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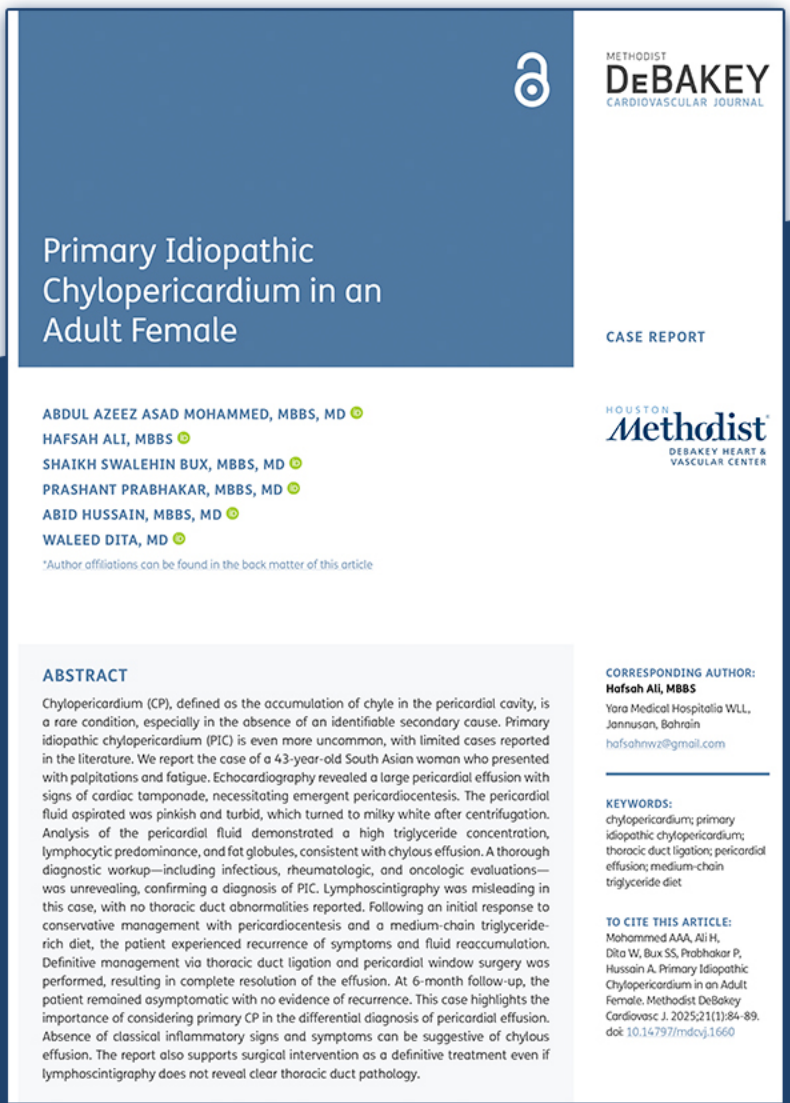
We are thrilled to announce that our latest case report, **Primary Idiopathic Chylopericardium** in an Adult Female, has been officially published in the prestigious **Methodist DeBakey Cardiovascular Journal**



METHODIST DEBAKEY CARDIOVASCULAR JOURNAL

Primary Idiopathic Chylopericardium in an Adult Female

CASE REPORT

ABDUL AZEEZ ASAD MOHAMMED, MBBS, MD
HAFSAH ALI, MBBS
SHAIKH SWALEHIN BUX, MBBS, MD
PRASHANT PRABHAKAR, MBBS, MD
ABID HUSSAIN, MBBS, MD
WALEED DITA, MD
*Author affiliations can be found in the back matter of this article

HOUSTON Methodist DEBAKEY HEART & VASCULAR CENTER

ABSTRACT

Chylopericardium (CP), defined as the accumulation of chyle in the pericardial cavity, is a rare condition, especially in the absence of an identifiable secondary cause. Primary idiopathic chylopericardium (PIC) is even more uncommon, with limited cases reported in the literature. We report the case of a 43-year-old South Asian woman who presented with palpitations and fatigue. Echocardiography revealed a large pericardial effusion with signs of cardiac tamponade, necessitating emergent pericardiocentesis. The pericardial fluid aspirated was pinkish and turbid, which turned to milky white after centrifugation. Analysis of the pericardial fluid demonstrated a high triglyceride concentration, lymphocytic predominance, and fat globules, consistent with chylous effusion. A thorough diagnostic workup—including infectious, rheumatologic, and oncologic evaluations—was unrevealing, confirming a diagnosis of PIC. Lymphoscintigraphy was misleading in this case, with no thoracic duct abnormalities reported. Following an initial response to conservative management with pericardiocentesis and a medium-chain triglyceride-rich diet, the patient experienced recurrence of symptoms and fluid reaccumulation. Definitive management via thoracic duct ligation and pericardial window surgery was performed, resulting in complete resolution of the effusion. At 6-month follow-up, the patient remained asymptomatic with no evidence of recurrence. This case highlights the importance of considering primary CP in the differential diagnosis of pericardial effusion. Absence of classical inflammatory signs and symptoms can be suggestive of chylous effusion. The report also supports surgical intervention as a definitive treatment even if lymphoscintigraphy does not reveal clear thoracic duct pathology.

