Journal of Cardiology Cases (2011) 3, e106–e110

available at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/jccase

Case Report

Primary chylopericardium treated by surgery: Report of two cases

Hidetoshi Hattori (MD)[a,*], Eijirou Dakeshita (MD)[b], Jun Nakazato (MD)[a], Takanori Takahashi (MD)[a], Minoru Wake (MD)[a], Kazuhito Hirata (MD)[a], Hiroshi Yasumoto (MD)[c], Toshiho Tengan (MD)[c], Hidemitsu Mototake (MD)[c]

[a] *Division of Cardiology, Okinawa Chubu Hospital, Okinawa, Japan*
[b] *Division of Thoracic Surgery, Okinawa Chubu Hospital, Okinawa, Japan*
[c] *Division of Cardiovascular Surgery, Okinawa Chubu Hospital, Okinawa, Japan*

Received 25 October 2010; received in revised form 12 December 2010; accepted 14 December 2010

KEYWORDS
Pericardial effusion;
Primary chylopericardium;
Video-assisted thoracic surgery (VATS)

Summary Primary chylopericardium is a rare condition. The etiology and the treatment remain unclear. We report two cases of primary chylopericardium successfully treated by surgery. Both cases were asymptomatic young women and were found to have cardiomegaly on chest X-ray at a routine annual health examination. An echocardiography demonstrated massive pericardial effusion and chylous fluid was obtained with pericardiocentesis. Lymphoscintigraphy demonstrated abnormal communication between the pericardial sac and the thoracic duct. Because of reaccumulation of chylous pericardial effusion after conservative treatment, we performed surgical ligation of thoracic duct and partial pericardectomy by video-assisted thoracic surgery (VATS) in one case and by thoracotomy in another case. After surgery, both patients are doing well without recurrence of pericardial effusion. Surgical treatment including VATS is effective and should be performed in case of primary chylopericardium.

Introduction

Primary or idiopathic chylopericardium is accumulation of chylous fluid in the pericardial space without any apparent cause. It is a rare clinical entity and etiology remains obscure. Optimal treatment of chylopericardium remains controversial. Here we report two cases of primary idiopathic chylopericardium successfully treated by surgery.

Case report

Case 1: A 15-year-old female was admitted to our hospital for evaluation of asymptomatic massive pericardial effusion. She had been previously healthy and a chest X-ray at the time of routine annual health examination showed enlargement of the cardiac silhouette (Fig. 1A). An echocar-

* Corresponding author at: Department of Cardiology, Tokyo Women's Medical University, 8-1, Kawada-cho, Shinjuku-ku, Tokyo 162-8666, Japan. Tel.: +81 3 3353 8111; fax: +81 3 3356 0414.
E-mail address: hattori-hide@mqb.biglobe.ne.jp (H. Hattori).

1878-5409/$ – see front matter © 2010 Japanese College of Cardiology. Published by Elsevier Ireland Ltd. All rights reserved.
doi:10.1016/j.jccase.2010.12.004

