Asian Journal of Cardiology Research

Volume 7, Issue 1, Page 328-332, 2024; Article no.AJCR.125596

Successful Management of Rare Idiopathic Chylopericardium Mimicking as Pyopericardium in an Infant

Pradeep Kumar Jain [a++*], Nimish Rai [b#], Vivek Kumar Athwani [c†] and Abhinav Agarwal [d†]

[a] Department of Pediatric Cardiology, NSCB Medical College Jabalpur, India.
[b] Department of Cardiovascular and Thoracic Surgery, NSCB Medical College, Jabalpur, India.
[c] Department of Pediatrics, SKG Medical College, Sikar, India.
[d] Department of Pediatrics, Era Medical College, Lucknow, India.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/125596

Case Report

Received: 15/08/2024
Accepted: 27/10/2024
Published: 31/10/2024

ABSTRACT

Chylopericardium is a rare entity which is characterised by accumulation of chyle in pericardial sac. The absence of an underlying etiology is defined as idiopathic chylopericardium. In pediatric age group chylopericardium can resemble pyopericardium. If suspected the diagnosis of chylopericardium could be easily made with fluid analysis. The management of idiopathic chylopericardium could be conservative or surgical, but there are no guidelines. We report a case of idiopathic chylopericardium initially diagnosed as pyopericardium and its successful management.

[++]Associate Professor;
[#]Professor;
[†]Assistant Professor;
**Corresponding author: Email: docpradeepjain@gmail.com;*

Cite as: *Jain, Pradeep Kumar, Nimish Rai, Vivek Kumar Athwani, and Abhinav Agarwal. 2024. "Successful Management of Rare Idiopathic Chylopericardium Mimicking As Pyopericardium in an Infant". Asian Journal of Cardiology Research 7 (1):328-32. https://journalajcr.com/index.php/AJCR/article/view/235.*

Keywords: Chylopericardium; triglyceride; pyopericardium; pediatric; pericardiectomy.

1. INTRODUCTION

Chylopericardium (CP) is an exceedingly rare condition characterized by the accumulation of chyle within the pericardium. It may be idiopathic, or primary, when no etiology is identified. The term was first coined by Groves and Effler in 1954 [1]. Secondary CP arises due to malignancy, lymphangiomatosis, trauma, or post-surgical complications [2]. In the pediatric population, CP can resemble infective pyopericardium, which is more common. However, idiopathic CP should be considered, especially in the absence of symptoms and signs of infection as treatment for both conditions differ significantly. Diagnosis of CP is straightforward with pericardial fluid cholesterol and triglyceride level analysis [3]. Currently, there are no consensus guidelines for managing idiopathic CP. This report presents a pediatric case of idiopathic CP initially diagnosed as pyopericardium and its successful management.

2. CASE PRESENTATION

A 10-month-old male presented with shortness of breath and was found to have a massive right-sided pleural effusion on chest X-ray. Initially treated for empyema with intercostal drainage and antibiotics following which his symptoms improved. Cardiomegaly on chest X-ray prompted for Echocardiography which revealed significant pericardial effusion. The child was then referred to pediatric cardiology department for suspected pyopericardium management.

There was no history of fever, trauma, surgery or tuberculosis contact. There was no significant family history. Physical examination revealed heart rate of 120/min, respiratory rate of 32/min, blood pressure of 95/65 mmHg and a regular heart rhythm with distant heart sound.. The patient was alert without jugular venous distension, had clear lung fields with equal bilateral air entry. The abdomen was soft without tenderness, hepatosplenomegaly or lymphadenopathy.

Chest X-ray confirmed an enlarged cardiac silhouette. Transthoracic echocardiography at our center showed a large effusion with a 'dancing heart' appearance and early tamponade signs i.e. partial collapse of the right atrium and ventricle free wall in diastole and a dilated inferior vena cava—yet normal ejection fraction (Fig. 1). Laboratory tests indicated normal blood counts, normal C-reactive protein levels, and an erythrocyte sedimentation rate of 14 mm/h (normal <15 mm/h). A purified protein derivative skin test was negative.

To address dyspnea and determine its cause, the patient underwent pericardiocentesis using a 6 F pigtail catheter and 280 ml of yellowish fluid was removed, which alleviated the symptoms (Fig. 2). The pericardial fluid analysis showed sugar levels at 122 mg/dl, protein at 6.1 g/dl, LDH at 150 U/L, ADA at 4.3 U/L, and a cell count of 1800/mm3 with predominantly lymphocytes. Bacterial culture and TB-PCR were negative.