CORRESPONDING AUTHOR:
Hafsah Ali, MBBS
Yara Medical Hospitalia W.L.L, Jannusan, Bahrain
hafsahnwz@gmail.com

KEYWORDS:
chylopericardium; primary idiopathic chylopericardium; thoracic duct ligation; pericardial effusion; medium-chain triglyceride diet

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INTRODUCTION

Chylopericardium (CP), an accumulation of chyle in the pericardial sac, is a very rare clinical entity. It can be primary (idiopathic) or secondary arising from mediastinal neoplasm, injury to thoracic duct, thrombosis of the subclavian vein, tuberculosis, nonsurgical trauma, or thoracic or cardiac surgery. Primary idiopathic chylopericardium (PIC), the type with no known clear etiology, is even rarer.[1] The term "primary CP" was first used by Groves and Effler in 1954 to describe a case of isolated accumulation of chyle in the pericardium without any evident cause.[2] The diagnosis of CP can be made by analyzing the pericardial fluid analysis, while lymphoscintigraphy and lymphangiography are used to identify the location of the chyle leak. Presentation can be symptomatic or asymptomatic. Common symptoms include cough, dyspnea, chest pain, palpitations, or cardiac tamponade depending upon the severity of CP.[2] Management of disease can be conservative or surgical, which is the most effective treatment and associated with favorable outcomes. We report a case of recurrent primary idiopathic CP in a 43-year-old woman that was successfully treated with pericardial window with thoracic duct ligation along with placement on a low-fat diet rich in medium chain triglycerides.

CASE DESCRIPTION

A 43-year-old South Asian woman presented to our emergency department with complaints of palpitations and fatigue for the past week. She had diagnosed hypertension and hypothyroidism that were well-controlled with medications. She had mild fever 2 weeks prior that lasted for 1 to 2 days. A week later, she started experiencing palpitations and fatigue and was evaluated in a local private hospital, where she was found to have moderate pericardial effusion. She was started on colchicine, presuming post viral pericarditis. Three days later, she presented to us with worsening of symptoms. There was no history of chest pain or systemic features such as loss of appetite, weight loss, or prolonged fever.

On arrival, she was conscious and alert but reported palpitations and easy fatiguability. She was afebrile. Her heart rate and blood pressure were 78 bpm and 110/80 mm Hg, respectively. Cardiac examination revealed distant heart sounds and raised jugular venous pressure. The rest of her physical examination was unremarkable. Electrocardiogram showed low voltage complexes. An emergency echocardiogram confirmed large pericardial effusion with impending cardiac tamponade (Figure 1), and urgent pericardiocentesis was done with aseptic measures under fluoroscopic and echocardiographic guidance. Needle placement was confirmed by aspiration of thick turbid pericardial fluid. A 6F pigtail catheter was placed in the pericardial cavity via fluoroscopy, which facilitated effective drainage of the effusion (Figure 2). About 300 mL of thick turbid pericardial fluid was aspirated and sent for analysis.

The catheter was left in the pericardial cavity for continuous drainage, and the patient was shifted to the

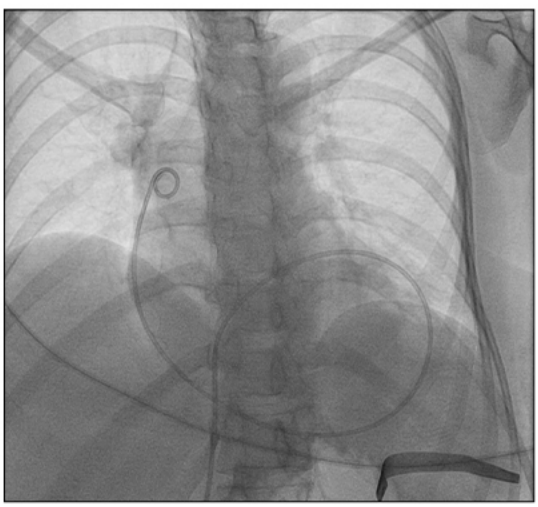

Figure 2 Fluoroscopic image showing pigtail catheter in situ in the pericardial cavity.

