Chylous Ascites and Pleural Effusion Secondary to Constrictive Pericarditis Presenting With Signs of Lymphatic Obstruction

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SUMMARY

Chylous ascites is a clinical entity characterized by accumulation of milky fluid containing high amounts of triglycerides in the peritoneal cavity. The cause is usually lymphatic obstruction secondary to neoplastic processes. Constrictive pericarditis rarely causes chylous ascites through elevated venous pressure and lymphatic stasis. To the best of our knowledge, there is no report of constrictive pericarditis leading to chylous ascites in a patient presenting with objective lymphangiographic findings of lymphatic obstruction rather than stasis. We present a case of chylous ascites and pleural effusion secondary to constrictive pericarditis presenting with signs of lymphatic obstruction in lymphangiography, in whom complete clinical and laboratory improvement was achieved after pericardiectomy. (Jpn Heart J 2004; 45: 535-540)

Key words: Chylous ascites, Constrictive pericarditis, Lymphatic obstruction

CHYLOUS ascites is a clinical entity characterized by accumulation of milky fluid containing high amounts of triglycerides in the peritoneal cavity. It usually results from lymphatic obstruction. In clinical practice, the most frequent cause of lymphatic obstruction is infiltration or compression of the abdominal or thoracic lymphatics by solid tumors.1,2) In this report, we present a case of chylous ascites and pleural effusion secondary to constrictive pericarditis, presenting with lymphatic obstruction signs in lymphangiography in whom complete clinical and laboratory improvement was achieved after pericardiectomy.

CASE REPORT

A 50-year-old Turkish woman presented with complaints of abdominal swelling and exertional dyspnea which had started 6 months earlier and increased...
in the last month. She did not have a history of liver disease, gastroenteric system disease, tuberculosis, rheumatic disorders, malignancies, or surgery.

On physical examination, her general appearance was not good (Karnofsky Performance Status 60%). Her temperature was 36.2°C, pulse 110 bpm, and arterial blood pressure 110/70 mm/Hg. She had jugular venous distention extending until the mandibular angle in the 30° supine position. Heart sounds were deep and rapid. The costophrenic angles were dull to percussion and lung sounds were faint at the basal level and absent at the costophrenic angles. She had abdominal distention, shifting dullness, and bilateral pretibial pitting edema.

Laboratory analyses were WBC: 9,000/mm³ (77% polymorphonuclear, 14% lymphocytes), RBC: 5,560,000/mm³, hemoglobin: 13.0 g/dL, hematocrit: 37.7%, erythrocyte sedimentation rate: 21 mm/hour, serum glucose: 64 mg/dL, BUN: 30 mg/dL, creatinine: 0.6 mg/dL, triglycerides: 73 mg/dL, total protein: 5.1 g/dL, albumin: 2.8 g/dL, AST: 26 U/L, ALT: 21 U/L, ALP: 611 U/L, and LDH: 405 U/L. Thyroid function tests were normal. Cardiac shadow was within normal limits in the chest X-ray. Nonspecific T-wave abnormalities were detected in electrocardiography.

There was nearly 12 liters of milky fluid in the peritoneal cavity and the biochemical characteristics of the fluid were as follows: glucose: 148 mg/dL, cholesterol: 73 mg/dL, triglycerides: 1008 mg/dL, protein: 4.2 g/dL, albumin: 0.9 g/dL, amylase: 22 U/L, and LDH: 230 U/L. These findings were consistent with chylous ascites. Cultures and direct microscopic examination of the fluid, including mycobacterial, were negative. Cytological examination revealed reactive mesothelial cells only.

The thorax, abdomen, and pelvis were scanned by computerized tomography for the etiology of the chylous ascites. The liver, gallbladder, spleen, other viscera, and main vascular structures were normal, no solid lesions were noted, and there was widespread ascites in the abdominal cavity. Besides these, there were plaque-shaped, dense calcifications in the pericardium and dilatation of the thoracic portion of the inferior vena cava. Echocardiography and cardiac MRI showed diffuse thickening and calcification of the pericardium.

