

Case Report

Primary Idiopathic Chylopericardium as Unusual Cause of Cardiac Tamponade

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Abstract: The chylopericardium is defined as the accumulation of chylous fluid containing high concentrations of triglycerides in the pericardial space. It is quite an uncommon condition first described in 1888 by Hasebroek. The absence of an underlying etiology defines it as primary idiopathic chylopericardium, term initially used in a study by Groves and Effler in 1954. Even when most patients are asymptomatic, constrictive pericarditis or cardiac tamponade may be referred. Cardiac tamponade, the most serious complication, represents an entity characterized by fluid accumulation in the pericardial cavity. Claudius Galen from Pergamum (131-201 D. C.) described pericardial effusions in gladiators with stab injuries of the chest and Richard Lowe (1669) described its physiology. It took two hundred years for the term "cardiac tamponade" was coined by German surgeon Edmund Rose. Cardiac tamponade can be presented from asymptomatic setting to life-threatening conditions. By the former reason, the early recognition of this entity has crucial importance. Idiopathic chylopericardium occurs in all age groups and affects both sexes equally. Although several mechanisms have been proposed in order to explain the development of chylopericardium, the underlying pathophysiology remains unclear. Damaged lymphatic vessels with abnormal communication of the thoracic duct to the pericardial lymphatics and elevated pressure in the thoracic duct can justify the presence of chyle into the pericardial sac. Diagnosis is confirmed by pericardiocentesis which must reveal at least two of the classical five criteria. Chylopericardium may be managed through conservative or surgical treatment. The conservative treatment consist a medium chain triglyceride diet or total parenteral nutrition but has a high degree of recurrence, for which reason the safest option is thoracic duct ligation with a pericardial window. The present study reports a case of chylopericardium wherein the leak was diagnosed by lymphoscintigraphy that was successfully treated by a low-fat diet, rich in medium-chain triglycerides and octreotide infusion.

Keywords: Chylopericardium, Cardiac Tamponade, Lymphangiography, Pericardiocentesis

1. Introduction

Constrictive pericarditis and/or cardiac tamponade are entities frequently seen in inpatients and its frequency can varies according the local epidemiology [1]. However, chylopericardium is a rare cause of this syndrome [2] and it is important emphasize that it must be suspected related the clinical setting [3]. Considering that constrictive pericarditis

and/or cardiac tamponade could be life threatening conditions it is important to act as soon as the diagnosis is made [4-6].

2. Case Presentation

In October 2018 a 31-year-old man was admitted to our hospital for a mild, oppressive, non-radiating chest pain of 4 days of evolution, which rapidly progressed to its maximum

intensity. The pain was not associated to positional changes or respiratory movements. Shortness of breath with the same time of evolution was referred. There was no significant medical or surgical history.

Upon examination heart rate was 88 beats per minute at rest, systolic blood pressure was 105 mmHg and diastolic blood pressure 70 mmHg. Elevated jugular venous pressure without inspiratory collapse, bilateral basal crepitations and distant heart sounds were found. The admission chest X-ray showed an enlarged cardiac silhouette with a water bottle sign (Figure 1).



Figure 1. Chest X-ray upon admission where it can be seen an enlarged cardiac silhouette with the typical water bottle sign.



Figure 2. Macroscopic characteristics of the pericardial fluid obtained via pericardiocentesis.

Electrocardiography showed no abnormalities. Transthoracic echocardiography revealed serious pericardial effusion with a swinging heart, partial collapse of the right atrium and right ventricle free wall and dilated inferior vena cava. The ejection fraction was normal. 420 mL of chylous fluid (Figure 2) were evacuated by pericardiocentesis, which showed a high level of triglycerides (505mg/dL) and a cholesterol–triglyceride ratio of less than 1.

Cytology demonstrated an abundance of lymphocytes (90% of 4,000 white blood cells). Cultures of the pericardial fluid were negative for common germs and acid-alcohol resistant bacilli. A drainage catheter was placed and an additional 500 mL of chylous fluid were evacuated in the first 12 hours. In the subsequent eleven days drainage decreased progressively until it was nil. Radiological and echocardiographic follow-up (Figure 3) showed no residual effusion.



Figure 3. Chest X-ray after drainage.

