Finally, although the initial placement of the catheter seemed to be uncomplicated, the free return of blood and the apparent radiographic location of the tip of the catheter within the central vein do not guarantee that the catheter was inside the vascular space immediately upon insertion; however, the fact that the patient was asymptomatic for 7 days after insertion of the catheter while hyperalimentation fluid was being infused makes an immediate complication of CVC insertion even less likely.

The patient's bilateral hydrothorax contained no red blood cells. Past case reports of hydrothorax as a delayed complication of CVCs are conspicuous for the absence of hemorrhage into the pleural space. It is difficult to understand the absence of a hemothorax associated with delayed perforation by the catheter. One would expect that since the CVC eroded through the vascular space, bleeding at least into the mediastinum would occur. Perhaps a thrombus forms around the site of central venous perforation, thus preventing clinically evident hemorrhage.

In summary, to our knowledge, this case report is apparently the first to describe a hydro pneumo mediastinum and bilateral hydro pneumo thorax as delayed complications of a CVC. The left internal jugular venous approach and the hypertonic infusate likely predisposed to the complication, although central venous rupture has been described with other approaches. When a CVC is inserted, particularly for parenteral nutrition, one must be certain that the tip of the catheter is placed into the SVC, lies parallel to the long axis of the SVC, and is proximal to the right atrium. This position should minimize the possibility of rupture by the catheter. A catheter which lies at an angle to the SVC is at risk for delayed perforation, which may have life threatening consequences. In retrospect, in placing the CVC in this patient, it would have been safer to advance the tip of the catheter further into the SVC and parallel to the long axis of the vein, thus minimizing mechanical forces on the venous wall. Finally, even a catheter composed of relatively flexible material such as silicone elastomer can rupture through the vascular space.

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Chylous Pleural Effusion Associated With Primary Lymphedema and Lymphangioma-like Malformations*

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We describe a patient with a chylous pleural effusion associated with primary lymphedema of his right leg and abdominal wall. On evaluation a generalized, severe hypoplasia of the lymphatic system turned out to be associated with hyperplastic, lymphangioma-like malformations.

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Although primary lymphedema of the extremities is known to be associated with pleural exudates,1,2 chylous effusions are extremely rare. To our knowledge, only three well documented cases have been published.3,5 We report a fourth case, not only remarkable because of the response on therapy, but also because of a thus far unreported association of a generalized hypoplasia of the lymphatic system and lymphangioma-like malformations.

CASE REPORT

A 28-year-old man was referred to our hospital because of a gradually progressive lymphedema of his right leg, which had started at the level of the knee after a minor sports-induced trauma at the age of 15 years. Thereupon, his lower leg became edematous. In later years, his thigh, scrotum, left upper thigh, and abdominal wall also became affected. The growth of the right leg lagged behind. Complaints about shortness of breath on exertion started 3 months before admission. The complaints were most prominent in the early morning and diminished during the day. He had no other respiratory symptoms, fever, night sweats, weight loss, or gastrointestinal complaints. There was no family history of congenital lymphedema, and no history of infection or trauma during pregnancy. The delivery had been normal, as was the patient's development until his 15th year.

On physical examination, the right leg was shorter than the left and more voluminous. On palpation, there was no temperature difference between both legs. The edema was nonpitting. His arms and nails showed no abnormalities. The findings from physical examination of the chest were remarkable for dullness and decreased breath sounds at the right lung base. Being most prominent in the early morning, this area was noticed to become smaller during the daytime.

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Chylous Pleural Effusion, Lymphedema and Lymphangioma-like Malformations (Bresser et al)
Lymphangiography of the right arm appeared to be hypoplastic. Lymphangiography of the left arm was technically impossible, because of an extreme macroscopic hypoplasia.

Chest radiography confirmed the presence of a large pleural effusion at the right side; also, a small amount of fluid was present on the left (Fig 2). By computed tomography of the thorax, no other abnormalities were detected. Chemical analysis of the pleural fluid showed a protein content of 3.1 g/dl (serum, 5.7 g/dl), a cholesterol content of 1.5 mmol/L, and a triglyceride content of 3.0 mmol/L (serum, 5.2 mmol/L and 1.6 mmol/L respectively); chylomicon were detected. These findings were consistent with chyle or at least the presence of an important chylous component. The findings from bacteriologic and cytologic examinations were normal. Subsequent computed tomography of the abdomen and pelvis revealed a tumor mass presacral at the right side with usurpation of the bone. On magnetic resonance imaging, this turned out to be a process in the bony structures of the right pelvis, with extension into the presacral soft tissues. By needle biopsy of the right iliac crest, only lymph was aspirated, containing more than 99 percent lymphocytes. Extraction of bone or marrow was not achieved. These findings are consistent with the presence of a lymphangioma-like malformation.

