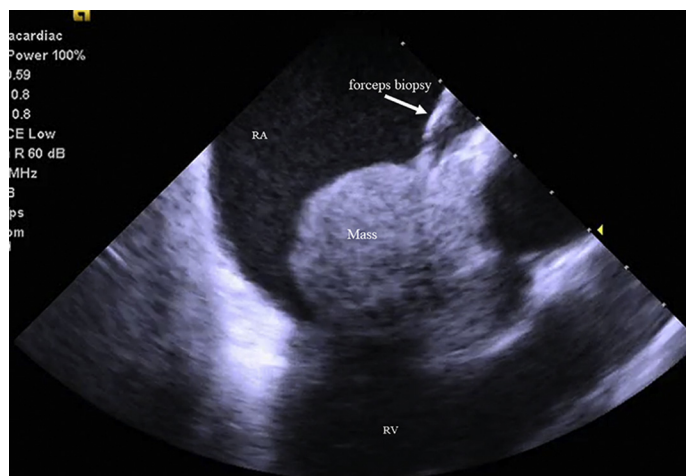
FIGURE 7 ¹⁸F-Fluorodeoxy Glucose-Positron Emission Tomography

¹⁸F-fluorodeoxy glucose-positron emission tomography showing a significantly increased uptake only by the mass.

histologically shows deposited crystals radiating from the cell membranes of epithelioid cells. This disease thus tends to occur at the sites of invasive procedures, including surgical sites (1,4).

To the best of our knowledge, only 14 cases of PGDD have been published up to June 2021 on PubMed. In most cases, phosphoglyceride crystal

deposition disease occurred at sites susceptible to invasion due to a previous injury. In our case, phosphoglyceride crystal deposition might have occurred from atrial septum closure and tricuspid valvuloplasty scarring. Severely functional tricuspid stenosis due to the mass led to exertional dyspnea and lower-extremity edema so that careful echocardiographic follow-up was needed. Ours was the first case in which PGDD had a direct impact on hemodynamics.

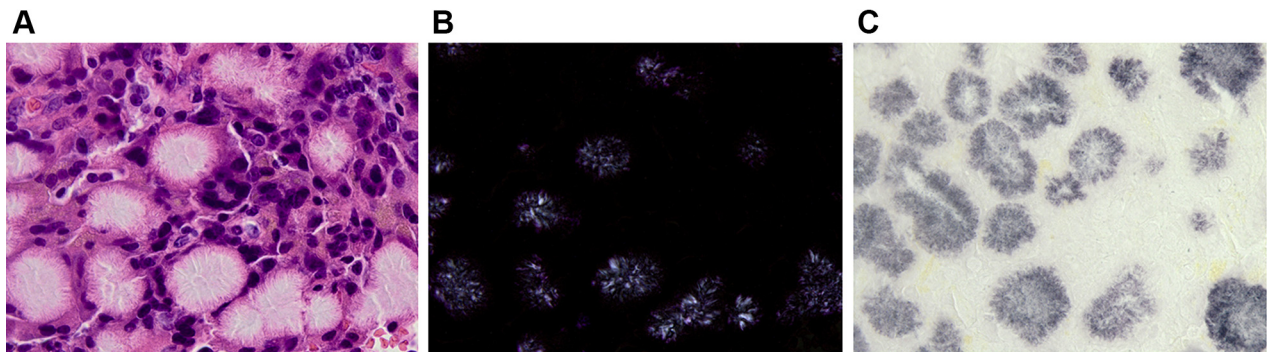
FIGURE 8 Percutaneous Transcatheter Biopsy Under Guidance of Intracardiac Echocardiography

Abbreviations as in Figure 1.

FOLLOW-UP

Before discharge, exercise stress echocardiography by means of supine bicycle exercise was performed to determine the association of functional severe tricuspid stenosis with exertional dyspnea. At a maximal work load of 25 W, mean trans-tricuspid pressure gradient increased to 15.1 mm Hg, and the increase in the peak tricuspid regurgitation pressure gradient from 14.6 mm Hg to 19.4 mm Hg. In addition, the decrease in right ventricular outflow tract velocity time integral changed from 11.8 to 12.9 cm. In addition, TTE was performed every 6 months after the diagnosis of PGDD because total resection of the mass could not be performed. In addition, based on the findings of exercise stress echocardiography we decided that no therapeutic intervention was required due to severe functional tricuspid stenosis caused by the mass. The size of the mass, 35 ×

FIGURE 9 Pathological Examination Findings



Pathological examination using hematoxylin-eosin staining revealed that a granular substance 50 mm in size was surrounded by multinucleated giant cells (A). Foreign body granulomas consisting of crystal deposition which characteristically showed an eosinophilic fibrinous rim and string-like crystals under polarized light (B). The deposition showed positive when the gold hydroxamic acid method was used for screening for phosphoglyceride (C).

37 mm, remained unchanged 12 months after the diagnosis of PGDD. In addition, functional tricuspid stenosis with a pressure-half time of 208 ms, and mean pressure gradient of 6 mm Hg, had not become worse.

CONCLUSIONS

PGDD is an extremely rare condition, but it should be taken into consideration in the differential diagnosis of an intracardiac mass in patients with a previous history of cardiac surgery. Percutaneous transcatheter biopsy under guidance of intracardiac

echocardiography proved to be useful for the diagnosis of PGDD.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS functional tricuspid stenosis, percutaneous transcatheter biopsy, phosphoglyceride crystal deposition

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