

Case Report

Pediatric Chylopericardium: Treatment Conundrum

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ABSTRACT

Primary chylopericardium is a rare entity in the pediatric population, which is characterized by chyle accumulation in the pericardial sac. Treatment of this rare problem is a management dilemma as no definitive treatment has been found to be curative. We share our experience of treating chylopericardium with minimally invasive surgery.

KEYWORDS: *Chylopericardium, minimally invasive surgery, pediatric*

INTRODUCTION

Primary chylopericardium is rare entity in pediatric population, which is characterized by accumulation of chyle in pericardial sac. Treatment of this rare problem is a management dilemma as no definitive treatment has been found to be curative. We share our experience of treating chylopericardium with minimally invasive surgery.

CASE REPORTS

We present two children aged 2½ years (Case 1) and 4 years (Case 2). Both children presented with a complaint of gradually increasing breathlessness over 2–3 weeks. There was no history of congenital heart disease, trauma, or fever. Both children were well-nourished, active, and alert with tachycardia and tachypnea. On systemic examination, the chest was clear and heart sounds muffled. Both children had nontender hepatomegaly. Chest radiograph showed globular heart with a cardiothoracic ratio of 0.6 and 0.58, respectively. Two-dimensional (2D) echo confirmed pericardial effusion. Therapeutic and diagnostic pericardiocentesis was done which drained milky pericardial fluid. Cholesterol and triglyceride levels confirmed chylopericardium. Cultures of the pericardial fluid for bacteria and tuberculosis were negative, and cytology showed no tumor cells. Routine serology showed no signs of systemic inflammatory reaction. Chest computed tomography (CT) scan did not reveal mediastinal neoplasm or lymphadenopathy that might have obstructed the thoracic duct. Both children were started on low-fat diet with medium chain triglycerides (MCTs).

Case 1

The fluid reaccumulated over a period of 2 months following which surgical option was sought. Under general anesthesia, left thoracoscopy was done using three 5-mm ports. The pericardium identified, and a 3 cm × 2 cm left pericardiostomy was done which drained the chylous fluid. A 20 F intercostal drainage tube was placed in the left side of the chest. The tube drained for 4–5 days following which it was removed. The child was monitored with echocardiography. Follow-up echo was done after 15 days and 3 months and there was no reaccumulation of the fluid.

Case 2

The fluid reaccumulated 3 weeks after pericardiocentesis. A left thoracoscopy and window pericardiostomy were done similar to the first case. The child however had recurrence of chylopericardium 6 weeks after thoracoscopy. Following this, ligation of the thoracic duct and open pericardiostomy was planned. The child was prepared with a high-fat diet from the day before surgery. Epigastric incision was placed. The pericardium identified from the abdomen and tube pericardiostomy done. The thoracic duct was identified from the abdomen and ligated and cut. Postoperative period was uneventful. The pericardial tube drained very small chylous fluid for 3 days, which then stopped. The tube was removed after 5 days.

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Follow-up echo was done after 15 days and 3 months and there was no reaccumulation of the fluid.

DISCUSSION

Chylopericardium is a clinical condition where chyle accumulates in the pericardial cavity. It arises due to thoracic duct disruption or obstruction, with both inadequate collateral drainage and reflux of chyle through the lymphatics draining the heart and pericardium.^[1] The term “primary chylopericardium” was coined by Groves and Effler,^[2] who described a case of isolated accumulation of chyle in a 31-year-old woman with mediastinal cystic hygroma. The etiology of chylopericardium is lymphatic leakage and communication with the pericardial sac as concluded by CT, lymphangiography, and intraoperative thoracic ductogram.^[3] Other mechanisms such as damage to the thoracic duct valves, abnormal communication between the thoracic duct and the pericardial lymphatics, abnormally elevated pressure in the thoracic duct, and increased permeability of the lymphatics can also lead to chylopericardium. Primary idiopathic chylopericardium occurs in all age groups and affects both sexes equally.^[4] The clinical manifestations may vary from an absence of symptoms to signs of cardiac tamponade. According to Mask *et al.*, the most common presentations of patients with chylopericardium are a lack of symptoms, cough, dyspnea, and fatigue.^[5] Usually, X-ray chest and 2D echo help in arriving at diagnosis of pericardial effusion. However, examination of the pericardial fluid clinches the diagnosis of chylopericardium. A milky fluid that contains cholesterol and high levels of triglycerides confirms the diagnosis of chyle accumulation.^[5] The management of chylopericardium includes prevention of the mechanical (cardiac tamponade or constrictive pericarditis), metabolic and immune-mediated consequences of chylopericardium, to eliminate lymphatic fluid losses and to reduce recurrence. Initially, patients should be treated with nonsurgical measures, such as (1) pericardiocentesis; (2) pericardial drainage; (3) dietary support with a low-fat diet and initiation of MCTs, which are absorbed via the portal vein, rather than via the lymphatic vessels, or even with total parenteral nutrition and (4) pharmacological therapy with octreotide, which has been shown to reduce intestinal absorption of fats.^[6] Nonsurgical treatment of idiopathic chylopericardium is usually not satisfactory, and a failure rate of 57%–60% has been reported.^[7,8] Han *et al.* reported that medical therapy should not be continued for longer than 3 weeks.^[9] If medical therapy proves to be ineffective, then surgical treatment should be considered, even in asymptomatic patients, to avoid subsequent progression to cardiac tamponade or constrictive pericarditis.^[10]

There are different approaches to the surgical treatment of chylopericardium: (1) pericardiectomy, (2) pericardial window formation and (3) ligation of the thoracic duct above the level of the diaphragm. Thoracic duct ligation and pericardial window formation are believed to be the most effective procedures to prevent recurrence.^[7,9] In our experience, we tried pericardial window formation in both patients; however, first patient showed resolution whereas second patient required ligation of thoracic duct above diaphragm. Looking at the rarity of this pathology and the unpredictable outcomes, a fixed management protocol cannot be determined. However, a thoracoscopic pericardiostomy should be the first option when surgical intervention is required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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