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Primary idiopathic chylopericardium presenting with cardiac tamponade

Additional material online

This article includes two additional Videos. You will find this supplemental at dx.doi.org/10.1007/s00059-013-3875-2.

Chylopericardium generally occurs after thoracic surgery or trauma. Primary idiopathic chylopericardium is an extremely rare condition especially in children and young adults. In recent years, a few case reports have been published on primary idiopathic chylopericardium due to lymphatic leak and fistula into the pericardium [1, 2, 3]. In this report, we described a 4-year-old boy with primary idiopathic chylopericardium presenting as cardiac tamponade.

Case report

A 4-year-old boy was brought to our hospital because of dyspnea, tachypnea, and hypotension. There was no history of trauma, operation, or infection. Physical examination showed respiratory distress, hypotension, and distant heart sounds. Chest radiography revealed a markedly enlarged cardiac silhouette and bilateral pleural effusions. Emergency echocardiography was performed with the suspicion of cardiac tamponade and it revealed a huge (35 mm) pericardial effusion, with diastolic collapse of the right ventricle (Video 1, **Fig. 1**). Pericardiocentesis was performed under echocardiographic control with drainage of 500 cc chylous fluid (**Fig. 2**). The biochemical characteristics of the chylous fluid were as follows: total cholesterol, 122 mg/dl; triglyceride, 181 mg/dl; low-density lipoprotein (LDL), 1120 mg/dl; very low-density lipoprotein (VLDL), 725 mg/dl; and chylomicrons, 1,440 mg/dl. Cultures were negative and there was no evidence of cytologic abnormalities. After pericardiocentesis, oral nutrition was stopped and total parenteral nutrition and octreotide treatment was start-

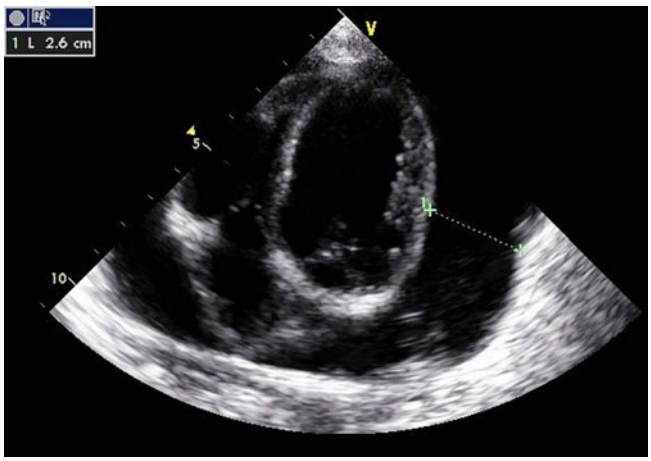

Fig. 1 ◀ Echocardiographic image after pericardiocentesis shows pericardial effusion. See also Video 1 online

Fig. 2 ▲ Chylous fluid (**a, b**)

ed. One day after pericardiocentesis, a 5 F pigtail catheter was placed in the pericardium under fluoroscopic guidance because of re-collection of chylous fluid. Despite the catheter drainage, 500 cc fluid drainage was continued daily. Therefore, a videothoracoscopic pericardial window was opened by thoracic surgeons using a two-hole method and tube drainage was performed after this procedure (Video 2). No etiological evidence was found with thoracic computerized tomography (CT), cardiac catheterization, or lymphatic scintigraphy. Conservative management consisted in maintaining effective tube drainage and octreotide therapy. A total of 1,700 ml chylous pericardial fluid was drained for 5 days. However, the effusion re-accumulated and over 400 cc of fluid per day was drained; thus, conservative treatment was deemed ineffective. Accordingly, lymphatic duct ligation of the thoracic duct was performed on the 14th day of admission. No problem was observed in the postoperative follow-up. Oral nutrition, including medium chain fatty acids, was started gradually and total parenteral nutrition and octreotide treatment was stopped. The patient was discharged on the 10th postoperative day. There was no re-accumulation at the follow-up visits, 1, 3, and 5 months after the surgery.

Discussion

Chylopericardium generally occurs after cardiac and chest surgery or trauma and is also associated with the obstruction of the thoracic duct by congenital lymphangiomatosis neoplasms or tuberculosis.

Idiopathic chylopericardium is a rare entity. It was first reported by Groves and Effler in 1954 [4]. Primary chylothorax and/or chylopericardium is rare, and this is mostly seen after mediastinal surgery. Although the exact pathophysiology of primary chylopericardium has not been established, the reflux of chylous fluid into the pericardial space was suggested as the etiology. Damage to the thoracic duct valves and the communication of the thoracic duct to the pericardial lymphatics or abnormally elevated pressure in the thoracic duct could cause chylous fluid reflux [5, 6].