The diagnosis of constrictive pericarditis was supported by cardiac catheterization. Pressure recordings were as follows: pulmonary capillary wedge 21 mmHg, pulmonary artery 46/22 mmHg, right ventricle 50/21 mmHg, right atrium 20 mmHg, and left ventricle 110/21 mmHg. No pathology was noted in the caval veins. Lymphangiography revealed stasis and irregular distribution in the upper paraaortic lymphatics at the level of the 12th thoracic vertebra, with only the initial part of the thoracic duct being opacified. Twenty-four hours later, there was no residual opacity in the thoracic duct and the thoracic lymph vessels, which was interpreted as lymphatic obstruction (Figure 1).
Total pericardiectomy was performed to treat the constrictive pericarditis. The heart was surrounded by a pericardial tissue of almost bone consistency and nearly 10 mm thick (Figure 2). Pathological examination of the pericardiectomy material showed nonspecific pericarditis. Symptomatic clinical improvement was observed at one-month follow-up visit; total clinical and laboratory recovery was achieved in six months.

Figure 1. Lymphangiograms obtained 24 hours apart (A: first, B: 24 hours later).

Figure 2. Pericardiectomy specimen with consistency of bone.
DISCUSSION

A diagnosis of chylous ascites is made when a milky fluid is obtained with paracentesis from a patient with ascites. The triglyceride concentration of the chylous ascites has to be over 200 mg/dL and higher than that of the plasma. Although a triglyceride concentration of 200 mg/dL is sufficient to produce a milky appearance in a fluid, the triglyceride concentrations of these patients are usually well over 1000 mg/dL.1) Our case had ascitic triglyceride levels in excess of 1000 mg/dL.

Chylous ascites is rare and generally results from obstruction of abdominal or thoracic lymphatics because of tumor, trauma, inflammation, or congenital abnormalities.2) Malignancies, especially lymphomas, have been the most frequent reason for chylous ascites in many series.3) Among the studies on the etiology of chylous ascites, malignancy-related obstruction was noted in 87.5% and 87% of patients in the series of Press, et al and Kelley and Butt, respectively.1,4) For this reason, these cases have to be investigated thoroughly for solid mass lesions. Cytological analysis of the ascitic fluid, computerized tomography, and lymphangiography are among the first-line methods of investigation. Lymph node biopsy, even laparotomy, can be employed when the etiology cannot be identified. In our case, solid mass lesions capable of causing compression were ruled out by detailed clinical evaluation and thoracoabdominopelvic tomography. The obstruction observed in the lymphangiography was thought to be secondary to the advanced stasis. This view is further supported by the patient's clinical status, complete cure, and disappearance of chylous ascites and pleural effusion after pericardiectomy.

Constrictive pericarditis causes chylous ascites probably through elevated venous pressure. Elevated venous pressure increases capillary filtration and leads to increased lymph production. Starling has demonstrated that obstruction of the inferior vena cava and portal veins in experimental animals leads to increased lymph production.5) Dumont, et al have reported that the thoracic duct diameter is increased 4-fold and lymph flow is increased 12-fold in patients with advanced stage heart failure and systemic venous hypertension.6) However, the rigid structure of the venolymphatic junction, where the thoracic duct and the subclavian vein meet, prevents the return of the increased lymphatic flow to the systemic circulation at the same rate.6,7) In addition, the elevated venous pressure resists the lymphatic inflow.8,9) Blalock and Burwell have demonstrated the development of increased pressure and dilatation in the thoracic duct in their experiments on the hemodynamics of constrictive pericarditis.10)

It has been shown in animal studies that, in the presence of normal venous pressure, even when the thoracic duct is ligated, the development of chylous
ascites is prevented by collaterals between the lymphatic and systemic circula-
tions. In cases where the lymphatic circulation is blocked, collaterals between
the thoracic duct and the azygous vein or the inferior vena cava provide decom-
pression of the lymphatic system. However, effective collateral circulation does
not develop when the venous pressure is increased. As a result, increased lym-
phatic pressure is reflected to the intestinal lacteals and, with the disruption of
these delicate structures, lymphatic leakage into the abdominal cavity and intesti-
nal lumen ensue. In a postmortem study of a case with dilated cardiomyopathy
and chylous ascites, dilated lymphatic channels and interconnecting, dilated
mucosal lacteals were observed in the submucosae of the duodenum, jejunum,
and ileum.13) Also, there are reports of cases with constrictive pericarditis associ-
ated with protein-losing enteropathy due to ruptured intestinal lacteals.14,15) The
hypoproteinemia in our case is probably the result of this same pathophysiologic
mechanism.

In conclusion, constrictive pericarditis is a very rare cause of chylous ascites
which can be cured totally with surgery.16-19) It should be remembered that con-
strictive pericarditis can lead to signs of lymphatic obstruction in lymphangiogra-
phy, although very rarely, and should be included in the differential diagnosis
especially when a solid mass lesion is not found.

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