The patient underwent extensive evaluation to find the cause of the chylous pericardium. Routine laboratory tests demonstrated normal blood counts, electrolytes (sodium, potassium, chloride, calcium, phosphate and magnesium), liver function, lipid profile and renal function. There was no sign of systemic inflammatory reaction (erythrocyte sedimentation rate, 11mm/hr; negative C-reactive protein; and total leukocyte count, 8600 cells/cu mm). Immune and infectious diseases (including tuberculosis) were ruled out by standard processes. Computed tomography of the chest revealed severe pericardial effusion, with bilateral peribronchovascular interstitial thickening in the posterior and basal regions. No mediastinal nodes or masses were found. As no obvious etiology was established for the effusion, the entity was labeled primary idiopathic chylopericardium.

A lymphangiography was performed. Technetium-99m bovine collagen-gelatin (particle size 100-400 nm) was injected intradermally in the webs between the first, second and third toes, 2 mCi per site.; anterior images of the knees,

thighs, pelvis, abdomen and chest were acquired at 60 and 240 minutes after the injection. Mediastinal activity was seen in the delayed images as the radio-tracer was detected in the

pericardial space (Figure 4) and in both infraclavicular regions, thus suggesting the presence of lymph as the cause of the pericardial effusion.

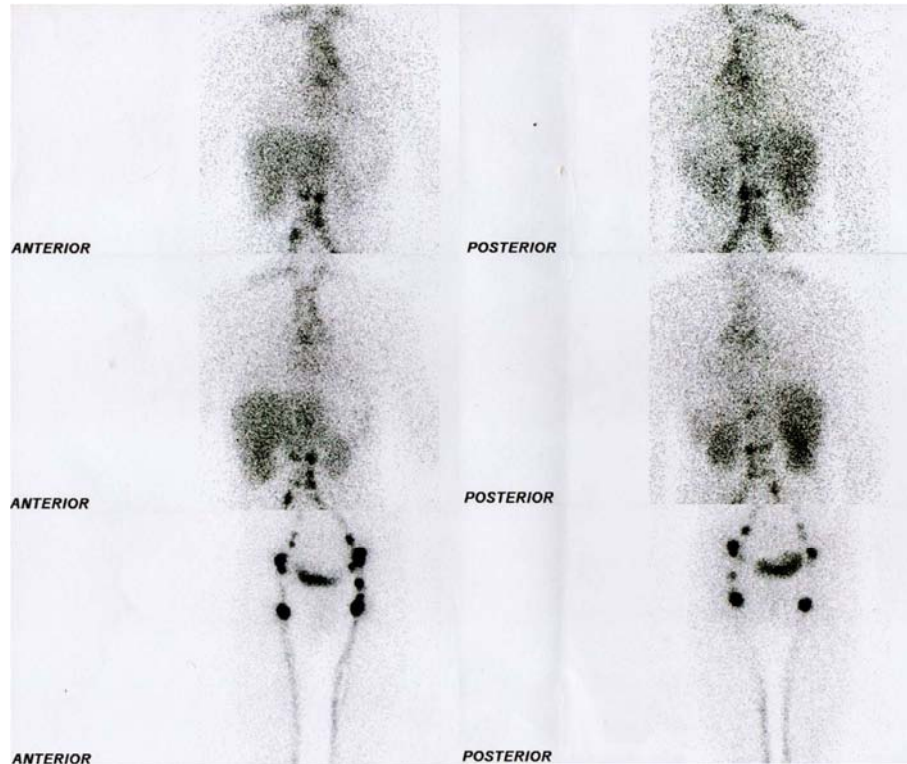


Figure 4. Lymphangiography evidencing chyle leak to the pericardial space four hours after tracer was injected.

A low-fat diet, rich in medium-chain triglycerides was indicated and an octreotide infusion was administered. For the good clinical evolution and lack of symptoms, patient was discharged with outpatient follow-up.

