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Primary idiopathic chylopericardium: a rare case with a synopsis of the literature

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ABSTRACT Primary idiopathic chylopericardium is a rare clinical entity characterised by the collection of chyle within the pericardial cavity without a definitive cause. This case report describes the history, physical examination, evaluation, diagnosis and treatment of a 19-year-old boy with primary idiopathic chylopericardium. Radiological findings and biochemical analysis of the pericardial fluid following pericardiocentesis sustained this diagnosis. Initial conservative management failed, and the patient was surgically treated subsequently. He recovered well postoperatively and remained asymptomatic thereafter. Primary idiopathic chylopericardium is a rare pathology with very few cases reported till date, and the symptoms are commonly due to cardiac compression. Computed tomography of the chest and bipedal lymphoscintigraphy are considered the standard methods for accurate diagnosis, and in cases of failed medical treatment, open and thoracoscopic thoracic duct ligation with pericardiectomy have been described as the best surgical options.

Keywords: chylopericardium, pericardiectomy, pericardiocentesis, thoracic duct ligation
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INTRODUCTION

Chylopericardium is the accumulation of chylous fluid in the pericardial space. It may be primary due to the absence of any clear aetiology or secondary to iatrogenic cardiothoracic trauma, malignant mediastinal tumours such as mediastinal dysgerminomas, gastric signet ring cell carcinoma, Gorham syndrome, infections such as tuberculosis, radiation, congenital lymphatic anomalies, blunt or penetrating trauma,[1] subclavian vein thrombosis, vena caval obstruction and filariasis.[2] This condition was first observed in 1888 by Hasebrock during an autopsy and later termed as 'primary' or 'idiopathic' chylopericardium by Groves and Effler in 1954.[3]

CASE REPORT

A previously healthy 19-year-old male patient was referred to our hospital for further investigations and management. His chief complaints were mild to moderate exertional dyspnoea, cough with yellowish expectoration, a mild fever with daily spikes for one week, with radiological findings of cardiomegaly, pericardial and plural effusions as well as pneumonia in the left lower lobe. There was no history of chest pain, palpitations, chest trauma, dizziness or mediastinal irradiation. Past medical, surgical and remaining histories were not significant. On physical examination, the patient was found to be of average size for his age, with mild abdominal distension and a slightly raised jugular venous pressure. Other general signs were normal and his vitals were stable. His apex beat was not visible, and there were no palpable thrills. Heart sounds were regular but slightly muffled, with no cardiac murmurs and pericardial friction rub. The lungs were clear.

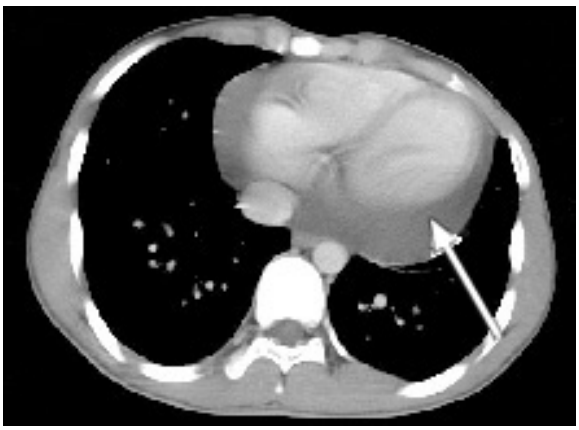

Fig. 1 Chest CT image shows an enlarged cardiomediastinal area with pericardial effusion (arrow).

All routine blood workup, enzyme assays, tumour markers and serological tests were within normal limits. Chest radiography showed cardiomegaly with a cardiothoracic ratio of 0.7. The presence of massive pericardial effusion was confirmed by echocardiography and chest computed tomography (CT); the latter also revealed the absence of mediastinal mass (Fig. 1). A subxiphoid pericardiocentesis yielded about 1,000 ml of milky fluid, which provided relief to the patient, and the catheter was left *in situ* for continuous drainage. Biochemical analysis of the pericardial fluid showed a high triglyceride level of 1,150 mg/dL and a cholesterol-triglyceride ratio < 1, which ascertained that the fluid was of lymphatic origin. Cytological studies reported a leucocyte count of 21 × 103/mm^3, and bacteriological stains, including acid fast bacilli and cultures, were negative. Since we could not discern its possible aetiology, a diagnosis of primary idiopathic chylopericardium was concluded.

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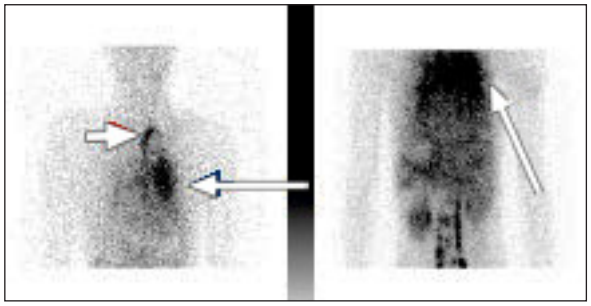

Fig. 2 Lymphoscintigraphy image shows increased radioactivity just before the thoracic duct joins the systemic circulation (arrowhead), with pooling of the tracer around the heart (arrows).

Initially, a conservative approach was adopted by keeping the patient nil by mouth with total parenteral nutrition (TPN). Somatostatin analogue 6 mg/day was used for seven days, along with continuous pericardial drainage. The effusion gradually decreased to about 50 ml per day. By the second week, normal diet was introduced, but the effusion restarted and the fluid accumulated to about 300 ml per day for seven days. Occluding the catheter for one day would result in cardiac tamponade. Lymphoscintigraphy using ^{99m}Tc-Dextran (DX) was then performed, which demonstrated an increased intrathoracic duct pressure and intense pooling of the tracer around the heart area (Fig. 2). Finally, definitive surgical treatment was decided upon, as conservative treatment was deemed ineffective.

