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

ABSTRACT

Chylopericardium is a rare entity which is characterised by accumulation of chyle in pericardial sac. In pediatric age group chylopericardium can resemble pyopericardium. If suspected the diagnosis of chylopericardium could be easily made with fluid analysis. Here we are reporting a case of idiopathic chylopericardium initially diagnosed as pyopericardium and its successful management.

Keywords- chylopericardium, mimicking, triglyceride, pyopericardium, pediatric

INTRODUCTION

Chylopericardium (CP) is an exceedingly rare condition characterized by the accumulation of chylous fluid within the pericardium. It may be idiopathic, or primary, when no clear 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 these 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 misdiagnosed as pyopericardium and its successful management.

CASE REPORT –

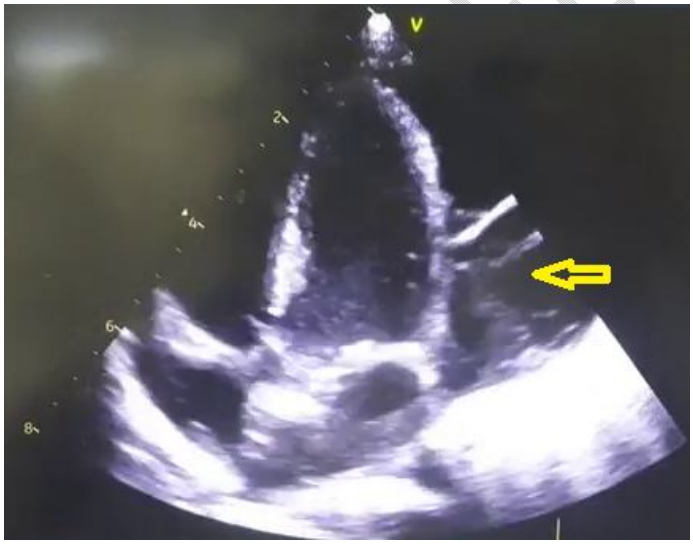
A 10-month-old male presented with shortness of breath and was found to have a massive right-sided pleural effusion on chest X-ray. Initially treated for empyema with intercostal drainage and antibiotics following which his symptoms improved. Cardiomegaly in chest X-ray prompted for Echocardiography

which revealed significant pericardial effusion. The child was then referred to our pediatric cardiology department for suspected pyopericardium management.

There was no history of fever, nor any history of trauma, surgery, tuberculosis contact or family medical issues. Physical examination revealed heart rate of 120/min, respiratory rate of 32/min, and blood pressure of 95/65 mmHg. The patient was alert without jugular venous distention , had clear lungs with equal bilateral airentry and a regular heart rhythm with distant heart sounds.The abdomen was soft without tenderness, hepatosplenomegaly or lymphadenopathy.

Chest X-ray confirmed an enlarged cardiac silhouette. Transthoracic echocardiography done at our centre 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.

Figure 1 – ECHO Image at the time of admission showing significant pericardial effusion



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.

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 diagnosisof pyopericardium wasreconsidered. 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.

Figure 2 – Pericardial fluid before(A) and after surgery(B)

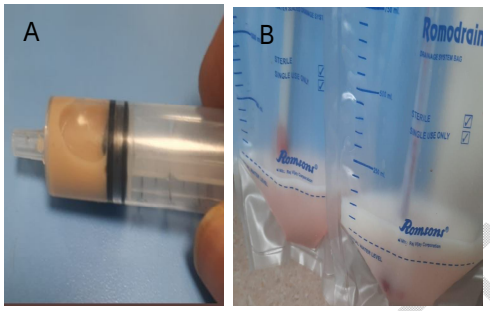
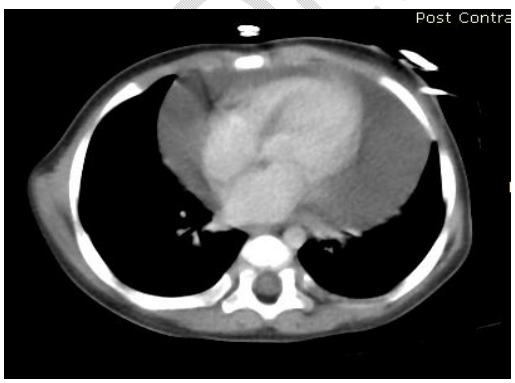


Figure 3 – CT scan showing significant pericardial collection

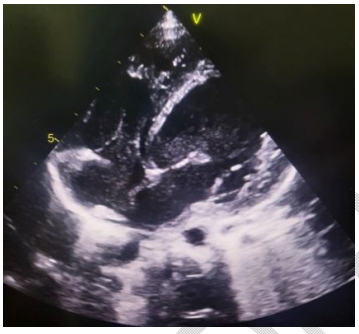


Despite minimal drainage after 48 hours and symptomatic improvement, echocardiography and CT scan revealed significant pericardial collection (Figure 3). No tumors or lymphangiomas were detected

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-op day and became serous in nature. Echocardiography showed minimal pericardial collection(Figure 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.

Figure 4 – ECHO at the time of discharge showing minimal pericardialcollection



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 absence ofclinical or lab 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 diagnostictools(3). Lymphangiography is invasive and requires injection of contrast agent into the cannulated lymphaticvessel whilelymphoscintigraphy 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 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 andthe 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 recommendedwhen dietary modificationsfail 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 pericardialwindow formation are believed to be the most effective procedures to prevent recurrence(6).

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

CONCLUSION

Chylopericardium is a rare disease and could be misdiagnosed as pyopericardium. In 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 cholesterollevels in pericardial fluid can confirm the diagnosis and facilitate prompt management. A combined approach consisting early surgicalintervention, pharmacological therapy, and MCT diet replacement is suggested for effective treatment.

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