Spontaneous idiopathic chylothorax in adults

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Chylothorax refers to the presence of chyle in the pleural cavity as a result of thoracic duct disruption. It is either spontaneous or traumatic. Typically, it appears as a milky, opalescent odorless fluid, rich in triglycerides. Herein, 2 cases of spontaneous idiopathic chylothorax are reported, with a brief review of the literature.

A 37-year-old Saudi man, who was admitted for investigation of a left basal opacity that was detected during medical fitness screening. He reported nocturnal episodes of dry cough and dyspnea with spontaneous resolution, however, he denied any medical treatment. On examination vital signs were within normal limits. There were old cautery scars over the right upper quadrant of the abdomen. Chest examination demonstrated signs of a left-sided pleural effusion. Chest radiography showed a left basal homogenous opacity suggestive of pleural effusion and right upper lobe linear shadow suggestive of azygus fissures **(Figure 1)**. Blood investigations including complete blood count (CBC), erythrocyte sedimentation rate (ESR), liver and renal function tests were all within normal limits. Thoracentesis yielded 800 ml of a pinkish milky fluid, its analysis confirmed the presence of chyle with high triglycerides and low cholesterol level **(Table 1)**. Cytology of pleural fluid revealed lymphocyte predominance, negative for malignant cells. Mantoux test was carried out and direct smear and culture for acid fast bacilli were negative. Abdominal ultrasonography (US), chest and abdomen computed tomography (CT) did not show additional abnormalities. Bronchoscopy was normal, apart from a mild but diffuse inflammation of bronchial mucosa. Lymphangiography showed ill defined thoracic duct, dye filled lymphatic channels, diffusely, with a leak at the level of carina. Conservative management, which consisted of bed rest, total parenteral nutrition and tube thoracostomy had failed, so he underwent left-sided thoracotomy with ligation of lymphatic varicosities. The post operative course was uneventful, follow-up visits at 2, 6 months and one year after discharge did not show recurrence.

A 30-year-old Sri Lankan house maid, was admitted for investigations of left basal opacity detected on chest radiography, carried for medical fitness screening. Past history was unremarkable. The patient was asymptomatic. On examination, vital signs were within normal limits, chest examination revealed signs of left-sided pleural effusion. Chest radiography showed left basal homogenous opacity suggestive of pleural effusion and a calcified nodular shadow in the right upper zone (**Figure 2**), abdominal US, chest and abdomen CT did not show any further abnormalities. Blood investigations, including CBC, ESR, liver and renal function tests were within normal limits. Thoracentesis yielded 900 ml of odorless, milky fluid, its analysis confirmed the chylous nature with high triglyceride and low cholesterol level compared with blood levels (**Table 1**). Pleural fluid cytology showed predominance of lymphocytes. Cytology for malignant cells, Mantoux test, revealed direct smear and culture for acid fast bacilli were all negative. Management consisted of

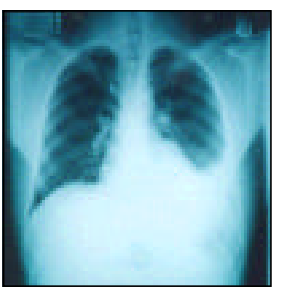

Figure 1 - Postero anterior chest x-ray shows left-sided pleural effusion and right upper lobe linear shadow suggestive of azygus fissures.

Table 1 - Comparative analysis of blood and pleural fluid.

Parameter	Reference Range*	Patient 1		Patient 2	
		B	F	B	F
Cholesterol	3.6 - 8.1	3.86	2.12	5.54	3.06
Triglycerides	0.55 - 3.21	0.53	7.65	1.52	18.3
Total protein	64 - 83	75	55	80	64
Glucose	3.89 - 5.83	4.9	6	5.5	5
LDH	210 - 420	230	182	72	176

* Reference range in International System of units for blood (B) and Pleural fluid (F). LDH - lactate dehydrogenase

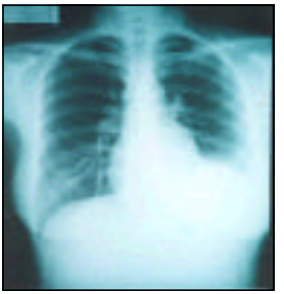

Figure 2 - Postero anterior chest x-ray shows left-sided subpulmonic pleural effusion.

bed rest and low fat diet. A second thoracentesis was performed 3 days later, it yielded 650 ml of grossly milky fluid. The patient was discharged 10 days later on the request of the sponsor against medical advice, however, chest radiography did not show an increase of effusion.

Spontaneous chylothorax in adults is a rare clinical condition. The most common causes are malignant tumors, where lymphomas represent approximately 75% of cases.[1] Among others, rare causes are pulmonary lymphangiomyomatosis[2] and tuberculosis.[3] Chylothorax is labeled idiopathic if no cause could be identified. Most cases are probably related to minor trauma such as a bout of severe cough, vomiting, stretching or hiccupping after the ingestion of fatty meals. The clinical and radiographic features of chylothorax are non-specific, and diagnosis could not be suspected before thoracentesis. The possibility of chylothorax should be considered whenever the turbid or blood-stained fluid remains turbid after centrifugation. Patients with chylothorax have a pleural fluid triglyceride level above 110mg/dl (1.24 mmol), a ratio of the pleural fluid to the serum triglyceride of greater than one, and a ratio of the pleural fluid to the serum cholesterol of less than one. If doubts persist after these measures, the demonstration of chylomicrons by lipoprotein analysis will confirm the diagnosis.[4]

Management of spontaneous chylothorax, depends on the etiology, the general condition of the patient, the speed of reaccumulation and local facilities. Radiotherapy or chemotherapy might control the chylothorax in lymphoma or metastatic carcinoma, but in case of failure, pleurodesis should be tried. Other examples of etiologic treatment are corticosteroids in sarcoidosis and specific antibiotics in tuberculosis. Conservative management of idiopathic chylothorax that is based on bed rest, low fat diet supplemented with medium chain triglycerides or total parenteral nutrition along with repeated thoracentesis or tube thoracostomy, is usually successful[5] suggesting a minimal laceration of the duct. Surgical management by ligation of the duct either by exploratory thoracotomy or video assisted surgery can control most cases of chylothoraces.[6]

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