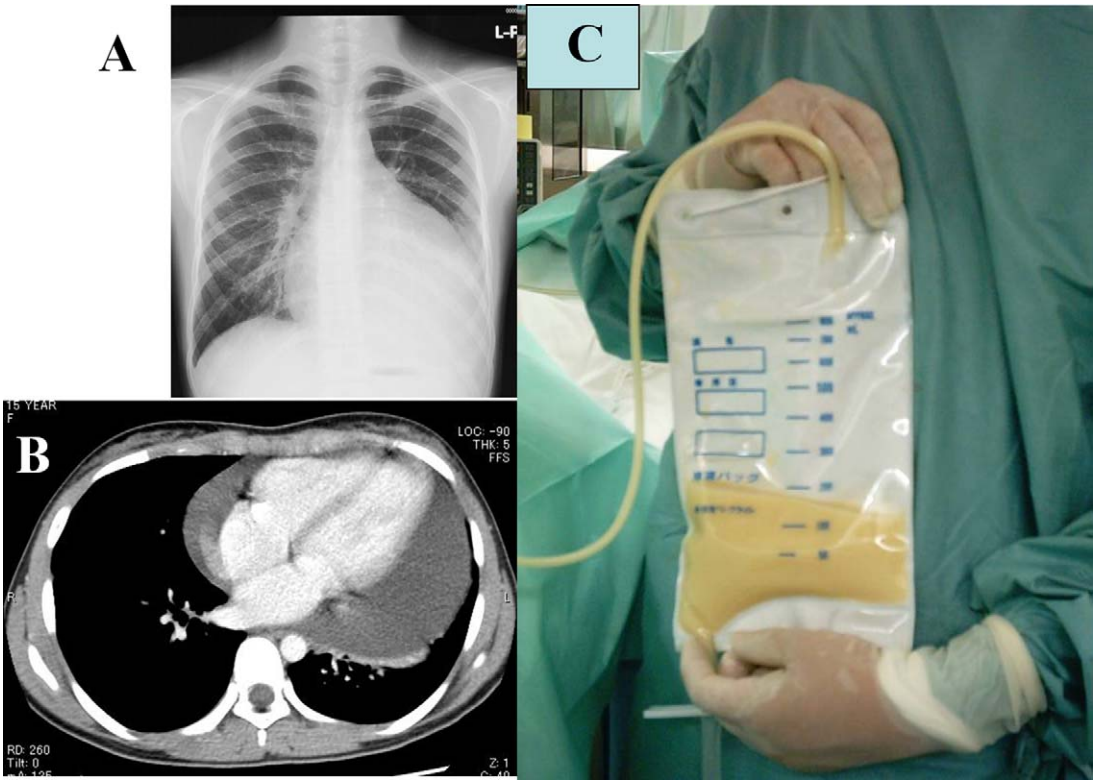

Figure 1 (A) A chest X-ray shows an enlarged cardiac silhouette. (B) Computed tomography demonstrates massive pericardial effusion. (C) 1100 ml of chylous fluid was drained.

diography and a computed tomography (CT) scan (Fig. 1B) revealed massive pericardial effusion. There was no past history of tuberculosis, trauma, or surgery. Her blood pressure was 118/70 mm Hg and the pulse rate was 80 beats per minutes. Cardiovascular physical findings were not remarkable and there were no signs of cardiac tamponade. Lipid profile demonstrated triglycerides of 32 mg/dl, total cholesterol of 188 mg/dl, low density lipoprotein (LDL)-cholesterol of 110 mg/dl, and high density lipoprotein (HDL)-cholesterol of 72 mg/dl. Blood count, electrolytes, kidney function, and liver function tests were normal.

After admission, pericardiocentesis was performed; 1100 ml of thick milky chylous fluid (Fig. 1C) was drained. Chemistry of the fluid showed increased triglycerides (specific gravity 1.034; total cholesterol 130 mg/dl; triglycerides 1600 mg/dl). Cultures for bacteria or acid-fast bacilli were negative. A cytology examination revealed no evidence of malignant cells. Since the fluid proved to be chyle, we performed ^{123}I-beta-methyl iodophenyl pentadecanoic acid (^{123}I-BMIPP) lymphoscintigraphy. Lymphoscintigraphy showed abnormal accumulation of ^{123}I-BMIPP in the pericardial space through the thoracic duct, which represented abnormal communication between the pericardial sac and the thoracic duct (Fig. 2A).

Because of the recurrence of the pericardial effusion after removal of the pericardial catheter, surgical treatment with ligation of the thoracic duct and partial pericardectomy (pericardial window) were performed with video-assisted thoracic surgery (VATS). Intraoperative thoracic duct angiography (Fig. 2B) confirmed abnormal communication between thoracic duct and pericardium. A ligation of the thoracic duct just above the diaphragm was successfully performed with VATS. The postoperative course was uneventful and she was discharged on the 7th post-operative day. She is well 1 year after operation with no signs of recurrence.

Case 2: A 30-year-old asymptomatic woman was hospitalized for evaluation of massive pericardial effusion (Fig. 3A and B). Her blood pressure was 100/60 mm Hg and the pulse rate was 84 beats per minutes. The findings of physical examination were not remarkable. Routine laboratory tests including lipid profile were all within normal limits (triglycerides 101 mg/dl; total cholesterol 124 mg/dl; LDL-cholesterol 60 mg/dl; HDL-cholesterol 44 mg/dl).

