Isolated Primary Chylopericardium : A Case Report

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Abstract

Isolated primary chylopericardium is a rare entity with an obscure etiology. The authors report a 10-week-old male infant presenting with tachypnea and enlarged cardiac silhouette. Echocardiography revealed a large pericardial effusion. A specific diagnosis of chylopericardium was made by pericardiocentesis and analysis of the fluid. Despite the pericardial tube drainage and medium-chain triglyceride diet, pericardial effusion reaccumulated. Ligation of the thoracic duct with the establishment of a pleuropericardial window was performed through a left thoracotomy. Follow-up echocardiograms have shown no reaccumulation of the pericardial fluid.

Key word : Chylopericardium, Pericardiocentesis, Thoracic Duct Ligation, Pleuropericardial Window

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Chylopericardium is a rare condition that usually occurs after cardiac surgery, trauma, irradiation, thrombosis of either the subclavian vein or superior vena cava, or obstruction of the thoracic duct by tuberculosis, neoplasm, or congenital lymphangiomatosis[1-3]. Isolated primary chylopericardium is extremely rare. In recent years, a few cases of lymphatic leakage and interconnections with the pericardial sac have been demonstrated in adults by computer tomography, lymphangiography or an intra-

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operative thoracic ductogram[4-6].This study reports a case of isolated primary chylopericardium in a 10-week-old infant who was treated with thoracic duct ligation and establishment of pleuropericardial window.

CASE REPORT

A 10-week-old male infant, who was first admitted at the age of 5 weeks and diagnosed as having viral pneumonia and pericarditis, was rehospitalized due to increasing tachypnea. There was no apparent history of trauma, surgery or central venous catheter placement. Chest radiography showed a marked enlargement of the cardiac silhouette. Echocardiography demonstrated a large amount of pericardial effusion compressing on the right atrium (Fig. 1). A diagnosis of chylopericardium was established, after aspiration of 150 ml of milky fluid with a white cell count of 150/mm^3 (80% lymphocytes and 20% neutrophils) by pericardiocentesis. The fluid triglyceride level was 1,009 mg per cent, cholesterol 33 mg per cent and protein 11.2 g per cent. Sudan stain was positive for fat globules. Gram stain, acid-fast stain and culture were negative. A pericardial drain was placed percutaneously using a pigtail catheter, and a low-fat diet enriched with medium-chain triglycerides was started. A CT scan of the chest did not reveal any tumor or congenital malformations. Five days after admission, the patient underwent subxiphoid drainage that removed 200 ml of milky fluid, and a chest tube was retained for 2 weeks. After removal of the chest tube, pericardial fluid reaccumulated, and a pigtail catheter was placed again for drainage, due to the parental refusal of surgery for thoracic duct ligation. As the amount of fluid decreased, the patient was discharged from the hospital. However, the fluid reaccumulated slowly, eventually the patient underwent thoracic duct ligation and pleuropericardial window creation through the left thoracotomy. Follow-up clinical examination, chest roentgenogram and echocardiogram showed no reaccumulation of the pericardial fluid 18 months post-operatively.

DISCUSSION

Since the first report of primary chylopericardium in 1954[10], there have been only a few cases reported in English literature particularly in infants [1,4-18]. Primary chylopericardium can occur in patients ranging from newborn to 77 years of age and it affects both sexes equally[1]. The symptoms of chylopericardium vary from asymptomatic to respiratory distress and cardiac tamponade[1,7,8]. The pericardial fluid that has a milky-white appearance and contains fat globules are diagnostic for chylopericardium. Protein and triglyceride are usually elevated in chylous fluid, but they may be similar to the

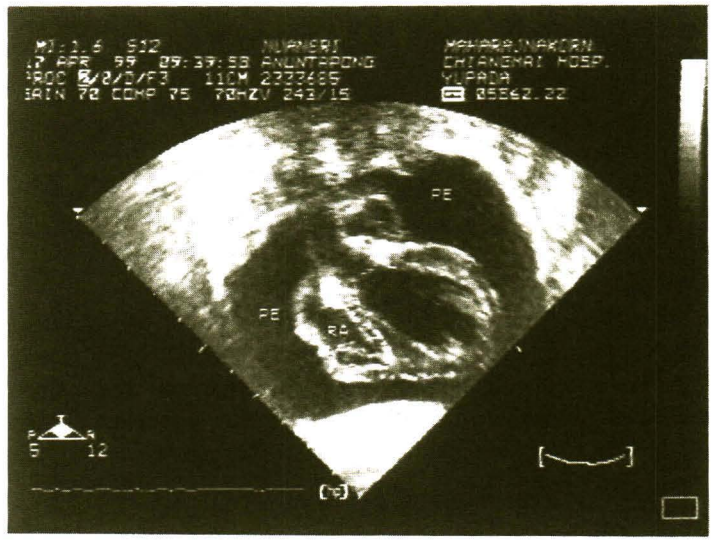

Fig. 1 Echocardiogram in subcostal view shows massive pericardial effusion. PE = pericardial effusion, RA = right atrium, S = superior, I = inferior, R = right, L = left.