3. Discussion

Chylopericardium is an entity defined as the accumulation of chylous fluid in the pericardial space. It is typically the result of chest trauma [7], thoracic surgical procedures [8], mediastinal radiotherapy, mediastinal neoplasms [7, 8], signet-ring cell carcinoma of the stomach [10], mediastinal tuberculosis, filariasis, thrombosis of the subclavian vein [7] and vena cava [11], Behcet's syndrome [12], congenital lymphangiomatosis, Gorham-Stout disease (osteolysis secondary to lymphangiomatosis with bone resorption) [13], congenital mediastinal lymphangiectasia and congenital lymphatic anomalies [12]. A recent case report describes a unique case of chylothorax and chylopericardium in end stage renal disease patient on hemodialysis with a left jugular tunneled catheter who developed superior vena cava syndrome [3], thus increasing the etiology of this illness.

In 1975 chylopericardium pathophysiology was proposed for the first time [14], suggesting an obstruction in the thoracic duct and abnormal chyle reflux as responsible factors, events associated to lymphatic hypertension [15] and subsequent pericardial accumulation of chylous fluid. Chylous transudate through small pathological channels between the thoracic duct

and pericardial lymphatic ducts [12] was proposed as another cause. Other potential factors are valve damage and increased permeability of the thoracic duct [16].

If no obvious cause can be established, the entity is labeled primary idiopathic chylopericardium [17, 18]. The primary idiopathic chylopericardium is a rare condition, with only 128 cases reported in 59 years from Groves and Effler's description [18] until November 2013 [16]. The first report of chyle in the pericardial space dates back to 1888, when Hasebroeck K [19] reported 22.6 ml of such fluid found in the necropsy of a man dead from asphyxiation. The term primary chylopericardium was first used back in 1945 by Groves and Effler [18], who described recurring accumulation of chyle in the pericardium of a woman in the presence of a mediastinal cystic hygroma and use the term primary idiopathic chylopericardium in a case without an obvious etiology.

Chylopericardium is found in equal shares between genders and in all age groups [1, 20].

Signs and symptoms are not indicative of an underlying etiology [12] and chylopericardium may range from being asymptomatic [5, 8, 9] to having the clinical manifestations of a cardiac tamponade [12, 21]. The most frequently described symptoms are cough, chest pain, dyspnea and fatigue [12, 16, 22] but in a recent literature review dyspnea was present in 53.90% followed by absent of symptoms (39.42%) and cough (10.58%) [1]. Cardiac tamponade is extremely rare [21, 23].

Diagnostic suspicion is based on an enlarged cardiac silhouette on the chest X-ray. The characteristic "swinging

heart" was reported in massive pericardial effusions with the particular pattern of electrical alternans [24].

Diagnosis is confirmed by pericardial fluid test, with a triglyceride concentration at least over 500 mg/dL, a cholesterol–triglyceride ratio characteristically less than 1, and an abundance of lymphocytes in the cell count and negative bacterial counts [12, 25]. A score of 2 (specificity and sensitivity of 100%) is required for a diagnosis of chylopericardium [9, 10, 26, 27]. Triglyceride concentration in the pericardial fluid is not universally accepted diagnostic criteria and some chylopericardium cases have been established with lesser values [12].

When the diagnosis of chylopericardium has been established, a search for the underlying cause should be made.

A history of chest trauma, the introduction of subclavian venous catheters, episodes of a sudden increase of intrathoracic pressure and pericardial involvement in systemic diseases should be ruled out. Bone X-rays may show extrathoracic lymphangiomas as patches in the skeletal system [20]. Cultures of pericardial fluid, sputum and any other fluid with diagnostic potential for infectious disease help rule out infections, particularly tuberculosis. Other diagnostic approaches prove useful, such as imaging (e.g.: computed tomography or magnetic nuclear resonance), to rule out lymphatic obstruction secondary to mediastinal masses. Oral administration of Sudan III dye with an assessment of its distribution inside the pericardial space [26], lymphangioscintigraphy [21], lymphangiography [28] and thoracic radionuclide imaging after oral administration of one oral dose of ^{131}I -triolein [26, 29] and marked erythrocytes [20] have been described.

Lymphangioscintigraphy and lymphangiography are the most widely used methods to rule out abnormal communications between the pericardial sac and the lymphatic system. Lymphangioscintigraphy is useful to identify lymphopericardial fistulas, anatomic variations in the lymphatic system and partial aplasia of the thoracic duct [17]. Lymphangiography is better at delineating the anatomy of the immune system, whereas lymphangioscintigraphy is less invasive and faster [30].

