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CHYLOPERICARDIUM: A CASE REPORT DEMONSTRATING UTILITY OF LYMPHOGRAPHY COMBINED WITH 3D COMPUTED TOMOGRAPHY FOR CORRECTIVE SURGICAL TREATMENT USING VATS

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ABSTRACT

We present a case of a 58 year-old woman with primary chylopericardium associated with chylothorax. Chylopericardium is a condition in which chylous fluid containing a high concentration of triglycerides accumulates in the pericardial cavity, and it can form for many different reasons. 3D computed tomography with lymphography precisely depicted the specific location of the lymphatic leak in this patient, which was successfully repaired using targeted video assisted thoracic surgery (VATS).

Keywords: chylopericardium, lymphography, video assisted thoracoscopic repair, lymphatic dysplasia, computed tomography

Chylopericardyum is an uncommon condition in which chylous fluid containing triglycerides in high concentration accumulates in the pericardial cavity. The etiology includes congenital mediastinal lymphangiectasia (1), iatrogenic after cardiac surgery (2-4), gastric signet ring cell carcinoma (5), Gorham-Stout syndrome (6), malignant tumors (7), blunt penetrating trauma, infection, radiation, congenital lymphatic anomalies (4), or primary idiopathic (8-13). We present a case of primary chylopericardium associated with chylothorax.

CLINICAL CASE

The patient is a 58 year-old woman referred to a peripheral hospital in December 2011 complaining of progressive breathlessness and palpitation. She had no fever, chest pain, cyanosis, weight loss or edema of the hands and feet. She presented a history of hypothyroidism medically treated with Levothyroxine and of paroxysmal tachycardia treated by verapamil. Chest radiography was suggestive for left pleural effusion and cardiomegaly, and computed tomography was confirmatory. Echocardiography revealed a significant pericardial effusion without signs of cardiac tamponade. Milky pleural fluid was aspirated from the left side, and initial conservative therapy was chosen.

In the following days, the patient complained of severe dyspnea, and her hemodynamic status became unstable. Imaging tests showed a significant left pleural effusion and massive pericardial effusion with signs of cardiac tamponade (*Fig. 1*). Pericardial aspiration was performed, and a pericardial

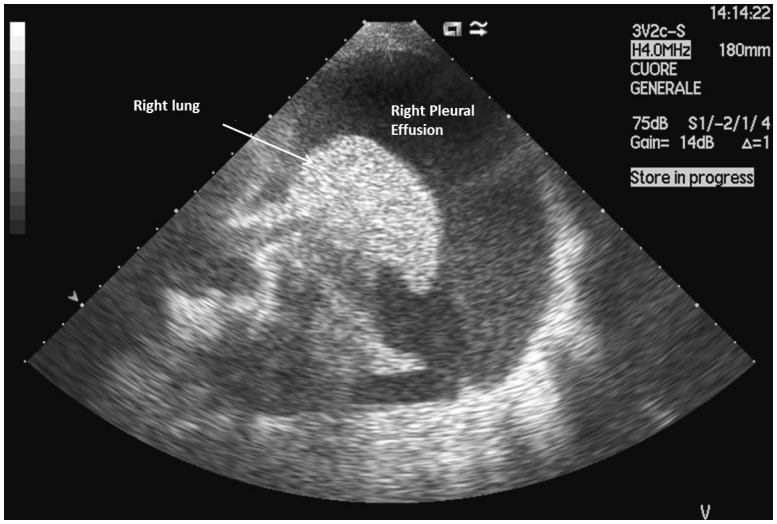

Fig. 1. Early echocardiogram of the patient demonstrating right pleural effusion.

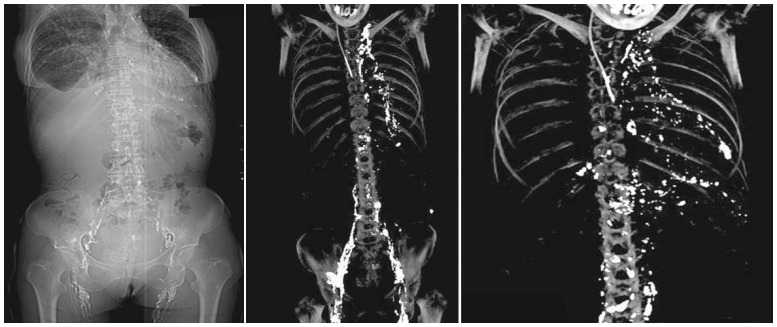

Fig. 2. Computed tomography (left) and early (middle) and late (right) lymphography demonstrating dilated and dysplastic pleuro-pericardial lymphatics and with chylous fistulas in the left pleuro-diaphragmatic region.

window was successfully carried out in combination with pleural aspiration and pleurodesis of the left pleura. Dyspnea reappeared after some months, and thoracic computed tomography (CT) and echocardiography showed moderate right pleural effusion and mild pericardial effusion. At this time, the patient was referred to our hospital for further investigation.

Her performance status was severely compromised, and clinical examination revealed hypotension, tachypnea, and desaturation. Six minute walking test demonstrated significant decrease of pulse capillary oxygenation at baseline and after physical activity (400 meters covered), and spirometry demonstrated a severe restrictive pattern.

