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European Journal of Cardio-thoracic Surgery 12 (1997) 319–321

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

Case report

Isolated primary chylopericardium

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Received 10 February 1997; accepted 8 April 1997

Abstract

Isolated primary chylopericardum is known to be a rare clinical entity. A 17-year-old girl was diagnosed as isolated primary chylopericardium. She was unresponsive to conservative treatment with pericardial tube drainage and medium chain triglyceride diet. At 2 weeks after the conservative treatment, ligation and resection of the thoracic duct with establishment of a pericardial window through a left thoracotomy was performed. At 6 months, follow-up showed no accumulation of the pericardial fluid. This case also supports that ligation and resection of the thoracic duct with establishment of a pericardial window is the treatment of choice in isolated primary chylopericardium. © 1997 Elsevier Science B.V.

Keywords: Chylopericardium; Primary

1. Introduction

Isolated primary chylopericardium is a rare entity in which chylous fluid accumulates in the pericardial cavity. In 1888 Hasebrock was the first to describe isolated primary chylopericardium and 'primary chylopericardium' was first reported by Groves and Effler in 1954 [1]. Although several reviews on the subject have been published recently [2,3], the cause remains unclarified. In this case, isolated primary chylopericardium in a 17-year-old girl is presented who was treated with ligation and resection of the thoracic duct with establishment of a pericardial window through a left thoracotomy.

2. Case report

A 17-year-old girl was admitted for easy fatigability and with the diagnosis of cardiomegaly on a chest X-ray. There was no apparent history of trauma, infection, radiotherapy or mediastinal neoplasm. The physical examination revealed a healthy appearing girl with normal physical examination findings except distant heart sounds. All laboratory findings were within normal range.

Her echocardiogram (Fig. 1) showed a large pericardial effusion. The diagnosis was established, after aspiration of 450-ml milky-colored chylous fluid obtained by pericardiocentesis. Cholesterol to triglyceride ratio of the fluid was 0.19. The sediment contained white blood cells. The fluid was sterile on culture and the cytological examination was normal. Sudan III staining of the fluid revealed the presence of fat globules. Computed Tomography (CT) of the thorax showed a large pericardial effusion. Unsuccessful lymphangiogram was attempted.

Through a subxyphoid approach, a pericardial tube was placed under general anesthesia. Meanwhile, the patient was placed on a high content of medium-chain triglyceride diet. Nevertheless the drainage continued for nearly 150 ml of chylous fluid per day. After 2 weeks, thoracic duct ligation was planned. Before oper-

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PII S1010-7940(97)00126-7

ation 250 ml of olive oil was infused into the stomach. After standard left thoracotomy, the pericardial sac was found to be distended and filled with 200 ml of chylous effusion. Following mobilization of the descending aorta, the thoracic duct and surrounding lymphatics were observed. No abnormality was found (Fig. 2), thereafter supradiaphragmatic ligation and resection of the thoracic duct and the concomitant fashioning of a pericardial window were performed. The postoperative course was uneventful and there is no reaccumulation of the pericardial fluid at 6 months follow-up.

3. Discussion

The accumulation of chyle in the pericardial sac can occur as a primary event or secondary to chest trauma, operative procedures, radiotherapy, thrombosis of subclavian vein, infections (e.g. tuberculosis) and neoplasm of the mediastinum (Hodgkin's and non-Hodgkin's lymphoma) [1,4]. If none of the mentioned causes can be found the disease is referred to as 'primary chylopericardium'. To our knowledge, there have been only 89 cases including our case. It has been reported that reflux of chylous fluid into the pericardial sac is the main mechanism involved [2]. There are a couple of possible explanations for the cause of this disorder: (1) the presence of both damaged valves of the thoracic duct and its communication to the pericardial lymphatics resulting in chylous reflux and (2) abnormally elevated pressure in the thoracic duct observed as lymphangiectasia. As it was pointed out by Akamatsu's review [2], most of the patients were young and were asymptomatic or were noted as having only cardiomegaly by chest radiography. Some patients had slight dyspnea (35%), cough (10%), and easy fatigability (4%).