According to the literature review conducted in one updated study in which 104 patients were recruited, imaging served an important role in the exploration of the leak site of chylopericardium. This study analyzed five different methods in order to diagnose this entity. In total, 42 (40.38%) cases underwent lymphangioscintigraphy and 24 of these cases presented abnormal accumulation within the pericardial sac. In addition, 24 cases (23.07%) underwent lymphangiography and 18 of them had abnormal finding; 35 (33.65%) cases underwent chest contrast-enhanced computed tomography scanning without any secondary disease finding [1].

Chylopericardium treatment is focused on preventing mechanical complications such as cardiac tamponade and constrictive pericarditis, preventing or treating metabolic and immune consequences, stopping lymphatic leaks and reducing recurrence. In the first instance, non surgical actions are required, such as pericardiocentesis, pericardial drainage,

low-fat diet with medium- and short-chain triglycerides, total parenteral nutrition and octreotide [16]. Pericardiocentesis with pericardial fluid drainage prevents mechanical complications by reducing intrapericardial pressure, improving preload and, therefore, systolic volume. A low-fat diet decreases the formation of chylopericardium and medium- and short-chain triglyceride supplementation is required for their preferential absorption through the portal vein rather than via the lymphatic vessels [11]. Total parenteral nutrition has been described as the method of choice for nutritional therapy as enteral nutrition promotes the formation of chyle. The administration of octreotide reduces intestinal fat absorption, concentration of triglycerides in the thoracic duct and in lymphatic vessel flow [16, 31, 32], although some reports does not agree with this concept [33].

Conservative treatment is not to exceed 14 days, and treatment failure is seen in up to 61% [34] of cases. There are no indicators that predict conservative treatment failure. In case of failed conservative treatment, surgical treatment is recommended, even in asymptomatic patients, so as to prevent sequelae such as constrictive pericarditis or progress to cardiac tamponade.

Surgical treatment should be indicated upon recurrence after pericardiocentesis and failed conservative treatment. Wurning et al indicate immediate surgical intervention in patients with hemodynamic instability [35]. Selle JG et al propose transthoracic ligation of the thoracic duct when chyle accumulation exceeds 1,500mL/day, no chyle flow reduction is found in the first 14 days of evolution and at the onset of imminent nutritional complications [36]. Dib et al propose a surgical procedure in chylopericardium secondary to neoplasm and when there is a daily chyle accumulation over 500mL over the first 5 days of evolution [12].

The average time between failed conservative treatment and the indication of a surgical procedure is 12 days [12]. The surgical procedure consists of pericardial window, pericardiectomy, and ligation of the thoracic duct just above the diaphragm, pericardioperitoneal shunt and thoracic duct embolization. The purpose of the pericardiectomy is to ensure complete drainage and prevent constrictive pericarditis. The pericardial window is a relatively simple technique, but has a high recurrence rate as it does not take care of the communication between the thoracic duct and the pericardial cavity. Thoracic duct ligation is effective as it decreases lymph flow to the pericardial sac through existing anomalous lymphatic vessels. The ligation should be performed at the level of the diaphragm as its anatomy varies above the diaphragm. Approximately 25% of the cases have multiple thoracic ducts above the diaphragm [37]. It should be emphasized that thoracic duct ligation induces collateral chyle drainage, independently from the level of the ligation, for the increased pressure in the duct. Up to 50 mm Hg increased duct pressure has been found after ligation, returning to normal values after 16 days [38]. That is the reason for the pericardial window [35].

The combination of thoracic duct ligation and a pericardial window seems to be the most effective procedure to prevent

recurrence [1, 4, 5].

The complications of this entity are the consequence of persisting chyle leak, which causes immune, metabolic and nutritional alterations [15].

4. Conclusion

Chylopericardium is a rare entity. Presentation may be acute, chronic, or even subclinical. Patients should undergo chest computed tomography followed by lymphangioscintigraphy/ lymphangiography to rule out secondary causes. Chylopericardium is treated by pericardiocentesis and diet according to its severity and etiology. This disorder can be resolved through dietary management, after initial drainage. Diet consists mainly in medium chain triglycerides. If, in spite of this treatment, chylopericardium persists surgical procedure is needed. Surgery should include ligation and resection of the thoracic duct just above the diaphragm with a pericardial window for subsequent drainage and prevention of complications such as constrictive pericardial disease.

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