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Case Report

Recurrent Primary Chylopericardium: An Unusual Cause of Cardiac Tamponade

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Abstract: Chylopericardium is a rare clinical entity in which chylous fluid accumulates in the pericardial cavity. We report a case of recurrent massive idiopathic chylopericardium with tamponade in a 3-year-old infant with no history of trauma, thoracic surgery, malignancy, infection or tuberculosis. Echocardiography showed a large amount of pericardial effusions with evidence of tamponade. And 450 mL of fluid was evacuated by pericardiocentesis. She successfully responded to 30 days of continuous pericardial drainage and 15 days of a medium-chain triglyceride diet. We conclude that if a patient is asymptomatic and can well tolerate daily life, surgery including surgical ligation of the thoracic duct and creation of a pericardial window is not necessarily required**.**

Keywords: Cardiac tamponade; chylous pericarditis; pericardial effusion; conservative treatment.

INTRODUCTION

Idiopathic chylopericardium is the accumulation of chylous fluid in the pericardial space in the absence of any apparent precipitating factor. In recent years, a few cases of idiopathic chylopericardium have been described wherein the lymphatic leak and the interconnections with the pericardial sac were visualized by lymphoscintigraphy [1]. We report a case of primary idiopathic chylopericardium in a 3-year-old infant successfully treated without surgical procedures. This is the rare case we have found in the literature of primary idiopathic chylopericardium resolved without surgery.

CASE REPORT

A 3-year-old infant was admitted to our cardio-pediatric department 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. She was asymptomatic except for some fatigability. Physical examination was unremarkable. Electrocardiogram showed only low QRS voltages. Echocardiography revealed large pericardial effusion with right atrial and ventricular collapse (fig. 1). Pericardiocentesis was performed and 1100 mL of milky fluid were removed, suggesting chylopericardium (fig 2A). The chylous nature of the fluid was confirmed by high content of triglycerides (1200 mg/dL) and by cholesterol/triglyceride ratio <1. Sudan III stain of the fluid revealed fat globules. Bacterial and tubercoulous cultures were negative. Cytology demonstrated abundance of lymphocytes, with no tumour cells. Subxiphoid exterior tube drainage was maintained (fig. 2B) and alipidic diet was started. After an initial success of this treatment, significant chylopericardium recurred. We did not demonstrate communication between the thoracic duct and the pericardial sac by chest computed tomography. She successfully responded to 30 days of continuous pericardial drainage and 15 days of a medium-chain triglyceride diet after 30 days of total parenteral nutrition. Follow-up echocardiography 6 months after treatment commencement showed a minimal reaccumulation of pericardial fluid without symptom.

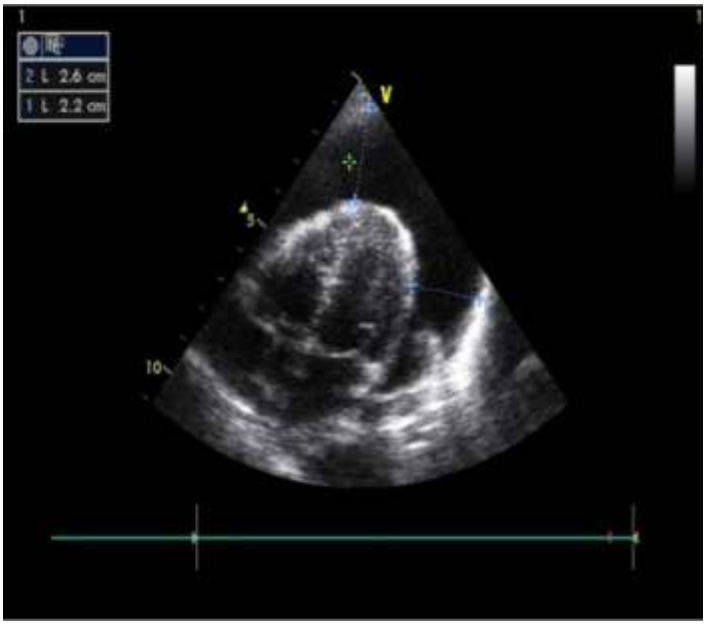

Fig-1: Echocardiogram shows a massive pericardial effusion

Fig-2: A) Subxiphoid exterior tube drainage, B) The milky fluid from pericardial effusion

DISCUSSION

Chylopericardium is sometimes a consequence of thoracic and cardiac surgery [2]. It may also occur as a result of chest trauma, mediastinal neoplasms, mediastinal tuberculosis, mediastinal radiotherapy, and thrombosis of the subclavian vein [2]. Idiopathic chylopericardium is a rare entity. It was first reported in 1886 by Hasebrock [3]. The term primary chylopericardium was first used by Groves and Effler [4], who described a case of isolated accumulation of chyle in the pericardium, with no obvious cause. In recent years, a few cases of primary idiopathic chylopericardium have been reported in which the lymphatic leak and fistula were seen by lymphangiography or CT [5].

