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Primary idiopathic chylopericardium: Report of two cases

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Primary chylopericardium is an extremely rare disease. This report presents two cases of this disease, in a 47-year-old man and 21-year-old woman. Both cases were given diagnosis of primary chylopericardium by chylous pericardial fluid examination and lymphangio-scintigraphy which demonstrated abnormal communication between the left thoracic duct and the pericardial cavity.

Key words: primary chylopericardium, lymphangio-scintigraphy, ^{99m}Tc-HSA-D, SPECT, Sudan III

INTRODUCTION

PRIMARY CHYLOPERICARDIUM is an extremely rare disease[1] with 32 cases reported since the initial report of Groves et al. in 1954.[2] We present two cases of this disease in which it was possible to demonstrate a communication between the thoracic duct and pericardial cavity using lymphangio-scintigraphy.

REPORT OF CASES

Case 1: A 47-year-old man presented with a feeling of persistent chest oppression, and shortness of breath in 1998. Chest X-ray demonstrated the cardiothoracic ratio (CTR: 56%). Echocardiography demonstrated a massive pericardial effusion and diastolic collapse of the right ventricle. Pericardiocentesis produced chylous fluid the biochemical characteristics of which were as follows: specific gravity, 1.04; total cholesterol, 122 mg/d*l*; triglycerides, 181 mg/d*l*; lipoprotein fractions (LDL, 1120 mg/d*l*; VLDL, 725 mg/d*l*; and chylomicrons, 1440 mg/d*l*). A total of 1325 m*l* of chylous pericardial fluid was drained over 5 days, resulting in marked symptomatic improvement. Pericardal fluid sampled after ingestion of a high-fat diet comprising 30 g of margarine and 50 mg of Sudan III on the previous day, became pink rather than chylous, indicating distribution of the ingested Sudan III into the pericardial effusion. Lymphangio-scintigraphy with 740 MBq of ^{99m}Tc-HSA-D (^{99m}Tc Human Serum Albumin Diethylenetriamine Pentaacetic Acid) was injected into the second or third subcutaneous web spaces of both feet. The whole body was scanned 10, 20, 50 min, 2 and 24 hours after the injection. An abnormal accumulation of the isotope was demonstrated in both lungs after 2 hours and migration toward the pericardial cavity after 24 hours on planar and SPECT images (Fig. 1a, b, c). On the same day, CTR on chest X-ray was improved and pericardial effusion had disappeared on echocardiograms. The patient was then discharged to be followed as an outpatient.

Case 2: A 21-year-old woman was admitted with dyspnea, and was noted to have a markedly enlarged cardiac silhouette and bilateral pleural effusions on chest X-ray. Echocardiography demonstrated a massive pericardial effusion, with diastolic collapse of the right ventricle. Pericardiocentesis produced yellowish white and turbid fluid. Microscopic examination demonstrated numerous Sudan III-stained red fat particles in histiocytes, and the biochemical characteristics of the pericardial fluid were as follows: specific gravity, 1.034; total cholesterol, 53 mg/d*l*; triglycerides, 245 mg/d*l*. A total of 1140 m*l* of chylous pericardial fluid was drained over 6 days, resulting in

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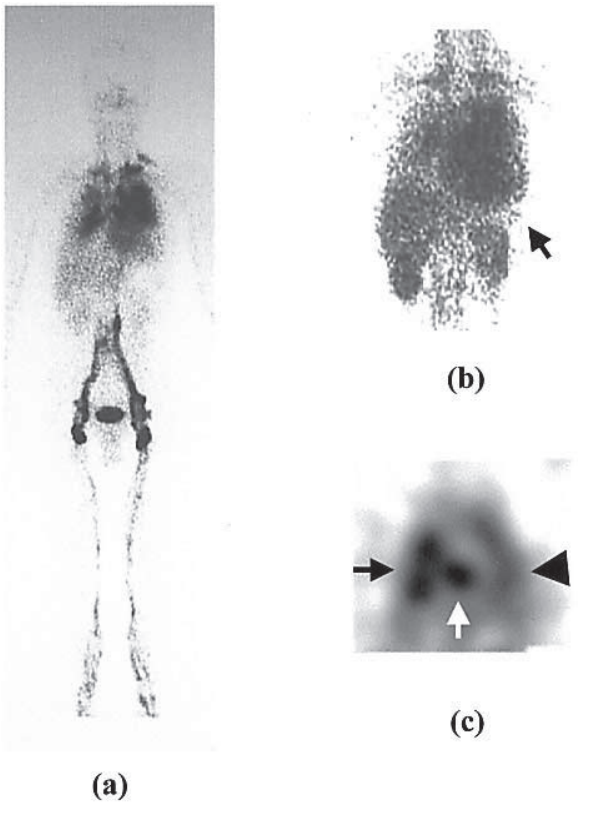