Diagnostic modalities essential to the evaluation of suspected chylopericardium include the following [3]:

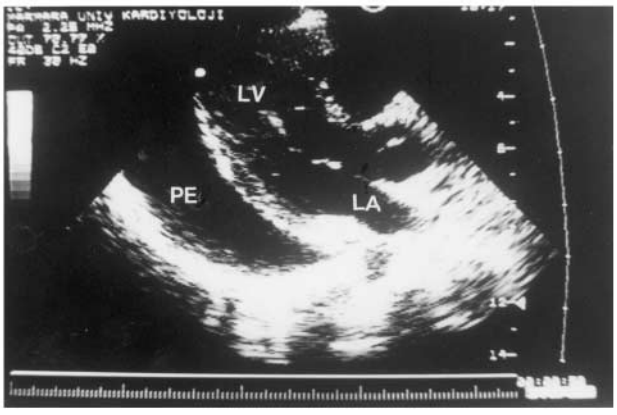

Fig. 1. Preoperative echocardiogram of the patient shows a large pericardial effusion (PE). (LV = Left Ventricle, LA = Left Atrium).

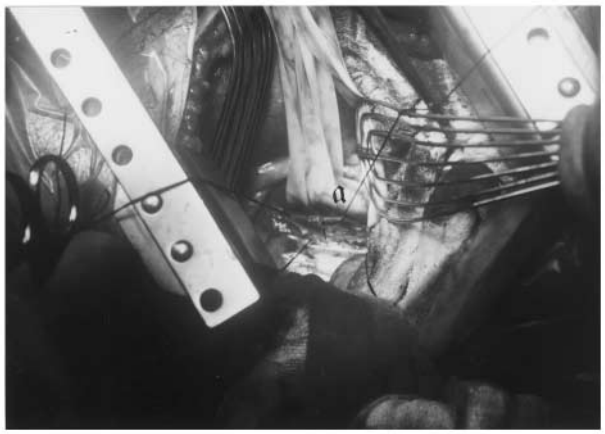

Fig. 2. Operative photograph of the patient. The thoracic duct (arrow) appeared after taping of the descending aorta (a).

(1) Chest radiography; (2) Echocardiograph; (3) CT of the chest (4) pericardiocentesis; (5) lymphangiography; (6) oral administration triolein labeled with iodine; and (7) a combination of lymphangiograph and computed tomography [5].

There is no agreement in the literature as to the amount of daily drainage that should be used as a criteria for operative intervention. Since the loss of lymph fluid may lead to a number of complications, such as compromise of nutritional and immunogenic status and impingement and aberration of imposing structures and their function [3] and risk of cardiac tamponade or constrictive pericarditis [6], it is our opinion that conservative management with a medium-chain triglyceride diet or parenteral nutrition supplemented by tube drainage may be excluded in management of the primary chylopericardium, except for patients who are unsuitable for surgical treatment [6]. We agree with other authors [2,4] that the certain treatment for the disease is ligation and resection of the thoracic duct with establishment of a pericardial window through a right or left thoracotomy. We performed left thoracotomy and we found this approach easy and less invasive to reach the thoracic duct and establish a larger pericardial window. Recently, thoracoscopic 'mass ligature' for isolated primary chylopericardium was reported by Furrer [7]. Due to occurence of pericardial effusion 1 month after the surgery, use of video-assisted thoracoscopic surgery in the management of isolated primary chylopericardium should be well documented. Giving the patient 100 to 200 ml of olive oil before the operation causes filling of the duct with milky chyle, which is readily recognized throughout the course of the operation [8]. After the operation, long-term follow-up is suggested [9].

In conclusion, primary chylopericardium is a rare disease for which ligation and resection of the thoracic duct with establishment of a pericardial window is the treatment of choice.

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