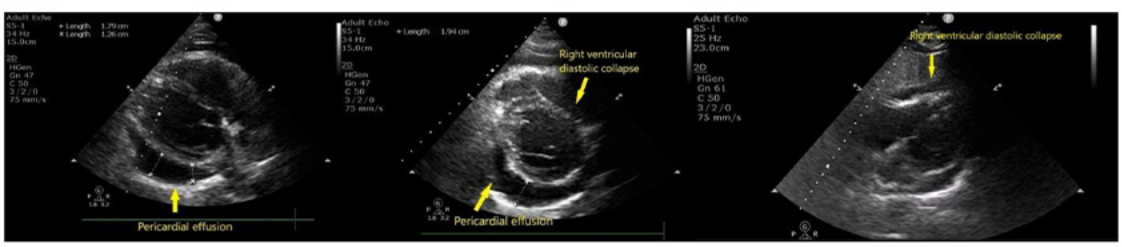

Figure 1 Two-dimensional echocardiographic still images. Left: subcostal view showing global pericardial with right ventricular diastolic collapse. Center: short-axis view showing large global pericardial effusion with right ventricular diastolic collapse. Right: parasternal long-axis view showing global pericardial effusion, more towards the inferior side, marked by arrow.

intensive care unit. About 100 mL of fluid was aspirated through the catheter overnight and then reduced to 5 to 10 mL every 2 hours. Confirmation of clearance of pericardial effusion was done by bedside echocardiography, and pericardial catheter was removed after 2 days.

Initial physical analysis of pericardial fluid showed pinkish turbid fluid. After centrifugation, this appeared milky white with deposition of red blood cells at the base of the tube (Figure 3). The pericardial fluid analysis revealed lymphocytic predominance with no malignant cells, high protein level, normal glucose levels, and very high triglyceride levels. Fluid cultures, viral and bacterial polymerase chain reaction (PCR), adenosine deaminase levels, and mycobacterium tuberculosis PCR were negative, as was gamma interferon. Special staining showed fat globules on microscopy. These findings were consistent with chylous pericardial effusion.[3] The composition of pericardial fluid is enumerated in Table 1.

An antinuclear antibody panel was done after rheumatologist consultation and was negative for connective tissue diseases. All other blood investigations including complete blood count, inflammatory markers, liver and renal function tests, and lipid profile were normal. Computed tomography (CT) of the chest confirmed the presence of a large pericardial effusion with no evidence of pulmonary or mediastinal pathology. Positron emission tomography CT showed no hypermetabolic activity suggestive of malignancy (Figure 4). Lymphoscintigraphy showed no evidence of major lymphatic structure abnormality (Figure 5).

Total volume	200 ml
Total WBC count	9000 cells/mm^3
Neutrophils	1%
Lymphocytes	98%
Total RBC count	58000 cells/mm^3
Adenosine deaminase	< 10 IU/L
Mesothelial cells	1%
Gram stain	Few neutrophils Gram positive Cocci in chains+
AFB* stain	Negative
PCR for Tubercule bacilli	Negative

Table 1 Pericardial fluid analysis. WBC: white blood cell; RBC: red blood cell; AFB: acid-fast bacillus; PCR: polymerase chain reaction
*Acid fast bacteria

During her hospital stay, the patient remained asymptomatic with no fever spikes, chest pain, or palpitations. She developed slow and steady reaccumulation of the fluid. Repeat pericardiocentesis was done after 2 weeks and 185 mL of thick pinkish turbid fluid was aspirated, which turned milky supernatant after centrifugation revealed lymphocytic predominance and fat globules on microscopy (Figure 5).

In view of recurrent accumulation of chyle in the pericardial sac, the patient was recommended surgical treatment. She underwent pericardial window and thoracic duct ligation in her native country under general anesthesia. After the uneventful procedure, she was started on injection octreotide and oral prednisolone and was kept on a low-fat, medium-chain, triglyceride-rich diet. The right pleural drain was removed on the seventh postoperative day. At her 6-month follow-up, there were no signs or symptoms of CP.

