Case report

Primary isolated chronic chylopericardium

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Abstract

Primary isolated chylopericardium is a rare entity. Its exact pathophysiology is still unknown. A case of chronic isolated primary pericardium diagnosed 12 years after the initial diagnosis of an asymptomatic pericardial effusion is reported. The diagnosis was established incidentally during surgery for resection of a papillary fibroelastoma of the aortic valve.

1. Introduction

Primary isolated chylopericardium is accumulation of chylous fluid in the pericardial space in the absence of any precipitating factor [1] such as: history of major systemic disease, trauma, body weight loss, mediastinal irradiation, generalized anomalies of the lymphatic system, exposition to tuberculosis, intrathoracic surgery, mediastinal tumors, filariosis [2], subclavian vein thrombosis [2], caval obstruction [3], or congenital lymphangiectasia [3].

We report a case of primary isolated chylopericardium discovered incidentally during surgery for resection of an aortic valve fibroelastoma.

2. Case report

A 68-year-old female was admitted for resection of a tumor of the aortic valve.

In 1989, a routine chest roentgenogram showed an unexpectedly large cardiac silhouette. Echocardiography at that time revealed an abundant (20 mm) circumferential pericardial effusion which was surprisingly well tolerated. She was totally asymptomatic and the effusion remained unchanged with no signs of compression of the cardiac cavities on serial echocardiographic studies performed at 6 months intervals. In spite of numerous work-up studies the etiology of this effusion was never elucidated. The patient always refused pericardiocentesis.

Prior to her present admission, an echocardiogram performed to control the pericardial effusion revealed the presence of a tumor of the aortic valve. A papillary fibroelastoma was suspected and the patient was referred for resection of the tumor.

Following median sternotomy the pericardium was found to be slightly thickened. Incision of the pericardium revealed abundant thick milky effusion. No other abnormalities could be noted.

Using standard cardiopulmonary bypass and myocardial protection techniques, an oblique aortotomy was performed and the tumor resected from the non-coronary cusp with care taken not to injure the aortic valve. The operation was terminated in the usual fashion. The pericardium was not resected and was left open.

Analysis of the pericardial effusion revealed: triglycerided at 860 mg/dl.

Pathological studies of the pericardium showed non-specific chronic inflammatory modifications. Pathology of the aortic valve tumor concluded to a papillary fibroelastoma.

Postoperatively, the chest tubes were frequently milked and examined in order to detect chylous drainage. Since no drainage was observed the tubes were removed at 48 h.

The patient was discharged on day 5 with an echo showing absence of pericardial effusion. Echocardiographic studies were obtained at 1, 3, 6 and 12 months and showed no recurrence of the pericardial effusion.

3. Comments

Primary isolated chylopericardium was first reported by
Groves and Effler in 1954 [4]. Its precise etiology is still to be elucidated [3,5]. Pathophysiological hypotheses incriminate abnormally elevated pressure in the thoracic duct as in lymphangiectasia or the coexistence of damaged valves of the thoracic duct together with its communication to the pericardial lymphatic vessels [5], directly or via suprarenal chorial lymph nodes [6], causing a chylous reflux [5].

Symptoms depend on the importance of the effusion and on compression of the cardiac cavities. Chronic effusions may remain totally asymptomatic for a long period of time. Whenever cardiac compression occurs symptoms are those observed with tamponade and include: recurrent syncope, exertional dyspnea [1,7], chest pain, fatigue, upper abdominal discomfort, cough, palpitations [2], chylous pericardium is usually diagnosed by pericardiocentesis that shows the presence of chylous fluid with a triglyceride level between 190 and 2000 mg/dl [1–3,6–8]. Pathological analysis demonstrates white yellow chylous fluid [1,7], with numerous foamy cells and fat globules shown by Sudan III staining [1]. Also noted are extra-cellular fat droplets [3] and predominance of lymphocytes [5].

Asymptomatic pericardial effusions are usually diagnosed on routine chest X-ray, echocardiography, computerized tomography scan or magnetic resonance imaging. However, these exams do not show any specific characteristics of the chylous effusion [1,2,5].

Whenever a chylopericardium is suspected following pericardiocentesis several techniques can be used to confirm diagnosis.

Oral administration of Sudan III may be interesting as the dye might appear in the pericardium. On the other hand, radionuclide imaging using 1131 triolein, or pedal 99m Tc-Sulfur colloid [1,7] may show a communication between the lymphatic system and the pericardial sac. In other cases these studies may reveal the absence of opacification of the left subclavian vein suggesting retention of the isotope in the upper mediastinum and accumulation of chyle in the pericardial sac. Concomittant accumulation in the pleural cavities may also be observed.

Non-surgical management includes dietary regimen based on medium chain triglyceridates [1,2,7] and serial pericardiocentesis. However, this conservative treatment alone is associated with reaccumulation of fluid [1–3,8]. Surgical treatment has been proposed to halt recurrence and progression for a later tamponade or constRICTIVE PERICARDITIS [8]. Surgical modalities include pericardial-peritoneal shunts, pericardiectomy and thoracic duct ligation [3]. Pericardiectomy alone results in a high recurrence rate [2,7]. Therefore this technique should always be coupled with ligation of the thoracic duct. This is obtained by ‘Mass ligature’ of all tissues between the descending aorta, the vertebral body and the azygos vein at the level of the diaphragm [3,5]. This procedure can be performed using right-sided video-assisted thoracoscopy and is associated with a zero recurrence rate at 4 year follow-up [1,3,5,7,8].

In our case the pericardial effusion was followed non-invasively by echocardiography for 12 years. The chylous origin was never suspected since the patient always refused pericardiocentesis and since all work-up studies were negative. The patient remained totally asymptomatic throughout this period with no compression of the cardiac cavities. Surgery was decided when a papillary fibroelastoma of the aortic valve was discovered incidentally on echocardiography. During surgery, this chronic effusion was found to be chylous with a triglyceride level of 860 mg/dl. Although no specific technique was used to treat this chylopericardium, and despite a normal postoperative diet, no chylous drainage in the chest tubes was observed. Moreover, Echocardiographic follow-up at 12 months was normal with no recurrence of the pericardial effusion.

No correlation could be found in the literature between the presence of chylopericardium and papillary fibroelastoma of the aortic valve.

References