The patient was put under general anaesthesia and placed in the left lateral position, with a double lumen endobronchial tube *in situ* for one-lung ventilation. Through a right minithoracotomy at the seventh intercostal space, a 6-cm incision was made and the thoracic duct was identified between the azygos vein and the descending aorta. It was isolated from the level of the inferior pulmonary vein down to the diaphragm, where it was doubly ligated 4 cm apart and resected. Then the 2-mm thickened pericardium was retracted upward in front of the right phrenic nerve, where a pericardial window of 2.5 cm × 1.5 cm was made longitudinally. The pericardial sac was empty and the catheter was *in situ*, with loose adhesions and no loculi between the visceral and parietal pericardium. Postoperatively, the patient recovered rapidly, and the chest drain was removed on the seventh day, as the volume drained was practically nil. This was confirmed by chest radiography and echocardiography. The patient was discharged after two weeks and has since been asymptomatic. Repeat echocardiography done during the follow-up at 12 months showed no recurrence of the effusion.

DISCUSSION

Primary idiopathic chylopericardium is an unusual cause of pericardial effusion and its aetiology is not well defined; its incidence is equal in both genders and across all age groups.[4] Only about 114 cases had been reported in the literature till 2007.[3] The pathophysiology of this condition is not very clear, but has been postulated as follows: (a) lymphangeal anomalies between the thoracic duct and pericardial cavity; (b) marked elevation of intrathoracic duct pressure due to obstructions causing chyle reflux;[5] (c) valvular incompetence or ruptured lymphatic valves due to a sudden rise in the intrathoracic pressure in blunt chest trauma;[6] and (d) increased permeability of the lymphatic vessel walls.[3] Nevertheless, the clinical features of this condition depend on the extent of the pericardial effusion and cardiac chamber compression. Symptoms usually resemble those of cardiac tamponade, i.e. exertional dyspnoea, cough, chest pain, palpitations, easy fatigability, upper abdominal discomfort and recurrent syncope.[2]

Investigations generally begin with an anteroposterior chest radiograph, which typically shows an enlarged cardiomediastinal shadow. Nonspecific T-wave flattening on electrocardiography necessitates echocardiography, which often reveals pericardial effusion. In order to elucidate its nature, a pericardiocentesis followed by biochemistry, cytological and culture analysis are of paramount importance. The most striking features of chylous fluid are a high triglyceride level of 110–2,000 mg/dL and a cholesterol-triglyceride ratio < 1.[1,7] The diagnosis can be confirmed with CT or magnetic resonance imaging to rule out any mediastinal disease causing compression and obstruction of the thoracic duct. Lymphangiography can show abnormal lymphatics and delineating anatomy of the thoracic duct, which is helpful for surgeons; however, the procedure is time consuming and painful.[6] Lymphoscintigraphy, a rapid and noninvasive procedure where the radioisotope can be traced from the lymph vessels to the pericardial cavity, can be used to identify fistulas, anatomical variations or partial aplasia of the thoracic duct.[8]

Treatment usually starts with a conservative or dietary approach, aimed to decrease lymph formation. Along with a tube pericardiostomy, the patient is kept fasting and given TPN, or put on an oral-enteral nutritional regime with low-fat diet and medium-chain triglyceride, which are directly absorbed by the portal system, thus avoiding the lymphatics and thereby decreasing lymph formation. Sympathomimetic drugs such as somatostatin and ethylephrine can be used as adjuvants to dietary measures, as they increase lymphatic drainage by inducing smooth muscle contraction of the thoracic duct.[9] However, this objective is not always achieved, and there are no proper guidelines governing when conservative treatment is considered to be a failure. Dib et al suggested that an average daily loss of chyle of 500 ml over five days should be an indication for surgery.[1]

The preferred surgical procedure is thoracic duct ligation with concomitant fashioning of a pericardial window to ensure adequate drainage, and this can be done by a right or left thoracotomy. The right-sided approach, which is our preference, allows easy access, as the duct ascends on the right side after emerging from the aortic hiatus of the diaphragm, but sometimes, mass ligation is required due to anatomical variation at this level. Dairy cream or olive oil given prior to surgery helps in the identification of the duct. Pericardiectomy alone has a high recurrence rate of 50%, and hence, should always be coupled

with thoracic duct ligation.[1,4] More recently, video-assisted thoracoscopic surgery is preferred over the traditional open approach, since magnification allows better identification of the mediastinal structures. Moreover, the former is associated with less postoperative pain and pulmonary dysfunction, with minimal scarring.[10] Another therapeutic option proposed by Chan et al is the pericardial-peritoneal Denver shunt.[2,7] This condition, if left untreated, may lead to complications such as compromised nutritional and immunological status, cardiac tamponade and constrictive pericarditis.[1,5]

In conclusion, our patient was symptomatically relieved following pericardiocentesis, but the nature of his nonspecific intermittent fever could not be explained. We advocate that medication should be the first choice of treatment, failing which definitive surgery should be considered. The preference for right thoracotomy is due to its easier access, and thoracic duct ligation just above the diaphragm along with pericardiectomy ensures the success of the procedure. However, extensive studies are warranted in order to clarify the aetiological hypotheses and to better define the therapeutic measures.

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