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CHYLOPERICARDIUM: A RARE CAUSE OF PERICARDIAL EFFUSION

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ABSTRACT:

A 21 year old man presented with asymptomatic, isolated chylopericardium. Despite echocardiography, radionuclide-angiography, computer tomography, and chemical analysis of the chylous effusion, the etiology remained obscure. After patent blue dye infusion into peripheral soft tissues, the appearance of coloring material in the effusion at 4 hours suggested direct communication of the pericardium with an apparently large thoracic duct. Fifteen months later, cardiomegaly persists in site of medium-chain triglyceride dietary restriction.

CASE REPORT

An active, asymptomatic 21 year old man was admitted to the hospital for evaluation of an enlarged cardiac silhouette noted on screening chest x-ray (Fig. 1a). Four years earlier he sustained a thoracic contusion without fracture following a motorcycle accident, but otherwise had no medical ailments.

Physical examination revealed a well-proportioned, healthy appearing man. Blood pressure was 135/90 mmHg. Heart sounds were normal to auscultation without murmurs or "snap". Cervical venous distension, hepatomegaly, splenomegaly and paradoxical pulses were notably absent. Electrocardiography disclosed normal voltage, mild bradycardia (55/min) with unremarkable QRS complexes and ST segments. Standard hemogram and urinalysis were normal. Chest x-ray revealed an enlarged cardiac silhouette (Fig. 1a) without pulsation at fluoroscopy. Echocardiography (M-mode and 2D) showed a large pericardial effusion but the myocardium was of normal thickness and contractility (Fig. 2).

Pericardiocentesis yielded 750 ml of yellowish, milky, sterile fluid with a specific gravity of 1.015. Of significance the cholesterol level was 88 mg/dl; triglycerides 3110 mg/dl and the VLDL-TG 1800 mg/dl levels compatible with chyle. Total protein of the fluid was 4.0 g/dl with 74.9% albumin. IgG was 588; IgA 101 and IgM 103 mg/dl. Cytologic examination revealed a few lymphocytes and polymorphonuclear leukocytes, occasional erythrocytes and many mesothelial cells.

After pericardial aspiration, the cardiac silhouette was much less prominent (Fig. 1b) but over the ensuing four weeks gradually increased again (Fig. 1c). Radionuclide-angiography (99mTc labeled pyrophosphate) showed an activity-free space around both ventricles ("halo sign") indicative of pericardial effusion and best seen in the left anterior oblique (LAO-45) position (Fig. 3). Right heart catheterization (Swan-Ganz) with exercise stress test (bicycle, upright — Bruce protocol) had to be terminated at 225 watts/min because of patient exhaustion at 87% of capacity. Measured values of heart rate, blood pressure, pulmonary arterial pressure and cardiac index are shown in the Table.

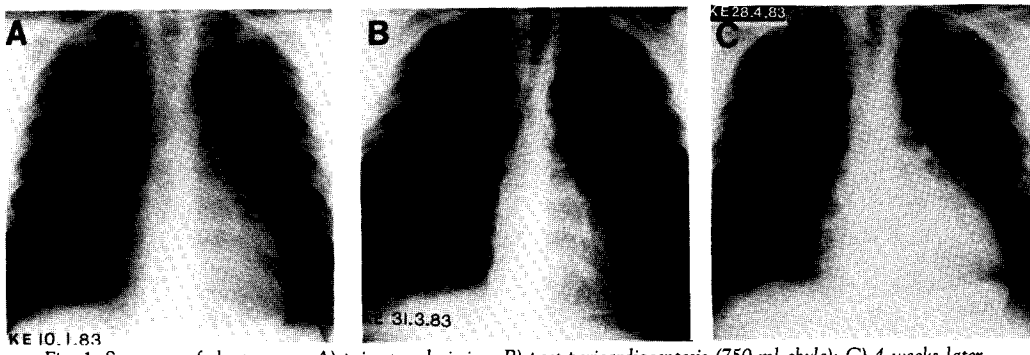

Fig. 1: Sequence of chest x-rays. A) prior to admission; B) post-pericardiocentesis (750 ml chyle); C) 4 weeks later showing reaccumulation of effusion.