After admission, pericardiocentesis was performed and 800 ml of chylous fluid was obtained (Fig. 3C). The chemistry of the effusion revealed high concentration of triglyceride (specific gravity 1.036; total cholesterol 106 mg/dl; triglycerides 3718 mg/dl). Bacteriological examination of the fluid was negative. ^{123}I-BMIPP lymphoscintigraphy showed abnormal accumulation of ^{123}I-BMIPP in the pericardial sac (Fig. 4A). Because of the possibility of unreliable effect with conservative management, surgical treatment was performed. First we tried to perform ligation of the thoracic duct and partial pericardiectomy by VATS. However, because of the difficulty in identifying thoracic duct, direct visualization under thoracotomy had to be done. Intraoperative thoracic duct angiography clearly revealed obstruction of the thoracic duct and abnormal communication to the pericardial space (Fig. 4B). A ligation of the thoracic duct and pericardial window were successfully performed. She was discharged on the 6th post-operative day without any complication. She has had no recurrence at 12 months follow-up.

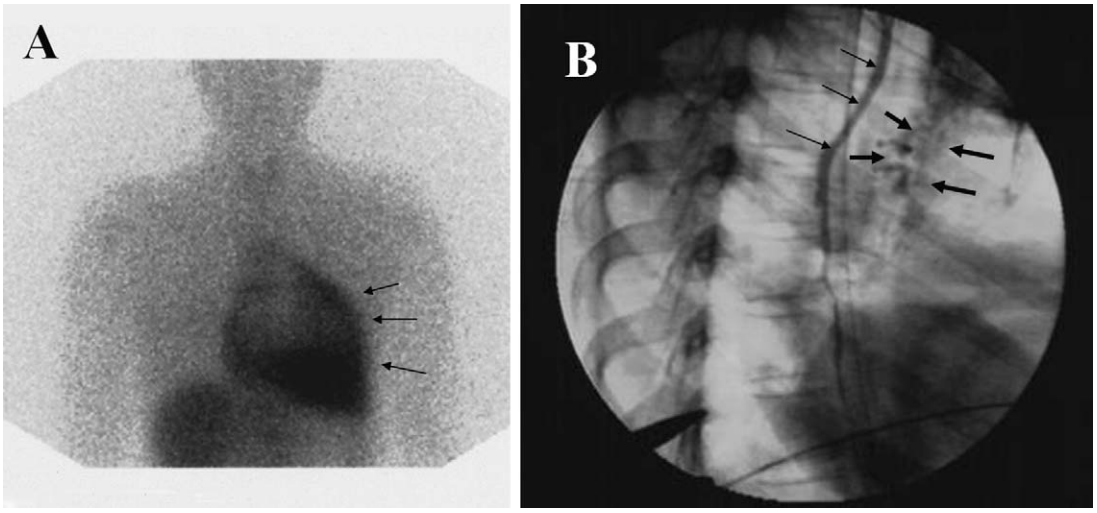

Figure 2 (A) ^{123}I-BMIPP lymphoscintigraphy taken 24 h after oral administration of ^{123}I-BMIPP. Arrows indicate accumulation of ^{123}I-BMIPP in the pericardial sac. (B) Intraoperative thoracic duct angiography shows abnormal reflux of the contrast dye into the pericardium suggesting obstruction of the thoracic duct. Black arrows indicate the thoracic duct and large black arrows indicate abnormal connection into the pericardium.