Fig. 1 Radioactivity of ^{99m}Tc-HSA-D was observed at the lung and then migrated to the pericardial cavity. (a) Radioactivity of ^{99m}Tc-HSA-D was observed at the lung. (2 hours after injection) (b) Radioactivity migrated to the pericardial cavity 24 hours after injection. (➡) (c) This image was the heart axial view of ^{99m}Tc-HSA-D SPECT image 24 hours after injection. Radioactivity was observed at the left ventricle (⇨), the right ventricle (➡) and the pericardial cavity (▼).

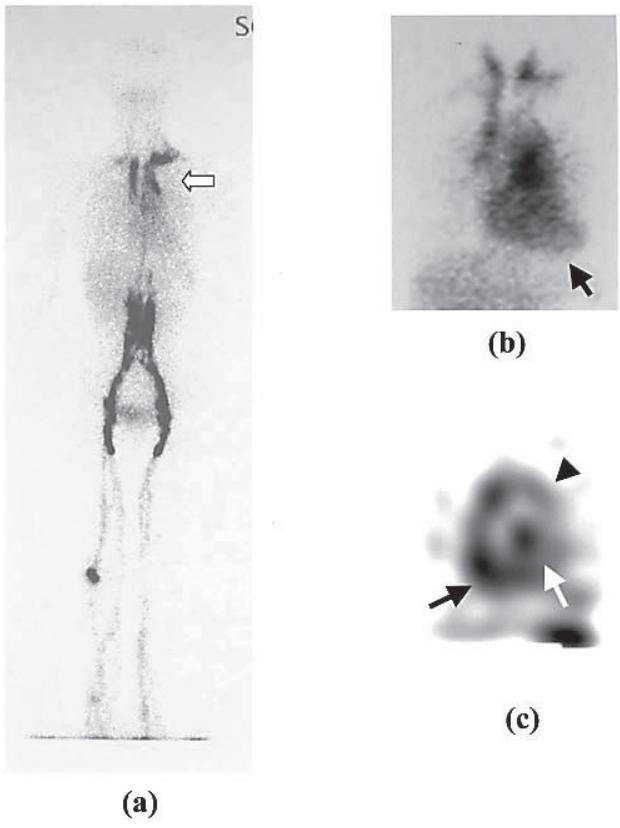

Fig. 2 Radioactivity of ^{99m}Tc-HSA-D was observed at the left brachiocephalic, subclavian veins and the pericardial cavity. (a) Radioactivity of ^{99m}Tc-HSA-D was observed at the bilateral ductus thoracicus, and an abnormal accumulation of the RI in the left brachiocephalic, subclavia vein caused back flow. (20 minutes after injection) (⇨) (b) Radioactivity at the left pulmonary hila then migrated to the pericardial cavity 24 hours after injection. (➡) (c) This image was the heart axial view of ^{99m}Tc-HSA-D SPECT image 24 hours after injection. Radioactivity was observed at the left ventricle (⇨), the right ventricle (➡) and suspect in the pericardial cavity (▼).

improvement of the symptom. Lymphangio-scintigraphy showed a bilateral thoracic ductus and an abnormal accumulation of the radio-isotope in the left brachiocephalic and subclavian veins caused back flow after 20 minutes post injection and migrated toward the left hilus and the pericardial cavity 24 hours post injection on planar and SPECT images (Fig. 2a, b, c). After 34 days, CTR on chest X-ray was improved and the patient was discharged. On the other day, she complained dyspnea again, CTR on chest X-ray was increased to 73%, and she was readmitted. And she repeated readmitted due to recurrence of pericardial effusion[3] by common cold.