The diagnostic algorithm of chylopericardium includes a primary chest x-ray that shows an enlarged cardiac silhouette and a subsequent echocardiography, which can explain the cause of cardiomegaly and pericardial fluid. Chest radiography and echocardiography are the first-line diagnostic tools with the addition of CT and cardiac magnetic resonance imaging (MRI). However, the final diagnosis is made by pericardial fluid analysis. The diagnosis can be confirmed with CT or MRI to rule out any mediastinal disease causing compression and obstruction of the thoracic duct. Lymphangiography is a time-consuming procedure and rarely delineates the diagnosis [7]. MRI ductography can be used instead of lymphangiography in experienced centers. Noncontrast fluid-weighted MRI sequences allow for visualization of not only the thoracic duct but also any congenital lymphatic malformations [8]. Some patients do not need surgical treatment and they can be treated with conservative therapy and effective drainage. If there is no tamponade and abundant pericardial fluid, oral and/or enteral medium-chain triglycerides or total parenteral nutrition is started. Octreotide, a somatostatin analogue, may reduce chyle production. Thus, it can be used for medical treatment of chylopericardium and chylothorax [9, 10]. Conservative treatment of idiopathic chylopericardium is usually not satisfactory and a failure rate of 57–60% has been reported. Thoracic duct ligation and pericardial window are the most effective procedures to prevent recurrence. Pericardial window alone is simple but carries a high incidence of recurrence [11, 12]. In our patient, thoracic duct ligation was performed after pericardial window, since he did not respond to medical treatment and pericardial window.

According to Akamatsu et al. [12], ligation and resection of the thoracic duct and construction of a pericardial window is the most effective treatment to prevent the recurrence of chylous fluid accumulation. Catheter embolization is another treatment option in selected patients. Nadolski et al. preformed thoracic duct embolization in 32 adult nontraumatic chylothoraces and 2 adult patients

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Abstract

Primary idiopathic chylopericardium is an extremely rare condition especially in children and young adults. Although the exact pathophysiology of primary chylopericardium has not been established, the reflux of chylous fluid into the pericardial space was suggested as the etiology. Damage to the thoracic duct valves and the communication of the thoracic duct to the pericardial lymphatics or abnormally elevated pressure in the thoracic duct could cause chylous fluid reflux. In this report, we described the case of a 4-year-old boy with primary idiopathic chylopericardium presenting as cardiac tamponade who was treated with video-assisted thoracoscopic window and then surgical duct ligation.

Keywords

Idiopathic chylopericardium · Duct ligation · Video-assisted thoracoscopy · Window · Children

Primäres idiopathisches Chyloperikard mit Herzbeuteltamponade

Zusammenfassung

Ein primäres idiopathisches Chyloperikard ist eine äußerst seltene Erkrankung, insbesondere bei Kindern und jungen Erwachsenen. Obwohl die genaue Pathophysiologie des primären Chyloperikards bisher nicht bekannt ist, soll der Reflux chylöser Flüssigkeit in den perikardialen Raum ätiologisch relevant sein. Eine Schädigung der Klappen des Ductus thoracicus und eine Verbindung des Ductus thoracicus mit den perikardialen Lymphwegen oder abnorm erhöhter Druck im Ductus thoracicus könnten einen Reflux chylöser Flüssigkeit verursachen. Im vorliegenden Artikel wird der Fall eines 4-jährigen Jungen beschrieben, bei dem ein primäres idiopathisches Chyloperikard, das sich als Herzbeuteltamponade manifestierte, vorlag und die Behandlung über eine videoassistierte thorakoskopische Fensterung mit anschließender chirurgischer Duktusligatur erfolgte.

Schlüsselwörter

Idiopathisches Chyloperikard · Duktusligatur · Videoassistiert thorakoskopisch · Fensterung · Kinder

with chylopericardium. They performed lymphangiography before the procedures and analyzed the thoracic duct patterns for embolization. Thoracic duct embolization was successful in 24 of 34 patients. The clinical success rates were low in cases with a normal duct pattern, with failure to opacify the thoracic duct [13].

Conclusion

Primary idiopathic chylopericardium is a rare condition in children and young adults. Video-assisted thoracoscopic window is a safe treatment but carries a high risk of recurrence. Surgical thoracic duct ligation and video-assisted pericardial window comprise the best treatment option for patients who are unresponsive to medical treatment.

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Conflict of interest. On behalf of all authors, the corresponding author states that there are no conflicts of interest.

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