The pathophysiology of primary chylopericardium may be related to an abnormal connection between the thoracic duct and the pericardium, with the presence of fistulas, chyle reflux associated with lymphatic hypertension with loss of the valve mechanism and increased permeability of lymphatic vessel walls [6].

The clinical presentation varies from asymptomatic state to signs of cardiac tamponade. The most common symptoms are dyspnea, fatigue, and cough.

Diagnosis is confirmed by the creamy white appearance of the pericardial fluid, the presence of fat droplets microscopically, and the confirmation of a high level of triglycerides in the fluid. The nature of the effusion is usually revealed by pericardiocentesis. However, the diagnosis of chylopericardium may be established noninvasively by precordial imaging using technetium- 99m–labeled red blood cells and oral administration of 131I-triolein [7, 8].

Diagnostic modalities essential for evaluating suspected chylopericardium include the following: A chest X-ray demonstrating cardiomegaly will lead to further investigations and eventually diagnosis. Pericardial effusion is usually diagnosed by echocardiography or computed tomography. Computed tomography and lymphangiography may be useful in the diagnosis of mediastinal lymphangiomatosis, which is the most common cause of primary chylopericardium [9].

Investigation specifically targeting malignant disease, lymphoma and tuberculosis should also be undertaken. Lymphangiography may in certain instances establish the fistulous connections and is also useful for delineating the anatomy of the thoracic duct [10].

In contrast to the success with conservative treatment of post-traumatic chylopericardium [11], such treatment of idiopathic chylopericardium is rarely successful.

Surgery is usually required, although occasional cases of successful nonsurgical treatment have been reported. The surgical procedure consists of ligation and excision of the thoracic duct just above the diaphragm, combined with partial pericardiectomy

A conservative management to chylotorax and posttraumatic chylopericardium is the use of oral-enteral MCT or total parenteral nutrition. Although most investigators report surgical procedure as a definitive treatment, they recommend starting with the conservative management consisting on drainage of the fluid, (pericardiocentesis or drainage catheter) and low-fat diet based on MCT or total parenteral nutrition [12].

REFERENCES

1. Akamatsu, H., Amano, J., Sakamoto, T., & Suzuki, A. (1994). Primary chylopericardium. *Ann Thorac Surg*, *58*, 262-6.
2. Nguyen, D. M., Shum-Tim, D., Dobell, A. R., & Tchervenkov, C. I. (1995). The management of chylothorax / chylopericardium following pediatric cardiac surgery: a 10-year experience. *J Card Surg.*, *10*, 302-8.
3. Hasebrock, K. (1888). Analyse einer chylosen pericardierlen Flussigkeit (chylopericardium). *Ztschr Physiol Chemie.*, *12*, 289.
4. Groves, L. K., & Effler, D. B. (1954). Primary chylopericardium. *N Engl J Med.*, *250*, 520-3.
5. Musemeche, C. A., Riveron, F. A., Backer, C. L., Zales, V. R., & Idriss, F. S. (1990). Massive primary chylopericardium: a case report. *Journal of pediatric surgery*, *25*(8), 840-842.
6. Fernandes, F., Arteaga, E., Carvalho, M. S. S., Ianni, B. M., Fernandes, P. P., & Mady, C. (1998). Quilopericardio idiopático. *Arq Bras Cardiol. 71*(2), 131-4.
7. Svedjeholm, R., Jansson, K., & Olin, C. (1997). Primary idiopathic chylopericardium— a case report and review of the literature. *Eur J Cardiothorac Surg*, *11*, 387-90.
8. Fujiseki, Y., Katsura, T., Goto, M., & Kawanishi, K. (1982). Non-invasive diagnosis of isolated chylopericardium using precordial pericardial imaging after oral administration of 131I-triolein: report of a case [in Japanese]. *J Cardiogr.*, *12*, 553-8.
9. de Menezes, I. C., Araujo, S. G., Damiao, A., Telo, M., Martins, F. M., & Macedo, M. M. (1990). Lymphangioma of the mediastinum as a cause of chylopericardium. *Acta Med Port*, *3*, 119-21.
10. Gallant, T. E., Hunziker, R. J., & Gibson, T. C. (1977). Primary chylopericardium: the role of lymphangiography. *AJR Am J Roentgenol.*, *129*, 1043-5.
11. Kannagi, T., Osakada, G., Wakabayashi, A., Kawai, C., Matsuda, M., & Miki, S. (1982). Primary chylopericardium. Chest, 81, 105- 8.
12. Allen, E. M., van Heeckeren, D. W., Spector, M. L., & Blumer, J. L. (1991). Management of nutritional and infectious complications of postoperative chylothorax in children. *Journal of pediatric surgery*, *26*(10), 1169-1174.