DISCUSSION

In an article by Schultz[4] published in 1980, chylopericardium was classified into two categories: primary and secondary. Primary chylopericardium is an extremely rare disease with a total of as few as 30 cases reported during the period up to 1991, and only 7 cases in Japan.[5,6] Generally, the lymph flow through the thoracic duct is as much as 1.38 m*l*/kg per hour,[7] and it is believed that lymphangial abnormalities or abnormal imbalance of pericardial cavity pressure, venous pressure and thoracic-ductal pressure leads to cardiac tamponade.[8] A suspicion of this disease is entertained in patients with a chronic course in the absence of subjective symptoms and positive physical findings, whose condition is accidentally discovered by chest X-ray and in whom massive pericardial effusion is demonstrated on echocardiography, is chylous in appearance and is characterized by lipid component constituents on biochemical tests.

Case 1; Chylopericardium was suspected because distribution of Sudan III dye into pericardial fluid was evident at 24 hours after ingestion of Sudan III and lymphangio-scintigraphy revealed an abnormal accumulation of the radio-isotope at both pulmonary hila with migration toward the pericardial cavity.

Case 2; Chylopericardium was suspected because lymphangio-scintigraphy revealed an abnormal accumulation of radio-isotope in the left vein brachiocephalic and subclavian veins and migrated toward the left hilus region and pericardial cavity. The diagnostic methods reported for chylopericardium include: 1) observation of distribution of Sudan III dye into the pericardial cavity following oral intake of Sudan III along with fat, 2) evaluation of chest radioactivity after an oral dose of ^{131}I-triolein,[9] 3) lymphangio-scintigraphy, and 4) lymphangiography upon thoracotomy. 1) permits observation of Sudan III dye distribution into the pericardial cavity, but the route is indistinct by this method, 2) is not permitted in Japan, and 4) is an invasive procedure. 3) lymphangio-scintigraphy is not invasive and has been demonstrated to facilitate a diagnosis of chylopericardium by abnormal accumulation of the radio-isotope. Using this method, the injected radionuclide can be traced from the lymph vessel to the pericardial cavity noninvasively and over time, hence suggesting its potential efficacy in the diagnosis of chylopericardium. In the therapy, we can select medical or surgical management. But this disease is rare, and so few examinations have been performed. We think first choice therapy is medical management by dietary treatment and think second choice is surgical the therapy, but we consider the age, gender and clinical prognosis which may be worsened by increased pericardial effusion in same cases. It is very important that we should not misjudge the optimal timing of surgical intervention in cases in which this is judged necessary.

REFERENCES

1. López-Castilla JD, Soult JA, Falcón JM, Muñoz M, Santos J, Gavilan JL, et al. Primary idiopathic chylopericardium in a 2 month old successfully treated without surgery. *J Pediatr Surg* 2000; 35: 646–648.
2. Groves LK, Effler DB. Primary chylopericardium. *N Engl J Med* 1954; 250: 520–523.
3. Chavez GM, Rodriguez GR, Gonn HJ. Isolated chylopericardium. *Am J Cardiol* 1973; 32: 352.
4. Schulz W, Schöeberger A, Trüber E, Reifart C, Sarai K, Kaltenbach M. Diagnose and Therapie des Chyloperikards. *Deutsch Med Woch* 1980; 45: 1573–1576.
5. Yoon YS, Shim WH, Chung TS, Lee YS. Primary idiopathic chylopericardium: Report of a case and review of the literature. *Yonsei Medical Journal* 1993; 34: 98–108.
6. Madison WM, Logue B. Isolated (Primary) chylopericardium: Due to anomalous communication with the thoracic duct of unknown causation. *Am J Med* 1957; 22: 825–830.
7. Bessone LN, Ferguson TB, Burford TH. Chylothorax collective review. *Ann Thorc Surg* 1971; 12: 527–550.
8. Usui S, Sakai K, Yamada T, Muraguchi K, Tsukamoto Y, Tei N. An operative case of primary chylopericardium: report of a case and review of the literature in our country. *Kyobu Geka* 1985; 38: 899–904.
9. Savran SV, Ratshin RA, Shirley JH, Naguwa SM, Googman L. Idiopathic chylopericardium: ^{131}I-triolein scan for noninvasive diagnosis. *Ann Int Med* 1975; 82: 663–665.