DISCUSSION

Chyle is the normal content of lacteals and the thoracic duct,[4] and CP is an accumulation of chyle in the pericardial cavity. Chylopericardium can be primary (idiopathic) or secondary CP. Before making the diagnosis of idiopathic CP, all secondary causes of CP must be excluded. The first case of idiopathic CP was reported in 1888 by Hasebroek. It is a very rare clinical entity, with only 104 cases of primary idiopathic CP reported in the past 65 years, between January 1950 and December 2015.[5]

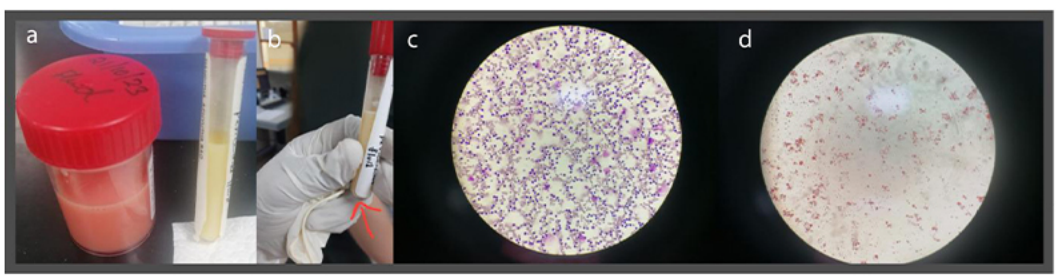

Figure 3 (A) Note pinkish turbid fluid immediately after pericardiocentesis **(B)** showing settling of red blood cells after centrifugation, leaving milky white turbid fluid with classical appearance of chyle; **(C)** lymphocyte predominance on microscopy, and **(D)** with fat stain of supernatant fluid positive for fat globules.

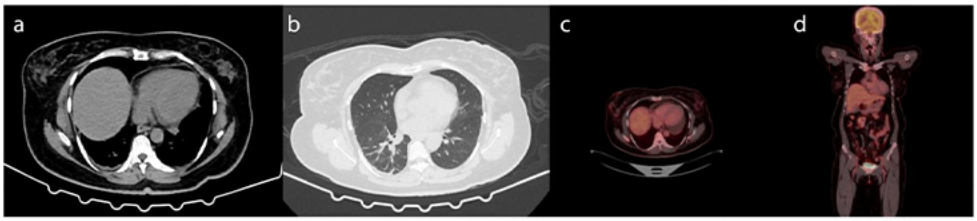

Figure 4 (A,B) Computed tomography (CT) chest shows pericardial effusion with normal lung and other thoracic structures; **(C,D)** positron emission tomography CT shows pericardial effusion with no active uptake of fluorodeoxyglucose.

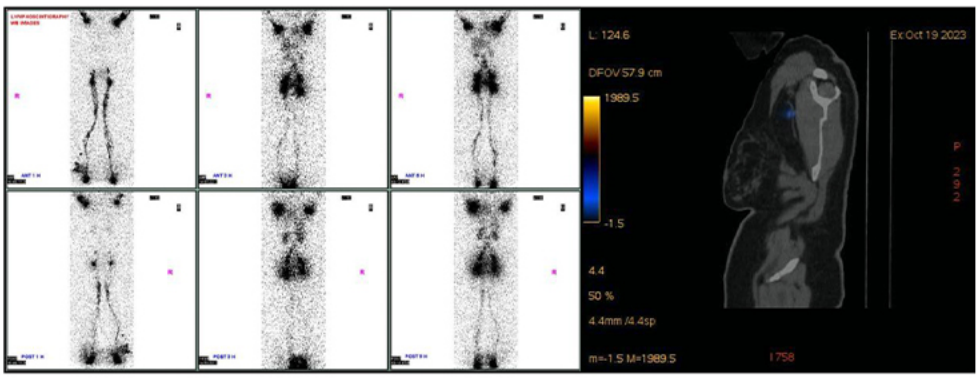

Figure 5 Lymphoscintigraphy shows normal lymphatic flow with normal appearance of thoracic duct.

Although its etiology is uncertain, the possible mechanisms include the following: (1) lymphatic valvular impairment on the branches that are connected to the thoracic duct and pericardium lymphatic vessels; (2) increased thoracic duct pressure that can occur in lymphangiectasia; (3) abnormal communication between lymphatic vessels and pericardial lymphatics leading to chylous reflux; and (4) congenital malformation.[5] Absence of systemic inflammatory features and chest pain suggest the possibility of chylous effusion. Diagnostic modalities for evaluating CP include chest x-ray to identify cardiomegaly and echocardiography, CT to help identify pericardial effusion, pericardiocentesis to reveal the nature of effusion, and CT and lymphoscintigraphy to help establish the cause of primary CP. It is important to rule out malignancy, lymphoma, and tuberculosis and ascertain a history of any type of trauma, previous thoracic surgery, and/or insertion of subclavian venous catheter.[1] In our case, we failed to visualize any communication between the thoracic duct and the pericardial sac by lymphoscintigraphy, possibly due to low or delayed uptake of the radiotracer.