Contrast lymphography and computed tomography 3D images showed dilated and dysplastic pleuro-pericardial lymphatics with chylous fistula at the left pleuro-diaphragmatic area (*Fig. 2*). A video- assisted thoracoscopic repair of the dysplastic area (chylous, pleuro-diaphragmatic fistula) was performed. A fatty meal (60 gm of butter melted in 250 ml of milk) administered to the patient 4 hours before surgery allowed visualization of the site of chyle effusion, which was leaking from a dysplastic pleural area at the inferior border of the mediastinal pleura. After the complete

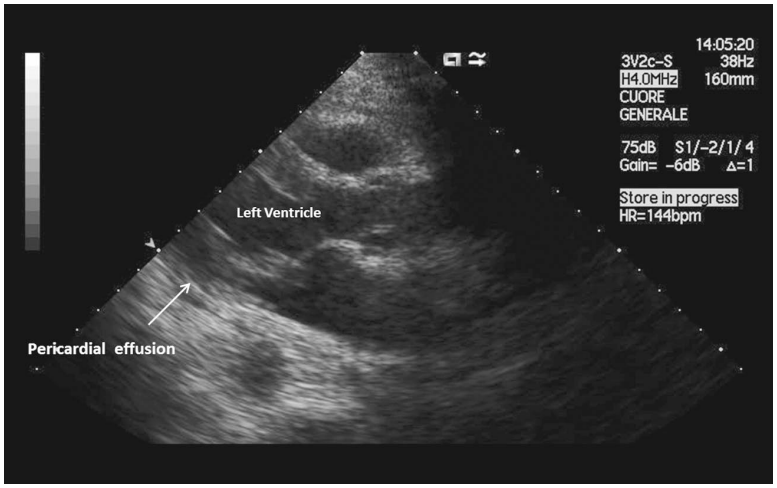

Fig. 3. Echocardiogram nine months after operation demonstrating minimal pericardial effusion.

drainage of chyle from the pleural cavity, two collagen sponges coated with human fibrinogen and thrombin (TACHOSIL) were used on the dysplastic area to seal the pleural surface, covering 1 to 2 cm beyond the edge of the area. The sponge was used to prevent bleeding and seals the tissue. Operation was completed with a chemical (talc poundage) pleurodesis. Visceral and parietal pleural apposition achieved pleurodesis. The talc was insufflated within the hemithorax to cover all visceral pleural surfaces.

After 9 months of follow up, the patient is asymptomatic without problems during ordinary life activities, and transthoracic echocardiography shows only a minimal pericardial effusion (*Fig. 3*).

DISCUSSION

Chylopericardium is an unusual disorder, and the diagnosis can be challenging with determination made often only after other causes have been ruled out. It may be primary or secondary according to involvement of lymphatic vessels. A removable "agent" can often be identified and for these occurrences, treatment is easy. In the case of primary idiopathic chylopericardium, the etiology is unknown and optimal treatment remains controversial (14). Cardiac tamponade is rare and doesn't seem to be related to etiology since it occurs both in surgical and non-surgical cases.

While many diagnostic tools have been described, we have found that enhanced CT combined with lymphangiography is a very reliable modality. Final diagnosis is ultimately made by pericardiocentesis followed by cytology, chemistry, and culture. Analysis of the fluid usually shows a whitish or milky aspirate with high triglyceride and protein content, a predominance of lymphocytes, and presence of fat globules by Sudan III staining.

Conservative therapy in the absence of hemodynamic instability is one option (15). Rarely, a single pericardiocentesis may prevent further recurrences, and pericardiocentesis followed by pericardiostomy, a medium chain triglyceride diet, or total parental nutrition can be tried especially if the patient is reluctant to undergo surgery or has concomitant life-limiting disease. Conservative treatment is effective in about 40% of patients (15), while surgery is routinely effective, so recurrence after pericardiocentesis or thoracentesis and failure of conservative treatment may be considered the indication for surgical treatment. Usual surgical intervention consists of thoracic duct

ligation, which generally leads to collateral circulation regardless of the level of ligation. To ensure the success of the surgical procedure, the thoracic duct should be ligated in the lower part of the thorax and a pericardial window should be created to allow adequate drainage to prevent complications such as constrictive pericarditis (16). The specific type of surgical procedure, Video Assisted Thoracic Surgery (VATS) versus open thoracotomy, depends on the local expertise. The VATS procedure is being used increasingly and is associated with less postoperative pain and pulmonary dysfunction (17).

In our patient, we arrived at the diagnosis in an unusual way with the chylous origin of the pleural/pericardial effusion obtained by pleural aspiration made soon after thoracic x-ray/echocardiographic evidence. CT combined with lymphangiography was only performed when the patient was admitted to our institution.

We recommend that lymphangiography be considered an indispensable diagnostic investigation that helps guide a selective solution to the lymphatic problem with a minimally invasive approach. Our case study demonstrates the critical role of lymphangiography in identifying the exact site of lymphatic defect. Following this precise identification, the surgeon is able to perform a VATS procedure, which represents the best option for the patient — definitive, minimally invasive, few side effects, and a fast recovery.

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