

Chylopericardium Secondary to Lymphangiomyoma

— A case report —

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Chylopericardium is a rare disease entity characterized by the accumulation of chylous fluid in the pericardial sac. It usually arises from mediastinal neoplasms, thrombosis of the subclavian vein, tuberculosis, nonsurgical trauma, thoracic or cardiac surgery. The spectrum of symptoms for chylopericardium varies from an incidental finding of cardiomegaly to dyspnea, upper abdominal discomfort, cough, chest pain, palpitation, fatigue. However, most of the patients are asymptomatic. The main purpose of treatment of chylopericardium is the prevention of cardiac tamponade and prevention of metabolic, nutritional, and immunological compromise due to chyle leak. Here, we report a case of chylopericardium secondary to lymphangiomyoma with review of the literature.

Key words: 1. Thorax
2. Chylopericardium
3. Lymphangioma
4. Secondary

CASE REPORT

A 37-year-old woman visited the Division of Cardiology, Department of Internal Medicine with an incidental finding of cardiomegaly during a routine screening test at her workplace. The vital signs were stable with a heart rate of 80/min, systolic blood pressure of 100 mmHg, respiratory rate of 18/min, body temperature of 37.0°C. Echocardiography showed an ejection fraction of 76.4% with normal cardiac structures including valves. However, it showed a moderate pericardial effusion, and the pericardial effusion size was measured at five different points. The diameters were 0.9 cm at the anterior site of the right ventricle, 1.5 cm at the posterior site of the left ventricle, 0.4 cm at the lateral site of the

left ventricle, 0.6 cm at the right atrial site, 1.7 cm at the apex site which was the largest. Other laboratory examination findings were within their normal ranges. The initial diagnosis was pericarditis with pericardial effusion. It was decided to treat the patient with oral diuretics. After 3 months, chest X-ray did not show any improvement in cardiac size, and hence chest X-ray was followed by chest CT (computed tomography). A cystic mass in the right cardiophrenic angle having homogeneous low attenuation and pericardial effusion without definite pericardial thickening or enhancement were detected (Fig. 1). The woman was transferred to the Department of Cardiothoracic Surgery for surgical resection of the cystic mass and confirmation of the diagnosis. The operation was performed by videoscopically assisted right thoracotomy

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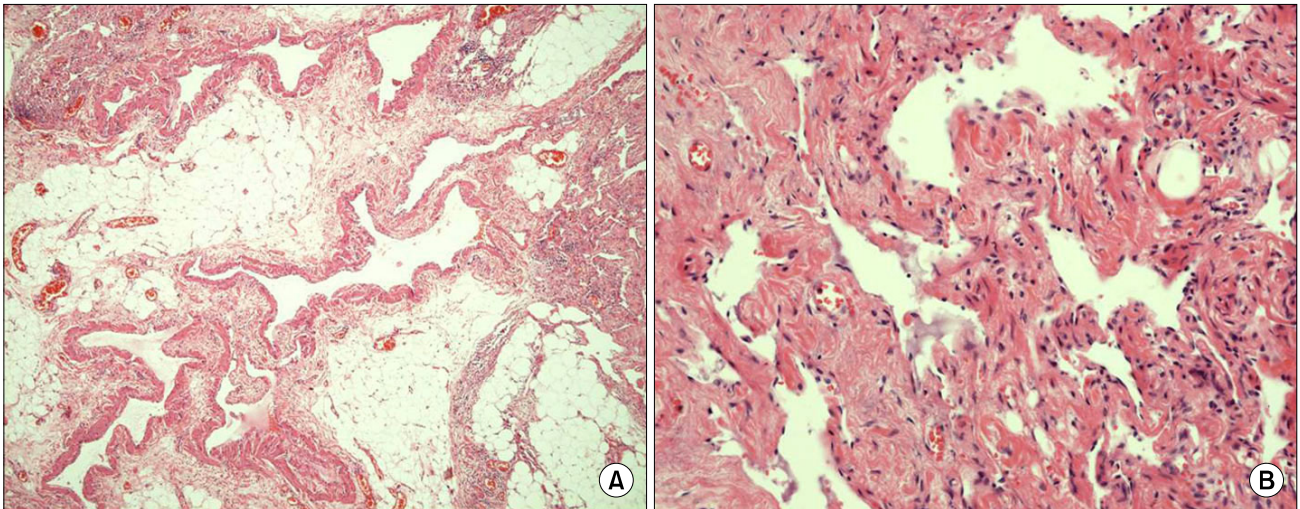


Fig. 2. (A) The mass is composed of slit-like vascular spaces and remaining lymphoid tissue on low-power view. (B) There are lymphoid aggregations suggesting pre-existing lymphoid accumulations (H&E stain, $\times 10$).