The diagnosis of CP depends on pericardial fluid analysis. Physical analysis should be done immediately as well as after centrifugation and may reveal the classical milky white appearance of chylous fluid. Chylous pericardial fluid that is milky white with triglycerides > 500 mg/dL, cholesterol/triglyceride ratio < 1.0, negative culture, and preponderance of lymphocytes is consistent with the diagnostic criteria of CP.[4] Treatment options for managing CP include pericardiocentesis, thoracostomy drainage using a catheter, dietary management with medium-chain triglyceride and a low-fat diet, pericardiectomy, pericardial window formation, ligation of the thoracic duct above the level of the diaphragm, and a pericardial-peritoneal (Denver) shunt.[1]

Initially, a patient should be managed conservatively by pericardiocentesis or pericardial drain supported with dietary modifications or total parenteral nutrition. Unfortunately, some patients do not respond to medical management and require surgical intervention. In most cases, surgery involving thoracic duct ligation is curative.[6] Surgical ligation of the thoracic duct with a pericardial window through right posterolateral thoracotomy has been reported by Putra et al. as a definitive treatment.[4] Chemical pericardiodesis using tetracycline to treat recurrent pericardial effusion has been advocated by Bhat et al.[3] External drainage of the pericardial sac via pericardiostomy in addition to dietary support with medium-chain triglycerides and low-fat meals resulted in absolute resolution of recurrent pericardial effusion as reported in another case series.[1] Although most authors reported surgery as the definite treatment method,

Lopez-Castilla et al. reported the first case of primary idiopathic CP in a 2-month-old boy successfully treated nonoperatively.[7] Of the 104 reported cases of primary or idiopathic CP from the past 65 years, the majority of patients (71.2%) required surgery for definitive treatment, with thoracic duct ligation being the preferred procedure (44.23%). Follow-up data from 64 patients showed 100% survival over an average follow-up period of 12 months.[5] Thoracic duct ligation above the diaphragm is considered the best choice, favored by most surgeons. Video-assisted thoracoscopic surgery is gaining popularity among cardiothoracic surgeons due to its minimally invasive nature and lower impact on pulmonary function. Thoracic duct embolization is an alternative option, particularly for patients who either refuse or are unable to tolerate more invasive surgery or anesthesia. A newer surgical technique involves reconstructing the occluded thoracic duct as another therapeutic option. In our patient, conservative management with pericardiocentesis and a medium-chain triglyceride diet initially alleviated symptoms; however, a recurrence occurred. As a result, thoracic duct ligation became the mainstay of treatment, ultimately leading to the complete resolution of the pericardial effusion.

CONCLUSION

Primary idiopathic CP is a rare disease. Clues to diagnosis include large recurrent effusion with absence of chest pain and other systemic features as well as milky white appearance and high triglyceride levels in the fluid. Secondary causes of CP must be ruled out first by thorough investigations. Conservative therapy is the first-line treatment for CP. While a medium-chain diet or total parenteral nutrition can be considered, most patients will still need pericardiocentesis for symptom relief. Definitive surgical treatment is recommended if recurrent CP occurs. Surgical management is the most successful treatment and is associated with a favorable prognosis.

ETHICS AND CONSENT

Patient consent was obtained for this study.

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COMPETING INTERESTS

The authors have no competing interests to declare.

AUTHOR AFFILIATIONS

Abdul Azeez Asad Mohammed, MBBS, MD orcid.org/0009-0009-0871-1310
Bahrain Specialist Hospital, Manama, Bahrain
Hafsah Ali, MBBS orcid.org/0009-0009-1928-6735
Yara Medical Hospitalia WLL, Jannusan, Bahrain
Shaikh Swalehin Bux, MBBS, MD orcid.org/0000-0002-7774-5700
Bahrain Specialist Hospital, Manama, Bahrain
Prashant Prabhakar, MBBS, MD orcid.org/0009-0000-3270-4772
Bahrain Specialist Hospital, Manama, Bahrain
Abid Hussain, MBBS, MD orcid.org/0000-0002-6984-7803
Bahrain Specialist Hospital, Manama, Bahrain
Waleed Dita, MD orcid.org/0000-0002-4642-4293
Awali Hospital, Awali, Bahrain

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