Table

RIGHT HEART CATHETERIZATION
EXERCISE STRESS TEST

Exercise Watts/sec	HR Beats/min	BP mmHg	PAP (mmHg) syst/diast/mean	CI l/min/m^2
Rest	65	135/90	23 / 13 / 15	3.8
75	93	130/90	25	
125	115	150/85	28	
175	145	190/100	33	16.3
225	170	200/105	60 / 20 / 36	9.3

HR = heart rate; BP = blood pressure; PAP = pulmonary arterial pressure; CI = cardiac index

Mediastinal computer tomography confirmed the presence of a pericardial effusion with a density of 15 HU (Hounsefield units) (Fig. 4a). There was no sign of malignancy or lymphatic malformation. At several intrathoracic levels a dilated vascular channel (diameter of 2 cm) was seen, and the image was not changed after intravenous instillation of 100 ml contrast material (Uromiero 300), and probably, therefore, was a dilated thoracic duct (Fig. 4b). Instillation of patent blue dye subcutaneously in the right foot appeared prominently in the pericardial fluid four hours later suggestive of a possible direct major lymph trunk (e.g., thoracic duct) pericardial communication.

Follow-up chest x-rays over the next 15 months revealed the cardiac silhouette to be enlarged but stable. The patient is still asymptomatic but remains on a medium chain triglyceride restricted diet.

DISCUSSION

Approximately 57 patients with chylopericardium have been reported (1-7). In 11 patients (19%) chylopericardium followed operative cardiotomy and an abnormality of lymphatic drainage was implicated (8). In 17 patients (29%) a discrete malformation of mediastinal lymphatics such as lymphangioma (7 patients) or lymphangiectasia (10 patients) was detectable. In the remaining 30 patients (52%) no explanation for chylopericardium was forthcoming despite extensive diagnostic ex-

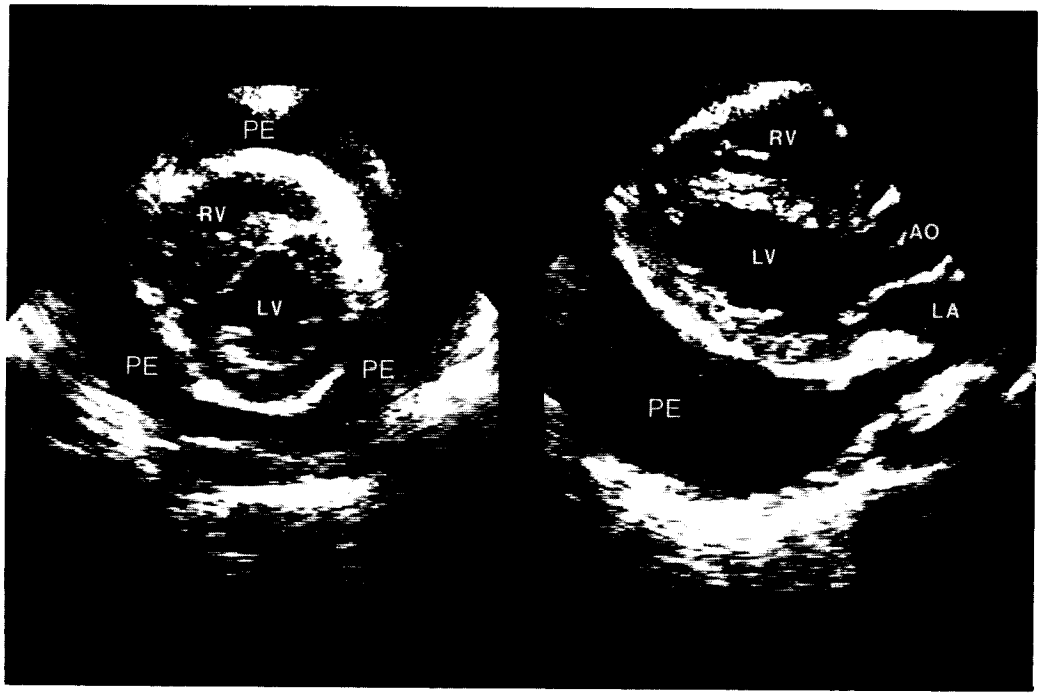

Fig. 2: 2D-echocardiographic images. Left-parasternal short axis view in the plane of the papillary muscles. Right-peristernal long axis view PE = pericardial effusion; LV = left ventricle; RV = right ventricle; LA = left atrium; AO = aorta.