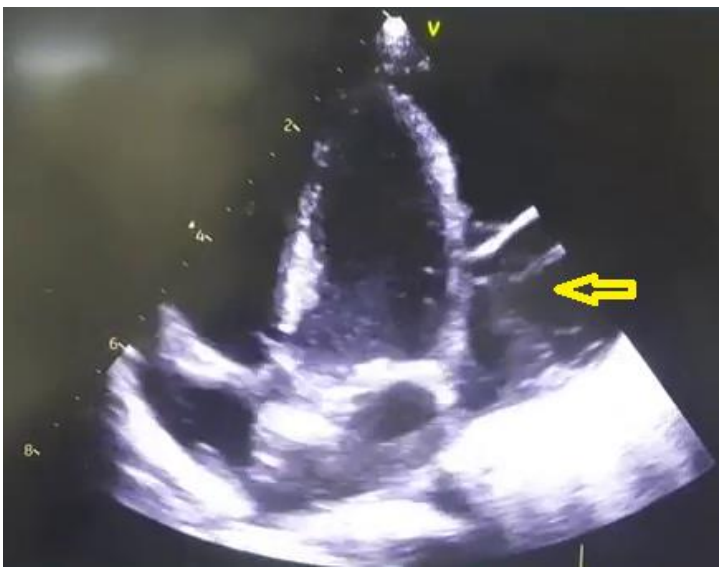

Fig. 1. ECHO Image at the time of admission showing significant pericardial effusion

Given the child's good health, absence of fever or signs of sepsis, normal blood counts, and lymphocyte-predominant pericardial fluid cell count, the initial diagnosis of pyopericardium was reconsidered. Subsequent testing for cholesterol and triglyceride levels was conducted with chylopericardium in mind. The pericardial fluid's triglyceride level was 1220 mg/dl and cholesterol was 169 mg/dl, yielding a cholesterol/triglyceride ratio of 0.13, confirming the fluid as chylous.

Despite minimal drainage after 48 hours and symptomatic improvement, echocardiography and CT scan revealed significant pericardial collection (Fig. 3). No tumors or lymphangiomas were detected on the CT scan as potential causes of chylopericardium. Due to resource limitations, lymphangiography or lymphoscintigraphy were not performed. With no history of trauma or surgery and other causes secondary CP ruled out and diagnosis of idiopathic chylopericardium was made. The patient was initially kept nil by mouth, followed by a fat-free diet enriched with protein, carbohydrates, and medium-chain triglycerides (MCT).

As conservative treatment showed no improvement, video-assisted thoracoscopic surgery (VATS) for pericardiectomy and pleuropericardial window creation was performed. Persistent significant drainage even after 72 hours post-surgery led to the initiation of octreotide infusion. Drainage gradually decreased from the 12th post-operative day and became serous in nature. Echocardiography showed minimal pericardial collection (Fig. 4), allowing for the removal of pericardial and pleural drains. The patient remained on a fat-free diet and was discharged on the 22nd day after surgery. A two-month follow-up showed no significant pericardial collection.

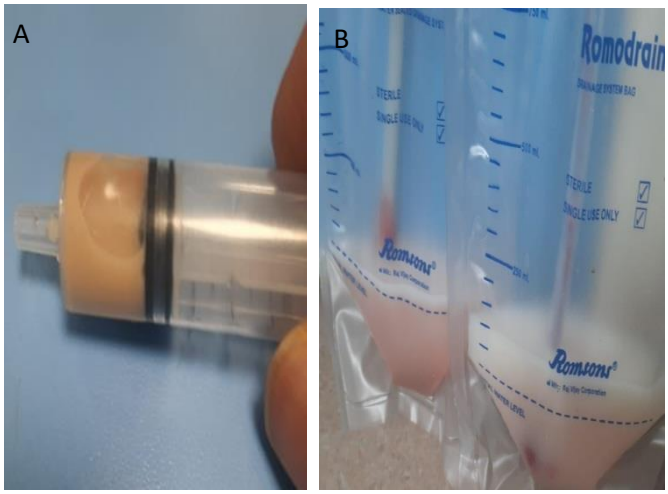

Fig. 2. Pericardial fluid before(A) and after surgical drainage (B)