Discussion

Chylopericardium is abnormal accumulation of chylous fluid in the pericardial sac. It usually occurs after thoracic or cardiac surgery or may be associated with chest trauma, mediastinal tumor, infection, and congenital lymphatic abnormalities [1]. Sagristà-Sauleda et al. reported only one of 461 patients with large pericardial effusion had chylopericardium [2]. Primary or idiopathic chylopericardium is diagnosed in the absence of any apparent precipitat-

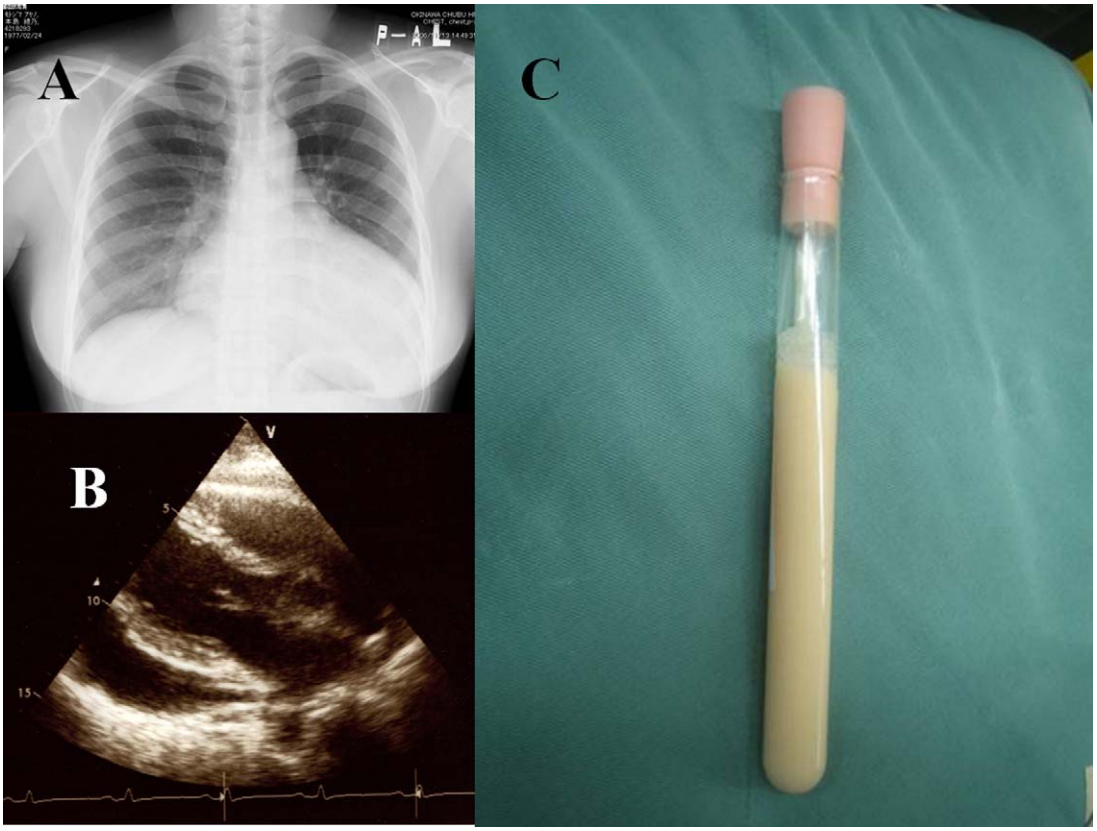

Figure 3 (A) Chest X-ray shows marked cardiac enlargement. (B) An echocardiography demonstrates large amount of pericardial fluid. (C) Chylous fluid obtained by pericardiocentesis.