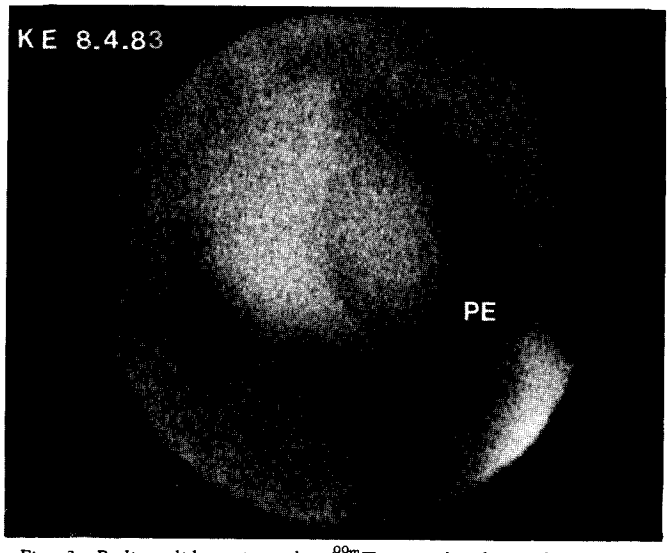

Fig. 3: Radionuclide-angiography (^{99m}Tc pyrophosphate) demonstrating pericardial effusion (PE). Projection LAO 45°.

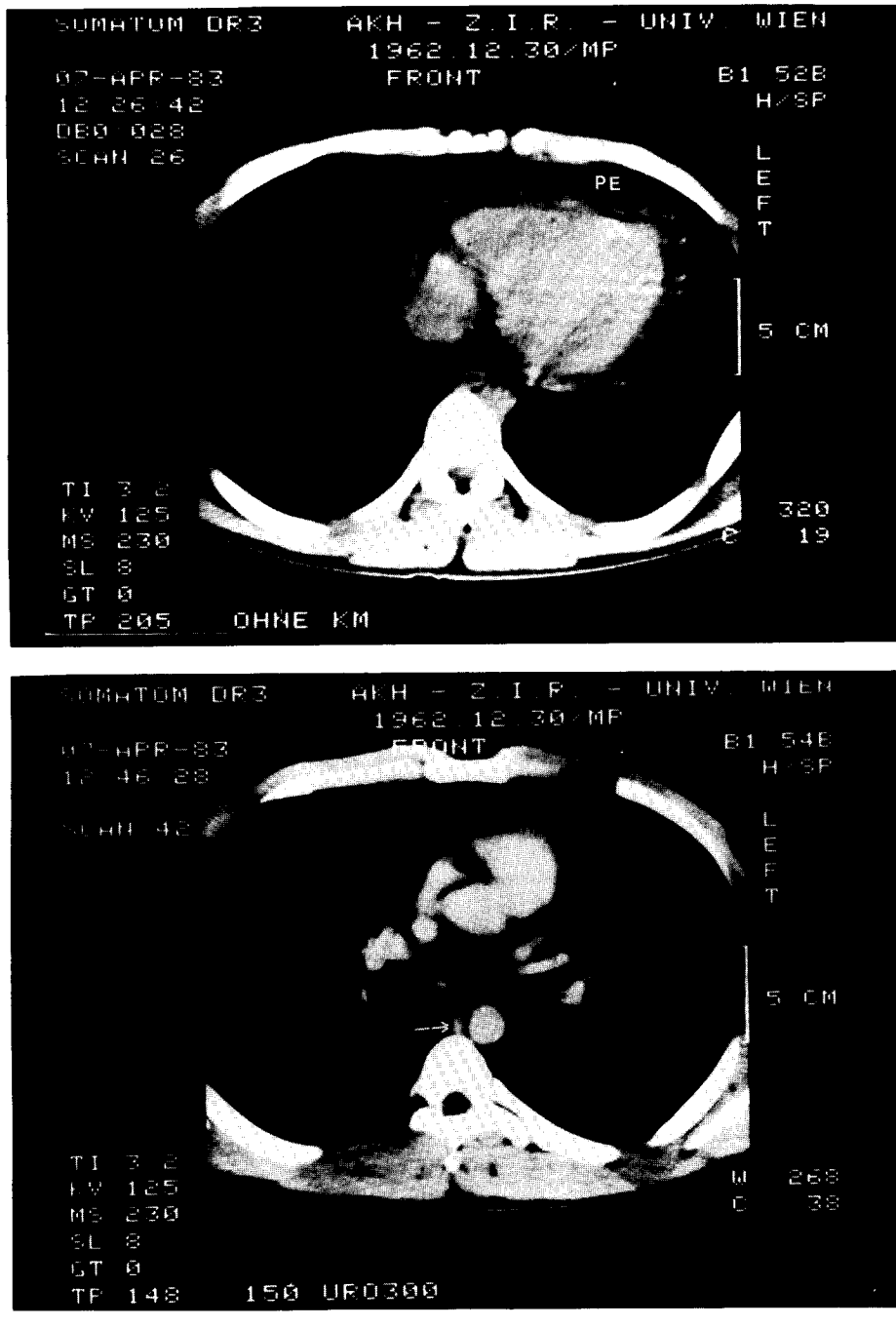

Fig. 4: Mediastinal computer tomography demonstrating (top) pericardial effusion (PE) (density 15 Hounsefield units) and (bottom) a large dilated channel (arrow) adjacent to the aorta. The image of this channel did not enhance after intravenous infusion of contrast (Uromiero 300) suggesting an abnormally large thoracic duct.