Thoracocentesis failed to clarify a specific site of leakage, and no dilated lymphatic vessels were seen. Tube drainage was performed, whereupon 15 L of fluid were drained within the next 24 h. At the same time the leg edema resolved almost completely. Subsequently, we successfully performed chemical pleurodesis with quinacrine, combined with a low-fat, medium-chain triglyceride diet, in order to slow down the chyle flow. The patient did very well for almost 2 years. The pleural effusion on the right did not recur. The patient was suffering from a slight, stationary edema of his right leg, for which he had to wear elastic stockings.

After 2 years the patient started complaining again about shortness of breath, caused by a gradually progressive exudate on the left. A chylous component could not be proven. Initially, we performed repeated thoracentesis in order to relieve the dyspnea. Finally, the edema of the right leg also increased. Because of severe dyspnea, not responding to repeated thoracentesis, we had to perform tube drainage, which caused a partial pleurodesis. After drainage the leg edema resolved within 1 day. Six months later, the patient is still doing well. If recurrent pleural fluid causes symptoms, we will consider implantation of a pleuroperitoneal shunt.

**DISCUSSION**

Primary lymphedema of the extremities can be classified based on lymphangiographic findings, according to Kinmonth et al., as hyperplastic, hypoplastic, or aplastic. In the case of hyperplasia, numerous large and tortuous incompetent, sometimes called “varicose,” lymphatic vessels are present. Lymphangiography will show small lymphatic vessels, reduced in number in the case of hypoplasia and a total absence of vessels in the case of aplasia.

Pulmonary manifestations in primary lymphedema are mainly characterized by pleural exudates. This combination may be associated with yellow dystrophic nails, a triad often referred to as the “yellow nail syndrome,” first described by Samman and White in 1964. The pleural exudate is thought to be secondary to the dysfunction of pleural and pulmonary lymphatic vessels. Once present, the exudate tends to persist and may reaccumulate within days or several months after thoracentesis.

Chylous pleural effusions in association with primary lymphedema, as seen in our patient, are rare; however, mediastinal lymphangiomas are known to be associated with
Chyloous pleural effusions. Lymphangiomas are rare malformations, considered to be developmental defects of the lymphatic system. In our patient, no mediastinal lymphangioma was found, but such malformations were present in the right pelvic bones and subcutaneously at the left hip. Lymphangiomatosis of the bone is rare; it may be limited to a solitary bone or be disseminated throughout the skeleton. In hyperplastic lymphedema, lymphangioma-like malformations have been described; however, there are no reports of such malformations in association with hypoplasia or aplasia.

Treatment of chylothorax is difficult. Chemical pleurodesis in combination with a low-fat, medium-chain triglyceride diet to slow down chyle flow or total parenteral nutrition is generally considered to be the initial approach. If this fails, surgical treatment by means of pleurectomy or ligation of the thoracic duct is indicated. In our patient, pleurodesis was very successful. Even the edema resolved almost completely, without recurrence for 2 years. Pleurodesis did not influence the lymphatic drainage from the lower parts of the body, since no reaccumulation of the lymphedema was observed. We suppose that there must be an alternative, as yet unknown, transport of lymph eventually draining on central lymphatic vessels.

In summary, we found primary lymphedema to be associated with a chyloous pleural effusion that was successfully treated by pleurodesis. Lymphangiographic studies revealed aplasia (right leg), hypoplasia (both arms), and hyperplastic, lymphangioma-like malformations (right pelvis and left hip). We conclude that our patient presents a thus far unreported combination of aplasia, hypoplasia, and hyperplasia of the lymphatic system, in association with a chyloous pleural effusion.

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Cytomegalovirus Pneumonitis
An Unusual Cause of Pulmonary Nodules in a Patient With AIDS*

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A patient with AIDS and a history of non-Hodgkin’s lymphoma developed a generalized illness associated with the appearance of multiple pulmonary nodules on a chest radiograph. Cytomegalovirus infection was demonstrated by needle aspiration cytology. The patient’s symptoms and radiographic abnormalities resolved completely on ganciclovir therapy. This unusual case (1) broadens the differential diagnosis of nodular pulmonary disease in patients with AIDS and (2) suggests that cytomegalovirus can cause clinically significant lung disease which may respond to standard antiviral therapy in patients with AIDS.

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