Chylothorax and Milroy Disease

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Declaration of conflicting interests

The authors declare that there is no conflict of interest.

Abstract

Background: Chylothorax is a type of pleural effusion resulting from accumulation of chyle in the pleural cavity. Milroy's disease is a congenital disease characterized by lymphedema, mostly in the legs and is caused by congenital abnormalities in the lymphatic system. It is a rare presentation of this disease. A 30 year old boy, accountant by profession in Saudi Arabia, presented to me in clinic with recurrent right sided pleural effusion. Some 5 months back pleural effusion was aspirated and some medicine were advised. We planned pleural biopsy, pleural fluid R/E and cytology. During procedure the fluid was found to be thick white and blood mixed. Biopsy was postponed and therapeutic tape and fluid was sent for triglycerides level as prime suspect of being chylothorax. Triglycerides were 759 mg/dl suggestive of Chylothorax.

Key Words: Chest Disease; Pleural Effusion; Biopsy

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Introduction

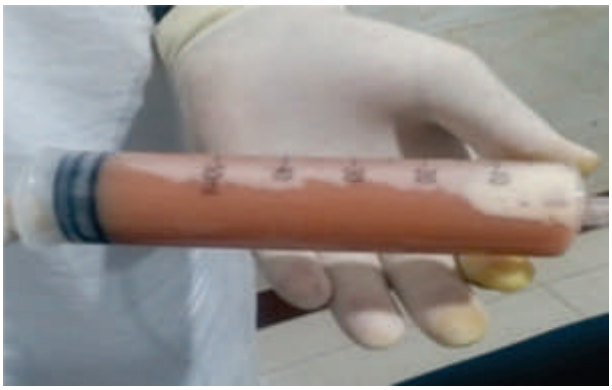
Chylothorax is a type of pleural effusion resulting from accumulation of chyle in the pleural cavity. Milroy's disease is a congenital disease characterized by lymphedema, mostly in the legs and is caused by congenital abnormalities in the lymphatic system. It is a rare presentation of this disease.

Case Report

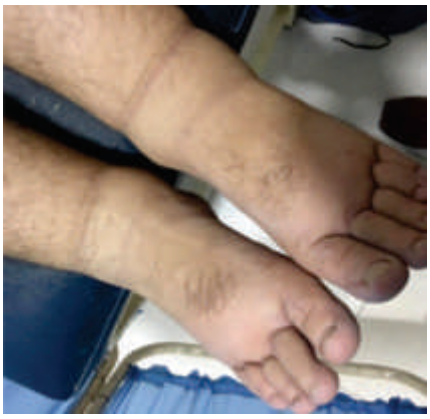
A 30 years old male presented to my clinic with complaint of recurrent pleural effusion. He is accountant by profession in Saudi Arabia. He was diagnosed as a case of Milroy disease since childhood with bilateral pedal edema. He used to apply pressure stockings, manual drainage and foot end elevation as part of management of his disease. From the last 6 to 8 months he has right sided chest pain and shortness of breath. He was diagnosed as case of pleural effusion and therapeutic tape was done in Saudi Arabia.

This time again, he presented with shortness of breath and right sided chest heaviness. He was not febrile and general physical examination was normal except bilateral pedal edema up to the shin. Percussion was dull and there were decreased breath sounds on right side of the chest auscultation. X-ray showed right sided homogenous opacity occupying around half of hemithorax. Pleural biopsy was planned and investigations were sent.

He was negative hepatitis B and C and HIV serology. LFTs, RFTs, CBC were all in normal range. At the time of procedure when local anesthetics were given and aspiration was done, the fluid was found to thick white stained with blood and non-odorous. Chylothorax was suspected and the plan was changed to chest drain insertion but the patient was not ready so therapeutic aspiration was done and fluid about 60ml



The patient was sent on antibiotics and advised pleurodesis, fat free diet and to continue compression stocking along with foot elevation and manual drainage.



was kept for stagnation and 10 ml for triglycerides level (chylomicron test was not available). About 1200 ml of fluid was drained. Test result showed Triglycerides level to be 759 mg/dl. After one hour of stagnation the fluid was homogeneous in contrast to empyema.

Discussion

Milroy's disease is a familial disease that is characterized by lymphedema. Common site is the legs. It is caused by congenital abnormalities in the lymphatic system. Disruption of the normal drainage of lymph leads to fluid accumulation and hypertrophy of soft tissues.[1,2] Milroy's disease is also known as primary or hereditary lymphedema type 1A or early onset lymphedema. It is autosomal dominant condition caused by a mutation in the FLT4 gene.[3] It is more common in women. FLT4 codes for VEGFR-3 which is implicated in lymphatic system development.

Chylothorax is one of the rarely reported complications of this disease in literature.[4] It presents with chest heaviness, pain and shortness of breath. Some people find this as an accidental finding. The most common causes of chylothorax are trauma and neoplasms, whereas spontaneous chylothorax is infrequently encountered. LAM (lymphangioleiomyomatosis) is another cause in childbearing age females.[5] Small subset of patients with primary lymphedema will present chylous complications of their disease such as chylous ascites, protein-losing enteropathy, chyluria, chylometrorrhea, or chylous vesicles of the lower extremity. Pleural effusion is attributed to a dysfunction of the pleural and pulmonary lymphatics.[6]

Pauwels and colleagues have reported a case of congenital lymphedema associated with chylothorax developing at an adult age.[7] The association of congenital lymphedema and chylothorax is found in a spectrum of generalized congenital lymphatic malformations. Generalized lymphatic disorders have also been described in patients with hereditary lymphedema.

Diagnosis relies on a high index of suspicion. Detailed history, physical examination and chest CT should be used to exclude other common pathologies. Lymphangiography may be helpful. Treatment in this case is to decrease chylous production by restricting fat rich diet and pleurodesis.

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