Abstract

Pleural disease in Non-Hodgkin’s Lymphoma is well documented and commonly presents with pleural effusion in 20% of patients. However, solid pleural involvement is less common and is usually a secondary event. Primary pleural lymphomas are extremely rare. Hereby we report a rare case of primary pleural lymphoma presenting as chylothorax.

Introduction

Malignant lymphoma of the pleura is mostly associated with chronic pleural inflammation.

Two types of primary pleural lymphomas have been described—the body-cavity based lymphoma (in HIV patients) and the pyothorax associated pleural lymphoma (in tuberculosis patients). The second entity, i.e. pyothorax associated pleural lymphoma is extremely rare. Pyothorax associated pleural lymphomas tend to present as a mass lesion consisting of a high grade NHL of B cell origin. Most cases have been described in Japan and Tazuko et al have experienced only six cases in 18 years. It has been postulated that the intense stimulation of B lymphocytes by asbestos and the decrease in the number of circulating T lymphocytes may lead to lymphoid/plasma cell hyperplasia.

Case History

18 year old boy from Palakkad got admitted in our pulmonology ward with left sided chest pain and mild exertional dyspnea for past 6 months. No history of fever, cough or breathlessness. No other significant illness in the past.

At the time of admission, he was conscious, oriented and afebrile. No dyspnea, clubbing, palor, lymph node enlargement or cyanosis was seen. Vital parameters were normal. Chest examination revealed features of left sided pleural effusion. There was no hepatosplenomegaly other system examination was unremarkable. He had left hydrocoele for which no treatment was sought before.

His blood investigations revealed Hb of 11.4 gm/dl and eosinophilia. Absolute tosinophil count: 750 cells/mm³ and ESR of 50 mm in first hour. Chest X-ray (Figure 1) showed evidence of pleural effusion on left side without parenchymal lesion or hilar adenopathy. Ultrasonography of external genitalia showed hydrocoele with normal testis and epididymis. Considering eosinophilia, hydrocoele and chylosus effusion, filarial antibody test was sent for and turned out to be positive.

Mantoux test was negative. Pleural fluid aspiration drained 600 ml of milky fluid and its study revealed 200 cells/mm³ and ESR of 50 mm in first hour. Chest X-ray (Figure 1) showed evidence of pleural effusion on left side without parenchymal lesion or hilar adenopathy. Ultrasonography of external genitalia showed hydrocoele with normal testis and epididymis. Considering eosinophilia, hydrocoele and chylosus effusion, filarial antibody test was sent for and turned out to be positive.

CT scan of the thorax was done which showed only left sided pleural effusion and did not detect parenchymal lesions or mediastinal pathology (Figure 2). Lymphangiogram showed no occlusion of major lymphatic vessels.

He was initially treated with di-ethyl carbamazine and other supportive measures including steroids but with poor response. Further he developed rapid recollection of the fluid which required repeated aspirations. Cytology of the fluid was repeatedly negative for malignant cells. During the hospital stay, he developed significant loss of weight and edema.

Considering his poor response to treatment, ATT was given empirically with oral prednisolone. But he presented again after 3 weeks without any satisfactory results. Finally pleural biopsy was done which revealed primary pleural NHL of diffuse large B cell variety (Figure 3). Thus the final diagnosis was -Primary pleural Non-Hodgkin’s lymphoma.
Hodgkin’s Lymphoma of diffuse large B cell variety presenting as chylothorax.

Discussion

Chylous pleural effusions are defined by the presence of chyle in the pleural space and usually result from disruption or obstruction of the thoracic duct. Chyle (from the Latin word chylus, or juice) consists of chylomicrons and very low density lipoproteins absorbed from the small intestine, secreted into intestinal lymphatics, and moved into cisterna chyli and then to thoracic duct. Chyle can accumulate in either the left or right pleural space, depending on where the thoracic duct is disrupted. If the disruption occurs below the fifth thoracic vertebra, then right-sided chylothorax occurs; and when above the fifth thoracic vertebra, then chylothorax is left-sided. These effusions were originally described in 1633 by Bartolet, and the diagnosis was initially based on the characteristic appearance of the fluid as demonstrated by lipoprotein electrophoresis which is generally considered the criterion standard in the diagnosis of chylothorax. Although lipoprotein electrophoresis can be performed on the fluid specimen, a simpler assay of quantifying triglyceride levels is widely used to document the presence of a chylous pleural effusion. Pleural fluid triglyceride level greater than 110 mg/dL proves it to be chylous. Conversely, a triglyceride level less than 50 mg/dL for a pleural effusion virtually excludes the diagnosis of chylothorax. These criteria continue to be used today in the diagnosis of chylothorax.

Malignant lymphoma arising from lung and pleura is very rare. It accounts for about 0.3% of all NHL. Most of the reports have been found to be associated with longstanding chronic Tubercular Pyothorax, or Pulmonary Tuberculosis treated with therapeutic Artificial Pneumothorax. Most of the reported case studies have suggested that chronic inflammation of the pleura poses a significantly increased risk of developing pleura based lymphoma.

The association of extranodal lymphomas in a background of chronic inflammation is well established—for example, the association of Helicobacter pylori and gastric lymphoma. In a similar scenario, this primary pleural lymphoma is associated with history of chronic pyothorax (2.2%), or as a result of chronic inflammation of the pleura. It is thought that long standing pleural inflammation is an important factor in the development of these lymphomas. Epstein-Barr virus and inflammatory cytokine stimulation are also thought to play a role.

Though Primary Pleural Lymphoma is extremely rare, pleural involvement secondary to systemic lymphoma is relatively common. Approximately 16% of NHL either present with, or subsequently develop pleural involvement, either unilateral or bilateral. In case of unilateral involvement it is more common on the left side. The patients are symptomatic, mostly presenting with dyspnoea, cough or chest pain. Pleural effusion commonly accompanies pleural lymphoma, which carries a poor prognosis.

The radiological abnormalities are; a pleura based mass, pleural thickening, pleural effusion, mediastinal lymph nodes and bilateral pulmonary nodules. Left pleural involvement is more common than the right. Pleural biopsy and pleural fluid cytological examination are the early steps in the essential diagnostic workup of pleural-based mass.

Conclusion

Ours is a rare case of pleural lymphoma i.e. primary pleural lymphoma with chylous pleural effusion. Neither there was any history of chronic pyothorax, nor any other form of chronic pleural inflammation, as has been described in the literature.

References