CASE REPORT

Chylopericardium associated with constrictive pericarditis assessed by multimodality imaging

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Abstract
Acute-onset presentation with breathlessness and calcific pericardial thickening encapsulating the heart. Extremely chylous pericardium, which is by itself rare, in combination with constriction assessed with multiple imaging modalities.

KEYWORDS
cardiac magnetic resonance imaging, cardiac surgery, chylopericardium, constrictive pericarditis, multimodality imaging

1 CASE REPORT

An 80-year-old male patient developed exertional dyspnea and bilateral peripheral edema while on holiday in Australia. Investigations including an echocardiogram, cardiac computed tomography, and cardiac magnetic resonance suggested calcific pericardial thickening encapsulating the heart with associated constriction. This is an interesting case as constriction was associated with a large chylopericardium of unknown cause.

There was no history of fever, productive cough, or joint pain. Initial investigations including an echocardiogram and a cardiac computed tomography (CT; Figures 1 and 2; Videos S1 and S2) suggested calcific pericardial thickening encapsulating the heart with associated constriction. A diagnostic angiogram was normal. The diagnosis of constrictive pericarditis was confirmed with right heart catheterization.

The patient was referred for further assessment with cardiac magnetic resonance imaging (CMR). Biventricular size and global systolic function were in the normal range with no evidence of myocardial necrosis. There was prominent pericardial thickening of both layers and mild to moderate pericardial effusion with inspiratory left ventricular (LV) septal bounce in mid-LV short-axis free-breathing sequences. The findings were consistent with a diagnosis of an effusive constrictive pericarditis.

The patient was referred for surgical treatment and underwent pericardial resection via median sternotomy without cardiopulmonary bypass. Multiple membranous cream pieces of tissue showing patchy areas of extensive calcification were identified, the largest one measuring 1.52 × 1.65 × 3 mm in thickness (Videos S3 and S4). No discrete nodule or mass was detected. A collection of chylous effusion was evacuated which is particularly unusual in association with constrictive pericarditis. Cytology demonstrated an abundance of lymphocytes. Further, biochemistry, histology, and microbiology assessment was sent. Biochemistry revealed an exudate, while histology ruled out a lymphoproliferative disorder and
other malignancies, as well as sarcoid. Fluid and tissue cultures were also negative for tuberculosis and other pathogens. The patient was subsequently followed up in cardiothoracic and cardiology clinic with yearly echocardiography. Also, a repeated CMR 12 months postsurgery showed preserved systolic function with no significant evidence of ventricular interdependence. With respect to symptoms, the patient experienced an immediate postsurgical improvement and has been stable thereafter.

2 | DISCUSSION

Chylopericardium is a pericardial effusion comprised of chyle, the normal content of the lacteals (lymphatics of the small intestine), and thoracic duct.\textsuperscript{1} Chylopericardium may be primary (idiopathic) or, much more often, secondary to a communication between the pericardial sac and the thoracic duct as a result of trauma, congenital anomalies, or as a complication of open-heart surgery, mediastinal lymphangiomas, lymphangiomatous hamartomas, lymphangiectasis, and obstruction or anomalies of the thoracic duct.\textsuperscript{2,3}

When the diagnosis of chylopericardium has been established, investigations specifically looking for malignant disease, lymphoma, and tuberculosis should be carried out. A history of trauma caused by thoracic surgery or blunt injury, the introduction of subclavian venous catheters, or episodes of vomiting or violent coughing should be looked for.\textsuperscript{4} Our patient did not have a history of treatment for tuberculosis nor any other conditions of the above-mentioned, which is consistent with primary idiopathic chylopericardium.

The treatment of chylopericardium is mostly conservative at the beginning, with failure of this approach as high as 57%. These patients most likely will need more invasive management like a pericardial window or surgery.\textsuperscript{1} More data are needed on clinical benefits of screening for chylous pleural and pericardial effusions in secondary causes such as primary lymphedema patients regardless of symptoms.\textsuperscript{5} However, extremely limited data exist in primary chylopericardium which is associated additionally with constrictive pericarditis as in our case; therefore, there is no widely accepted guidance.\textsuperscript{6} We suggest an individualized approach that constrictive pericarditis should be considered in the differential diagnosis of chylous ascites and chylothorax and could actively be sought with a suitable imaging modality (ie CMR, CT) additionally to echocardiography in the presence of relevant evidence. Specific final identifiable causes (nonviral–nonidiopathic) should be considered as part of the investigations, as well as high-risk features, including tamponade, recurrences, and constriction.\textsuperscript{1} Further data from case series/reports are required which would be of paramount importance for our better understanding and management of this very rare pathophysiological presentation.
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CONFLICT OF INTEREST
None declared.

AUTHOR CONTRIBUTIONS
EA: is corresponding author and took full responsibility for the contents of this manuscript; KB: diagnosed and followed up the patient and conceived of the case report; TPC: diagnosed and follow-up the patient; VC: operated and was the responsible surgeon.

ETHICAL APPROVAL
All procedures were in accordance with the ethical standards of the institutional and national research committee and with the Helsinki Declaration and its later amendments or comparable ethical standards. Also, informed consent was obtained from the patient involved in this case report.

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SUPPORTING INFORMATION
Additional supporting information may be found online in the Supporting Information section.

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