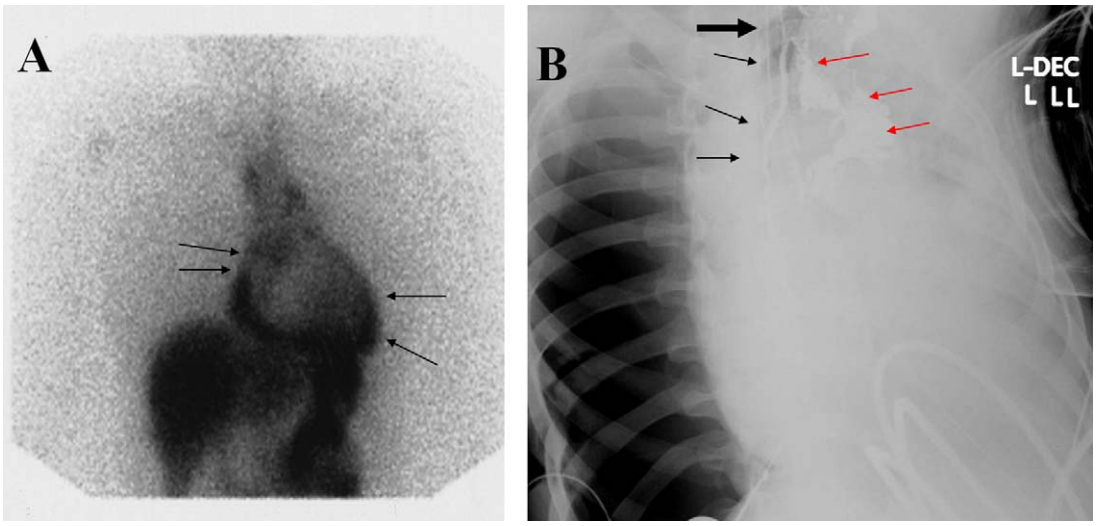

Figure 4 (A) ^{123}I-BMIPP lymphoscintigraphy shows accumulation of ^{123}I-BMIPP in the pericardial sac (arrows). (B) Intraoperative thoracic duct angiography demonstrated abnormal reflux of the contrast dye into the pericardium. Black arrows indicate the thoracic duct. A large black arrow indicates the obstruction of the thoracic duct. Red arrows indicate abnormal reflux of the contrast dye through lymphatic channels.

ing factor [3]. Primary chylopericardium was first reported by Groves and Effler [3]. Since then 114 cases have been reported up to 2007 [4].

Primary chylopericardium occurs in all ages but usually in children or young adults with equal frequency in males and females. Symptoms at presentation may vary such as dyspnea, chest pain, and cough. Some patients remain asymptomatic like our cases. Patients with cardiac tamponade are rare [5].

The underlying pathophysiology remains uncertain. Fatty acids absorbed in the gastrointestinal tract are incorporated into chylomicrons and ultimately reach the left subclavian vein through the thoracic duct. The thoracic duct is indirectly connected to pericardial cavity through mediastinal lymph node. Volume of chyle ranges between 1500 and 2400 ml daily and flow rate varies. A fatty meal provides the maximal stimulation for chyle production. Pathophysiolosy of chylopericardium was firstly proposed by Dunn [6]. The thoracic duct obstruction and abnormal reflux of chyle with elevated pressure in the thoracic duct cause accumulation of chylous fluid in the pericardium. Other lymphatic malformations such as lymphatic valvular incompetence, lymphangiectasia could lead to the accumulaton of chyle in the pericardium [7]. Also a recent report described the theory that accumulation of chyle is due to slow transudation of fluid from small, pathologic lymphatic channel rather than through direct communication with a main thoracic duct [8]. Itkin et al. demonstrated occlusion of the thoracic duct and multiple lymphatic collaterals by CT lymphangiography [8]. Intraoperative thoracic duct angiography in our cases demonstrated obstruction of the thoracic duct and reflux of the contrast dye through abnormal lymphatic channels connecting into pericardium (Figs. 2B and 4B). The cause of obstruction of the thoracic duct could not be identified in either of our cases. Slow speed of accumulation may explain the absence of signs and symptoms of cardiac tamponade.

Many diagnostic modalities have been reported, but lymphangiography or lymphoscintigraphy is used in most cases to detect the abnormal connection between the thoracic duct and the pericardium. Lymphangiography implies direct administration of contrast agent into cannulated lymph vessels. A combination of lymphangiography and CT is useful for detecting lymphatic vessels and their relationships to pericardial cavity [8]. Lymphoscintigraphy is an imaging technique which utilizes radionuclides as contrast agent. Lymphoschintigraphy may be an alternative to lymphangiography as it is a faster and less invasive procedure, although lymphangiography is useful for delineating the anatomy of the thoracic duct. In our patients, we performed lymphoscintigraphy using orally administered ^{123}I-BMIPP, which is generally absorbed from the intestine and reaches the venous circulation through the thoracic duct [9]. In both of our cases, lymphoscintigraphy was very useful to prove abnormal accumulation of ^{123}I-BMIPP in the pericardium. Oral administration of I-triolein is also useful for detecting abnormal communication between the pericardial sac and the thoracic duct [10].

