Large Pericardial and Pleural Effusions Associated with Familial Lymphedema

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Lymphedema, usually confined to the legs, is attributed to abnormal drainage of the lymphatic system. Hereditary forms of lymphedema, also referred to as Meige-Milroy disease, are usually of autosomal dominant transmission [1]. Hereditary lymphedema is classified into three types according to the age of presentation: namely infantile hereditary lymphedema (Milroy disease); lymphedema precox, or juvenile hereditary lymphedema, occurring at puberty (Meige's disease); and hereditary lymphedema tarda, occurring after the third decade of life [2].

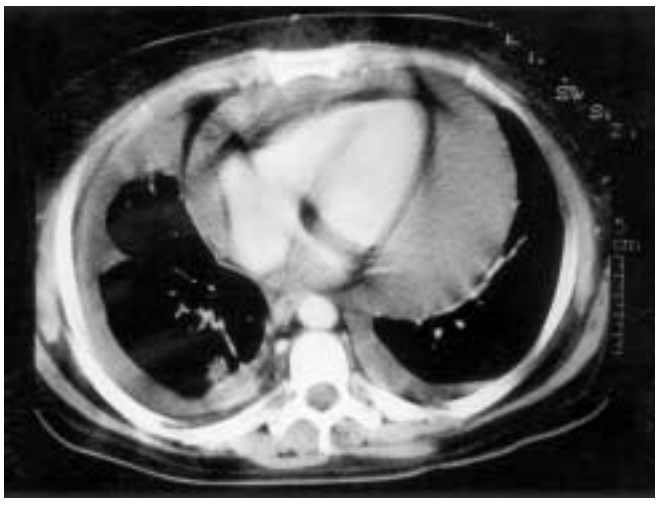
Association of lymphedema with chylothorax and yellow nails has been reported in the literature in both familial and sporadic cases [2]. However, cases of the simultaneous occurrence of lymphedema, chylothorax and pericardial effusion have been reported only sporadically [3–5]. We describe here the first case of a familial triad of lymphedema, pleural and pericardial effusion.

Patient Description

A 40 year old Caucasian woman was admitted to the Medicine Ward due to recurrent cellulitis of the legs. Her past history was relevant for swelling of the right leg since childhood. Large pleural effusion was noted for the first time at a previous admission 10 years earlier. On that occasion 3 L of chylous fluid were drained (Sudan III positive, total protein 4.8 g/L, triglyceride 443 mg/dl, cholesterol 90 mg/dl). A lymphography performed on the opposite limb did not reveal any abnormalities in lymphatic drainage up to the thoracic duct.

At the present admission, physical examination revealed a normal appearing young woman, with signs of cellulitis and lymphedema in the right leg below the knee. Body temperature was 39°C, pulse 100 beats/min, blood pressure 100/80 mmHg, and respiratory rate 20. Pulsus paradoxus was absent, and the neck veins were not distended. Chest examination showed dullness and decreased breath sounds on the right side; the heart sounds were faint. In the abdomen the liver and spleen were not palpable. Examination of the lower extremities revealed no abnormalities apart from the right leg lymphedema; the nails were of normal color and texture. The electrocardiogram was normal. Chest X-ray showed a large right and a small left pleural effusion; the cardiac silhouette was markedly enlarged (cardiomegaly of lesser extent was present on the chest X-ray taken 10 years earlier). Chest computerized tomography scan showed massive pericardial and bilateral pleural effusions [Figure]. A transthoracic echocardiogram revealed a large pericardial effusion with signs of tamponade (collapse of right atrium and right ventricle, and respiratory variation of mitral diastolic flow). Despite the pathologic echocardiographic and radiographic findings, the patient was free of cardiorespiratory symptoms. Pleural (800 ml) and pericardial (500 ml) drainage was performed, demonstrating chylous fluid that contained chylomicrons and triglycerides (207 and 28 mg/dl respectively).

The patient was discharged after becoming afebrile. At the follow-up visit one year later, she remained asymptomatic despite a recurrence of pleural and pericardial effusions as revealed by



Chest CT scan showing large pericardial and bilateral pleural effusion.

subsequent chest X-ray and echocardiogram.

According to the family history the patient's parents (of Jewish Moroccan origin) were not consanguineously related. Medical information on the proband's mother (deceased at age 70) was obtained from hospital records. She had been diagnosed with leg lymphedema, recurrent pleural effusion and pericardial effusion, and was treated for congestive heart failure and suspected tuberculosis. The proband's siblings were examined by one of the authors. While the proband and her mother presented with the full syndrome (right leg lymphedema, pleural effusion, pericardial effusion), four of her seven siblings had one or more of the above-mentioned signs: three had leg lymphedema and another also had pericardial effusion.

Comment

To the best of our knowledge this is the first case of familial pericardial effusion associated with pleural effusion and leg lymphedema. Our case has similarities to but also some differences from Meige's (yellow nail) syndrome. It is an autosomal dominant trait and presents with lymphedema of the lower limbs as well as pleural effusion. It differs in that the nails are not yellow (an optional aspect of the syndrome) and, most importantly, some members of the family showed large pericardial effusion.

Although some members of the family presented the full syndrome (proband and mother), others showed partial features. Altogether, five of the eight siblings showed some features of the syndrome. Thus, our data suggest autosomal dominant inheritance with variable penetrance. This is in contrast to previous descriptions of large pericardial effusion with or without pleural effusion and lymphedema, which were all sporadic [3–5].

The etiology of the chylothorax and pericardial effusion described is likely due to anomalous lymphatic drainage in these patients. The absence of clinical tamponade in this case is explained by the gradual accumulation of chylous fluid into the pericardial space, which allowed adaptation of the pericardium to this condition.

We conclude that in patients presenting with lymphedema, the coexistence of pleural and pericardial effusion should be sought and a comprehensive family history obtained.

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