patient's serum, depending on dietary intake. Chylous effusion has a low cholesterol level and high neutral and fatty acids, contrary to cholesterol pericarditis. Several underlying disorders such as, previous cardiac surgery, mediastinal irradiation, neoplasm of the mediasternum, lymphangiomatosis, and thrombosis of the subclavian vein or superior vena cava, need to be excluded in order to establish the diagnosis of primary chylopericardium.

Most investigators now agree that the cause is due to some abnormalities in the lymphatic connections between the lymphatic system and the pericardial sac[4,5,9]. These abnormal connections arise most frequently from the left suprabronchial lymph nodes or directly from the thoracic duct[18]. Reflux of chyle from damaged valves or elevated pressure in the thoracic duct has also been mentioned as a mechanism of this entity[8]. Lymphangiography or computer tomography (CT) combined with lymphangiography is useful for the evaluation of lymphatic connection abnormalities in adults or older children [4,5], but is unsuitable for infants or small children. Some radionucleotide imaging techniques have been used recently to diagnose chylopericardium and lymphodynamic such as oral ^{131}I-triolein and pedal ^{99m}Tc-SC lymphoscintigraphy[19]. Chest CT or MRI is a useful noninvasive investigation to rule out mediastinal tumor or lymphangiomatosis.

In contrast to post-traumatic chylopericardium conservative treatment of idiopathic chylopericardium is rarely successful[1]. Surgical therapy is usually required, although occasional cases treated conservatively have been reported[1,18]. The surgical procedure consists of thoracic duct ligation and pericardial window creation performed by either video-assisted thoracoscopy or through thoracotomy[1,4-17,20]. It is the authors' opinion that conservative management with medium chain triglyceride diet or parenteral nutrition may be eliminated in the treatment of isolated primary chylopericardium to minimize the loss of lymph fluid and hospital stay, except for patients who are not suitable for operation.

Chylopericardium can be diagnosed by a careful analysis of the pericardial fluid. Once diagnosed, secondary causes should be ruled out. Ligation of the thoracic duct with pleuropericardial window creation is the treatment of choice for isolated primary chylopericardium.

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ภาวะมีน้ำเหลืองปนไขมันในถุงหุ้มหัวใจชนิดปฐมภูมิ : รายงานผู้ป่วยและทบทวนวารสาร

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ภาวะ chylopericardium ชนิดปฐมภูมิเป็นภาวะที่พบไม่บ่อย รายงานนี้กล่าวถึงผู้ป่วยทารกอายุ 3 เดือนมาโรงพยาบาล ด้วยเรื่องหายใจเร็ว และพบมีเงาหัวใจโตจากภาพรังสีทรวงอก การตรวจคลื่นสะท้อนความถี่สูงของหัวใจพบว่ามีน้ำในช่องเยื่อหุ้มหัวใจเป็นจำนวนมาก การวินิจฉัยภาวะ chylopericardium นั้นได้จากการเจาะของเหลวจากช่องเยื่อหุ้มหัวใจมาตรวจพบว่ามีลักษณะคล้ายนมและพบหยดไขมันจากการตรวจด้วยกล้องจุลทัศน์หรือย้อมด้วยน้ำยา Sudan ผู้ป่วยได้รับการรักษาด้วยการใส่ท่อในช่องเยื่อหุ้มหัวใจเพื่อเอาของเหลวออกและให้รับประทานนมที่ไขมันต่ำซึ่งประกอบด้วย medium chain-triglyceride แต่กลับมีของเหลวมาคั่งในเยื่อหุ้มหัวใจอีก หลังจากเอาท่อระบายของเหลวออก ผู้ป่วยจึงได้รับการผ่าตัดโดยเย็บผูก thoracic duct และ ทำช่องระหว่างเยื่อหุ้มหัวใจกับช่องอก หลังผ่าตัดได้ติดตามผู้ป่วยนาน 18 เดือนไม่พบว่ามีอาการหรือตรวจคลื่นสะท้อนความถี่สูงของหัวใจพบของเหลวผิดปกติในช่องเยื่อหุ้มหัวใจเกิดขึ้นอีก

คำสำคัญ : ภาวะมีน้ำเหลืองปนไขมันในถุงหุ้มหัวใจ, การเจาะเยื่อหุ้มหัวใจ, การผูกท่อน้ำเหลืองใหญ่ของช่องอก, การเปิดช่องระหว่างเยื่อหุ้มหัวใจและช่องอก

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