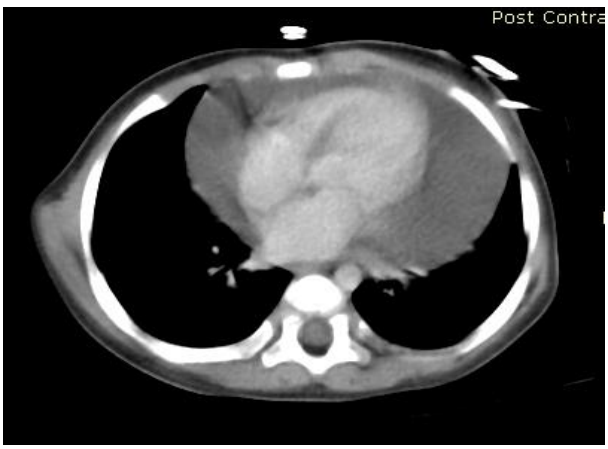

Fig. 3. CT scan showing significant pericardial collection

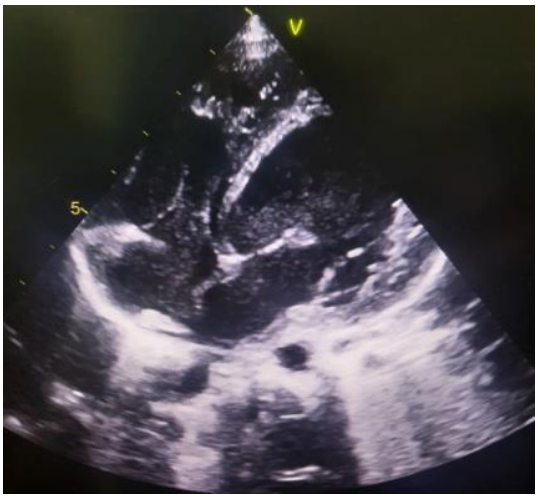

Fig. 4. ECHO at the time of discharge showing minimal pericardial collection

3. DISCUSSION

Primary chylopericardium is uncommon in children with few reported cases. Symptoms vary from asymptomatic to cough, dyspnea, chest pain, and fatigue [4]. The diagnosis can be challenging, as the appearance of pericardial fluid may resemble pyopericardium which was seen in our case. In the absence of clinical or laboratory parameter of sepsis, a high index of suspicion for chylous collection should be kept in mind. Key diagnostic indicators include a pericardial fluid triglyceride level above 500 mg/dL, a cholesterol/triglyceride ratio below 1, and lymphocyte predominance on cytologic examination [2]. Chest CT scans are used to exclude mediastinal masses or neoplasms. Lymphoscintigraphy and lymphangiography are primary diagnostic tools [3]. Lymphangiography is invasive and requires injection of contrast agent into the cannulated lymphatic vessel while lymphoscintigraphy is a noninvasive alternative. In our case these could not be done due to limitation of resources.

The pericardial space typically contains 25–35 ml of fluid which is produced by the pericardium and is similar to lymph. It is drained by lymphatic vessels of the heart into the left subclavian vein via the mediastinal lymph vessels, lymph nodes and the thoracic duct. The main lymphatic flow from the pericardium, pleura, entire right lung and the lower portion of the left lung meet at the bronchomediastinal lymphatics. Thus, regurgitation of lymphatic flow can occur simultaneously in multiple sites in the thoracic cavity, such as the pleura, lungs and pericardium and thus leading to chylothorax and chylopericardium [3].

Treatment for CP lacks consensus; initial management includes pericardiocentesis and a MCT diet [2,5]. Dietary restriction with a low-fat diet and MCT decreases the formation of lymph and absorption of these nutrients occurs via portal system thus bypassing the lymphatics. Medical management involves the use of sympathomimetic drugs such as octreotide which increase smooth muscle contraction and increase lymph drainage. The dose and duration of treatment with octreotide have varied in different case reports. CP is difficult to manage conservatively and it failed in 57.1% of reported cases [6]. Surgery is recommended when dietary modifications fail to bring any improvement even after 2-3 weeks [7]. Surgical management involves thoracic duct ligation, pericardiectomy, pericardial windows, and pericardial peritoneal shunt [8]. Thoracic duct ligation and pericardial window formation are believed to be the most effective procedures in cases of recurrences [6].

In our case VATS was performed as pericardial effusion was persisting despite 2 weeks of medical management and persistent drainage post-surgery prompted use of octreotide. The VATS procedure is associated with lesser postoperative morbidity [9]. Regular follow-up with echocardiography is crucial to monitor for constrictive pericarditis or recurrence [7].

4. CONCLUSION

Chylopericardium is a rare disease and could be misdiagnosed as pyopericardium. In the absence of clinical history, physical examination findings, or evidence supporting an infection, the diagnosis of pyopericardium should be reconsidered, with chylopericardium as a

differential diagnosis. Measuring triglyceride and cholesterol levels in pericardial fluid can confirm the diagnosis and facilitate prompt management. A combined approach consisting early surgical intervention, pharmacological therapy, and MCT diet replacement is suggested for effective treatment.

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

CONSENT

As per international standards, parental written consent has been collected and preserved by the author(s).

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of this manuscript.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/125596