aminations. Among these latter patients, two (like our patient) had a prior history of external chest trauma but its relation to chylous pericardium was unclear (5,9). Of 47 patients with "spontaneous" (i.e., non-surgical) chylopericardium the sex distribution was nearly equal with most patients clustered between 15 and 30 years. Only 2 patients were older than 45 years. In contrast to pericardial serous effusion where other severe illness almost always coexisted, chylous pericardium was often an isolated phenomenon and patients were often asymptomatic. Some had dyspnea on exertion but seldom angina or palpitations. In only 5 of 47 patients were there signs of cardiac tamponade. Two-thirds of this group had normal (i.e., not muffled) heart sounds and this unexpected finding has been attributed to chyle and presumably better transmission of sound (10).

The simplest, but nonetheless invasive method to differentiate pericardial effusion is fluid analysis after pericardiocentesis. Echocardiography and radionuclide-angiography are useful non-invasive methods, but cannot differentiate the type of effusion. Computer-tomography is a promising technique to differentiate types of effusion by use of density measurements in HU. Theoretically, a large fat content of chyle should generate less density than a simple transudate ($<$15 HU), but in our experience with other chylopericardium, lymphangiomas and lymphatic cysts (e.g., after kidney transplantation) we found density readings between 10 and 20 HU. Density of chyle depends not only on fat content, but also on protein concentration. Proteins per se demonstrate a density $>$20 HU. Therefore, the density of chyle is variable and depends on the relation between its fat and protein content. To establish a possible connection between the thoracic duct and pericardium different methods have been used. Lymphangiography on occasion may visualize malformations in the mediastinum but the oily contrast medium remains undesirably in lymph nodes for months thereafter (3,9,11,12). Patent blue dye infusion into the peripheral soft tissue, as used in this patient, may signify a major lymphatic-pericardial connection but further anatomic localization is not possible. Other techniques include injections (13) or oral ingestion (4) of tracers such as 131-I-triolein with precordial radioactivity followed for 12-24 hours (4,13). As with dye infusion, however, anatomic delineation of direct lymphatic-pericardial shunting is not possible. In our patient we were reluctant to use lymphangiography because of lack of symptoms and possible side effects. Nonetheless, a major lymph-pericardial communication is likely although not clarified further.

The pathogenesis of "non-surgical" chylopericardium is unclear although two major theories have been proposed. Either abnormal congenital or hamartomatous communication exists between mediastinal lymph vessels and the pericardium (12,14), or obstruction of the thoracic duct with limited collateralization and progressive valvular incompetence and congestion leads to reflux or frank rupture of chyle into the pericardium (1,15).

Almost all previous patients, symptomatic and asymptomatic, have undergone thoracotomy. The prime operative indications included rapid refilling of the pericardium after aspiration (4) or suspicion of a mediastinal mass or malformation (9). Intraoperatively the pericardium was noted to be extremely thin and dilated. Instillation of patent blue dye into the thoracic duct has been advocated to outline a fistula or detect a ruptured lymph vessel (11,16). In most (42 of 47 patients) treatment consisted of a fenestration of the pericardium combined with thoracic duct ligation (16). These patients did well without relapse for up to two years follow-up (5). Four of 47 patients had a pericardial "window" only. Two died of intractable chylothorax and the others were lost to follow-up. Despite this dramatic theraputic difference, reluctance to ligate the thoracic duct has persisted because it is feared that abnormalities of the central lymph trunks contribute to

idiopathic chylopericardium and ligation of the thoracic duct may exacerbate the condition (16). Accordingly, ligation of abnormal patent blue dye-colored lymphatics thought to communicate with the pericardium has been recommended (16). In one asymptomatic 8-year-old girl, treatment consisted of repeated pericardial puncture, cortisone and dietary substitution with medium-chain triglycerides (5). In contrast to long-chain triglycerides, short-chain fatty acids are absorbed directly into the portal vein thereby reducing the amount of chyle formed (9). After four years this patient was well without sign of cardiac silhouette enlargement. A similar non-operative approach has been successful in four patients after "surgically-induced" chylopericardium (9). In light of these observations we opted to treat our patient expectantly. After 15 months the cardiac shadow remains enlarged, although he is still without symptoms.

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