Optimal treatment for primary chylopericardium is not well established. Current treatment options are conservative therapy including total parenteral nutrition, a low-fat diet with medium-chain triglycerides or surgical exploration including thoracic duct ligation, pericardial-peritoneal shunts and pericardectomy. Conservative treatment failed in 61% of the cases reported and single pericardiocentesis often leads to reaccumulation of chylous fluid [5]. As these data show, single pericardiocentesis did not prevent recurrence in case 1. The precise natural history of primary chylopericardium is difficult to establish, but later progression of cardiac tamponade or constrictive pericarditis is reported when pericardial effusion persists [10]. In addition, a continuing leakage of chyle can lead to serious metabolic, nutritional, and immunologic abnormalities [5]. Because of the high probability of recurrence with conservative treatment, the efficiency of surgical treatment and avoiding later complications, we believe surgical treatment should be considered if there is no contraindication to surgery.

Surgical exploration includes thoracic duct ligation and partial pericardectomy for complete drainage and prevention of constrictive pericarditis. Mass ligation just above the diaphragm is recommended, because anatomic variations of the thoracic duct are common. Partial pericardectomy alone is associated with reaccumulation of chylous fluid in some cases [11]. Recently, VATS has been introduced for treatment. Furrer et al. first reported the successful treatment of primary chylopericardium by VATS [12]. The VATS procedure is being used increasingly, but VATS or thorocotomy depends on the local expertise.

In summary, we present two cases of primary chylopericardium. The surgical treatment including thoracic duct ligation and pericardectomy was effective for the primary chylopericardium in these two cases.

References

[1] Campbell RM, Benson LN, Williams WW, Adatia IA. Chylopericardium after cardiac operations in children. Ann Thorac Surg 2001;72:193—6.
[2] Sagristà-Sauleda J, Angel J, Permanyer-Miralda G, Soler-Soler J. Long-term follow-up of idiopathic chronic pericardial effusion. N Engl J Med 1999;341:2054—9.
[3] Groves LK, Effler DB. Primary chylopericardium. N Engl J Med 1954;250:520—3.
[4] Silva MA, Martins AS, Campos NL, Andrade RR, Tohi LM, Hueb JC. Primary idiopathic chylopericardium. Arq Bras Cardiol 2009;92:40—3.
[5] Dib C, Tajik AJ, Park S, Kheir ME, Khandieria B, Mookadam F. Chylopericardium in adults: a literature review over the past decade (1996—2006). J Thorac Cardiovasc Surg 2008;196:650—6.
[6] Dunn RP. Primary chylopericardium, a review of the literature and an illustrated case. Am Heart J 1975;79:369—77.
[7] Akamatsu H, Amano J, Sakamato T, Suzuki A. Primary chylopericardium. Ann Thorac Surg 1994;58:262—6.
[8] Itkin M, Swe NM, Shapiro SE, Shrager JB. Spontaneous chylopericardium: delineation of the underlying anatomic pathology by CT lymphagiography. Ann Thorac Surg 2009;87:1595—7.
[9] Qureshy A, Kubota K, Ono S, Sato T, Fukuda H. Thoracic duct scintigraphy by orally administered I-123 BMIPP: normal findings and a case report. Clin Nucl Med 2001;26:847—55.
[10] Morishita Y, Taira A, Furoi A, Arima S, Tanaka H. Constrictive pericarditis secondary to primary chylopericardium. Am Heart J 1985;109:373—5.
[11] Yüksel M, Yildizeli B, Zonüzi F, Batirel HF. Isolated primary chylopericardium. Eur J Cardiothorac Surg 1997;12:319—21.
[12] Furrer M, Holp M, Ris HB. Isolated primary chylopericardium: treatment by thorascoscopic thoracic duct ligation and pericardial fenestration. J Thorac Cardiovasc Surg 1996;112:1120—1.