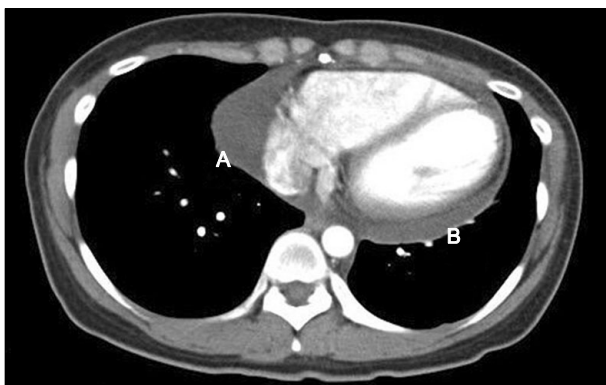


Fig. 1. (A) Cystic mass in the right cardiophrenic angle with homogeneous low attenuation. (B) Pericardial effusion without definite pericardial thickening or enhancement.

through three trocar holes. The cystic mass was removed and pericardiotomy and pericardial biopsy were performed. After pericardiotomy, massive amount of milky fluid about 950 cc was drained. A sample of the milky fluid was sent for laboratory examination, and pericardial tissue sample was obtained for pathologic examination. After sampling and biopsy, the pericardial sac was cleansed with normal saline irrigation. Wounds due to trocar insertion were closed layer by layer, after positioning one chest tube into the pericardial sac, and the other into the thoracic cavity. Laboratory examination findings of the milky fluid were as follows: pH 7.5, RBC $6,900/\text{mm}^3$, WBC $9,700/\text{mm}^3$, 90% lymphocyte dominance in

the differential WBC count, blood glucose 83 mg/dL, LDH 164 U/L, albumin 3.5 gm/dL, triglycerides 1,508 mg/dL. The pathologic report of the cystic mass was lymphangiomyoma and that of the pericardial tissue sample was chronic inflammation (Fig. 2). Based on the laboratory examination findings and pathologic report, we confirmed the diagnosis of chylopericardium due to lymphangiomyoma. The chest tube was removed from the thoracic cavity on the 13th day after surgery and from the pericardial sac on the 14th day after surgery. The patient was discharged on the 15th day after surgery, and was followed up in the outpatient department. At the 3-month and 1-year follow-up, there were neither any signs of cardiomegaly nor of pericardial effusion.

DISCUSSION

Chylopericardium is a rare disease entity characterized by the accumulation of chylous fluid in the pericardial sac. Since Hazebrock first described the term, chylopericardium with chylous fluid in the pericardial sac in 1888, and Groves & Effler used the term primary idiopathic chylopericardium in 1954, many suggestions about the etiology of chylopericardium have been presented. However, the definite cause of chylopericardium has not been established [1]. Chylopericardium usually arises from mediastinal neoplasms, thrombosis of the subclavian vein, tuberculosis, nonsurgical trauma,

thoracic or cardiac surgery [1]. The main mechanism responsible for the development of chylopericardium is similar to that in chylothorax, i.e. the mechanical obstruction of the thoracic duct or the disturbances in the drainage of chyle into the subclavian vein. The spectrum of symptoms for chylopericardium varies from an incidental finding of cardiomegaly to dyspnea, upper abdominal discomfort, cough, chest pain, palpitation, fatigue. However, most of the patients are asymptomatic [2]. The diagnosis of chylopericardium is made as follows: most cases show cardiomegaly on chest X-ray, and further evaluation could be performed. The presence of pericardial effusion is usually diagnosed with echocardiography or computed tomography. If a pericardial effusion is detected, the characteristics of the pericardial effusion should be analyzed with pericardiocentesis. If the macroscopic examination of the pericardial effusion shows the presence of a milky fluid and the laboratory examination of the milky fluid shows a high level of triglycerides, the diagnosis of chylopericardium should be made. The main differential diagnosis of pericardial effusion is cholesterol pericarditis, in which the pericardial fluid contains cholesterol crystals, and the macroscopic examination mimics that of chylopericardium [3]. The characteristics of the fluid are milky-white appearance which clears promptly after addition of ether, presence of fat droplets demonstrated by a Sudan III stain, relatively high triglyceride content compared with cholesterol, high protein content and predominance of the lymphocytes [2]. If a diagnosis of chylopericardium is made, further evaluation should be carried out to detect any communication between the lymphatic channels and the pericardium [2]. Direct lymphangiography and radio-nuclide lymphangiography should be carried out in such cases, however no communication between the lymphatic channels and the pericardium is detected in most cases. If there is any communication between the lymphatic channels and the pericardium, it could demonstrate a direct communication between the pericardial sac and the lymphatic channels [4] or show pooling of contrast in the pericardial sac [5]. The presence of communication could exclude any mechanical obstruction. This can be very useful in planning an operation such as thoracic duct ligation. The main purpose of treatment of chylopericardium are the prevention of cardiac tamponade and prevention of metabolic, nutritional, and immunological

compromise due to chyle leak [2]. The therapeutic modalities are as follows; pericardiocentesis for both the diagnostic and therapeutic purposes, pericardial window formation, pericardiectomy, thoracostomy drainage, ligation of or resection of the thoracic duct in the lower part of the chest, dietary support with medium or short-chain triglycerides and low fat meals [6], pericardial-peritoneal shunt [7]. Although, chylopericardium does not respond well to the conservative treatment, and surgical treatment is required in most cases, Szabados et al. reported a case treated with conservative treatment using octreotide [8]. In this case, chylopericardium was associated with lymphangiomyoma. Lymphangiomyoma is characterized by a proliferation of lymph vessels and smooth muscle elements. The accumulation of chylous fluid in the pericardial sac is considered to be the result of abnormal proliferation of the lymphatic vessels due to lymphangiomyoma in the pericardium. After surgical resection of the lymphangiomyoma, chest X-ray showed no cardiomegaly. If any changes may occur during follow-up, an operation such as thoracic duct ligation can be considered.

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