Letter to the Editor—II

Bilateral Chylothorax Secondary to Acute Lymphoblastic Leukemia (ALL)

Sir,

Chylothorax, the presence of chyle in the pleural space, is infrequently associated with lymphoma. However, association with acute lymphoblastic leukemia is rare.1 A 12 years old male child was admitted to our department with the complaints of breathlessness, fever, loss of appetite and bone pains for 1.5 months. The resting pulse rate was 102/min and his respiratory rate was 26/min. His general examination revealed pallor and multiple, bilateral lower cervical lymph nodes (1 x 1 cms, firm and palpable) and bilateral pleural effusion. On aspiration pleural fluid was milky colour suggesting chylothorax. Pleural fluid protein-5.2 gm%, sugar 55mg%, total leukocyte count 7,800 cells/mm³, differential: neutrophils 35%, lymphocytes 65%, pleural fluid triglyceride: 550.6 mg %, pleural fluid cholesterol: 30.8 mg %. Pleural fluid for malignant cytology- negative. Pleural fluid adenosine deaminase (ADA)- 18 IU/L. Culture for pyogenic organism was sterile. Mantoux test showed no induration. Serum triglyceride and Serum cholesterol were 84.5 mg% and 89.1 mg%, respectively.

He was put on conservative management along with low fat diet but effusion re-accumulated and repeat thoracentesis was performed. Meanwhile his Blood examination was done that revealed Hb 8.5 gm%; WBC-56,900/mm³; differential lymphocytes 91%, neutrophils 9%; platelets 54,000 /mm³. Bone marrow examination revealed: hyper cellular marrow; 48 % blast cells suggestive of acute lymphoblastic leukemia L1 subtype. His renal and liver function test were within normal limits. CT scan thorax revealed bilateral pleural effusion (more on left side) with mediastinal lymphadenopathy.

A diagnosis of acute lymphoblastic leukemia with chylothorax was made.

Comments:

Chylothorax is defined as the accumulation of chyle-containing lymphatic fluid resulting from obstruction or disruption of the thoracic duct. Sassion et al2÷ divided the causes of chylothorax into four major categories: tumour, trauma, idiopathic, and miscellaneous. Malignancy accounted for approximately one half; lymphoma being the most common malignancy (almost 75%) followed by bronchogenic carcinoma.3,4

Chylothorax usually develop as the result of extrinsic compression or direct invasion of the thoracic duct by a tumour or as the result of obliteration of the lymphatics following radiation therapy.3,4 An extrinsic obstruction or an infiltration of the thoracic duct causes an increase in back pressure. This elevated pressure promotes dilatation of collateral channels and renders lymphatic valves incompetent, which produces regurgitant flow of chyle. Consequently, seepage may occur through erosion or perforation of a diseased thoracic duct or one of its tributaries into the mediastinum. Mediastinal chyle then penetrates the mediastinal pleura, producing a chylothorax; alternatively, leakage may occur directly into the pleural space from the dilated intrapulmonary lymphatics because of the regurgitant flow from the bronchomediastinal trunk or posterior intercostals lymphatics.3,4

Diagnosis of chylothorax is established via direct analysis of the pleural fluid. The fluid is characteristically “milky” in appearance,
although not all milky effusions are chylous in nature and not all chylous effusions are milky. Best way to establish the diagnosis of chylothorax, is to determine the concentration of triglyceride in pleural fluid. The triglyceride concentration greater than 110 mg/dl (in our case it was 550.6 mg %), a ratio of pleural fluid to serum triglyceride of greater than 1.0 (in our case it was 6.52), and ratio of pleural fluid to serum cholesterol of less than 1.0 (in our case it was 0.35) usually confirms chylothorax. Chylothorax will be excluded if pleural fluid triglyceride concentration is less than 50 mg/dl. However, in case of levels in between 50 to 110 mg/dl, a lipoprotein analysis of pleural fluid should be performed and demonstration of chylomicrons in fluid confirms the diagnosis of chylothorax.

The first step in the management of chylothorax demands a review of the history and physical examination. Since lymphoma is the most common cause of chylothorax in the non-traumatic etiology, a computed tomography of the chest and abdomen should be performed to evaluate the mediastinum and abdominal lymphadenopathy. Primary treatment in case of chylothorax should be directed towards correction of malnutrition and compromised immunologic status which is due to repeated pleural fluid aspirations of chyle with its high levels of protein, fat, electrolytes and lymphocytes. The defect in thoracic duct often closes spontaneously in case of traumatic injury. In case of severe dyspnoea, placement of pleuro-peritoneal shunt or chest tube drainage is mandatory. If chylothorax persist for more than four weeks, consideration should be given to surgical exploration with ligation of the thoracic duct. In some situation it may be impossible to identify the duct, especially when there is malignancy or radiation fibrosis obscuring the duct. In such situation, other modalities, including pleurodysis and pleurectomy, may be used. Finally, other nontraumatic chylous effusion may be amenable to medical therapy. In lymphomas or other neoplasms responding to chemotherapy/radiation therapy effusion may be resolved.

REFERENCES:

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