Chylothorax and Chylopericardium as Initial Presentation of Thymoma: A Case Report

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SUMMARY

The rarity and simultaneous commonness of thymoma in a 27-year-old female presenting with chylothorax and chylopericardium is highlighted in this case report.

Overall, incidence of thymoma is less than 1% of all neoplasms. However, thymic malignancies are difficult to segregate because of their great variability in cytological and structural patterns. In the Philippines, unfortunately, there is limited published literature on which to base a review of the epidemiology of this condition due to relatively small number of population-based studies.

Chylothorax and chylopericardium is defined as the presence of chylous effusion in pleura and pericardium respectively. Chylothorax is a rare clinical condition, while chylopericardium is also rarer, its incidence is less than 1%. On further work-up, an anterior mediastinal mass was discovered on chest CT and biopsy findings were consistent with thymoma as the cause of effusion. To date, the most frequent malignant cause of chylothorax is lymphoma. This includes cases of rare transformation to lymphoma from a diagnosed case of thymoma. Bilateral chylothorax and chylopericardium has been reported in a few case reports only.

CASE REPORT

A 27-year-old woman presented with progressive difficulty of breathing for a month. Salient features revealed decreased breath sounds bilateral, no lymphadenopathy nor masses, and had unremarkable past medical and family history. Chest radiograph revealed massive bilateral pleural effusion (Figure A). Chest ultrasound showed a free-flowing pleural effusion amounting 3120 and 650 cc, on the right and left, respectively. Thoracentesis was done and 2500 cc of milky, odorless pleural fluid was drained (Figure B). Biochemical analysis of the pleural fluid confirmed the diagnosis of chylothorax based on high triglyceride level of 620 mg/dl. The patient underwent bilateral pleural catheter drainage due to recurrence of effusion (Figure C). Also, a low-fat diet in the form of medium-chain triglycerides was advised. Post-drainage, CT of...

Figure 1: Initial chest xray showing bilateral pleural effusion
the chest revealed an anterior mediastinal mass measuring 15.7x7.4x15.3 cm. There are non-calcified lymph nodes in the cardiophrenic, right lower and mid internal jugular chain.

Notably, there is presence of moderate pericardial effusion and still with minimal pleural effusion (Figure D). A CT guided biopsy of the anterior mediastinal mass was done. Recommended immunohistochemistry studies (CK7, CD30) showed negative results. Hence, the final histopathological report was Thymoma. Echocardiogram showed large circumferential pericardial effusion with signs of tamponade. Patient did not present with hemodynamic instability, a pericardial drain was inserted and noted chylous effusion compatible with thymoma. She immediately underwent radiotherapy. Postradiation, there is cessation of chylous effusion. Therapy was continued as an out-patient basis.

**Discussion**

Thymoma is a rare tumor of unknown malignancy with estimated prevalence of only 0.2% to 1.5% of all malignancies. (1) Although rare, it is the most common anterior mediastinal mass. Thymic tumours are a heterogeneous group of malignancies with a range of clinical presentation, often limited to the anterior mediastinum and without an aggressive disease profile. It is asymptomatic in majority of cases. Chylothorax from thymoma were not mentioned in literature. However, in a journal by Raya et al (2017), a 28-year-old Hispanic woman diagnosed with thymoma presented with chylothorax and chylopericardium. On work-up, it showed a rare transformation of classical Hodgkin’s lymphoma from previous thymoma. Based on the journal of thoracic oncology, association between thymoma and subsequent development of non-Hodgkins lymphoma is established to be due to the disruption of T-cell function.
caused by either the thymoma itself or its treatment. Several studies have documented associated malignancies however, small population were used in the study. It is not possible to draw an absolute conclusion, but there may be a small increased risk of subsequent malignancies in those individuals who develop a thymoma. (2)(3)(4)

Chylothorax is a rare clinical condition characterized by pleural fluid with a turbid or milky appearance due to high triglyceride content in the form of chylomicrons that enter the pleural space. Malignancy is a leading cause of non-traumatic chylothorax responsible for 63% of non-traumatic causes. Lymphoma is the single most common cause responsible for 50% of the non-traumatic causes. Other common causes of chylothorax are Castleman's disease, sarcoidosis, histoplasmosis, lymphangioleiomyomatosis, Noonan syndrome, filariasis, heart failure, lymphangitis of the thoracic duct, aneurysm of the thoracic aorta that erodes the duct, Cirrhosis of the liver, childbirth and idiopathic. (5) Surgical procedures and chest trauma are the most common causes of the traumatic chylothorax. Diagnosis of chylothorax is established by direct analysis of the pleural fluid obtained from thoracentesis. The fluid is characteristically milky in appearance. The triglyceride concentration greater than 110 mg/dl, a ratio of pleural fluid to serum triglyceride of greater than 1.0, and ratio of pleural fluid to serum cholesterol of less than 1.0 usually confirms chylothorax. Chylothorax will be excluded if pleural fluid triglyceride concentration is less than 50 mg/dl. (6) This criteria was fulfilled in the case presented. The cornerstone of treatment are adequate fluid and electrolyte replacement along with appropriate nutrition. In cases of non-traumatic chylothorax, treatment of the underlying disease such as chemotherapy and irradiation can lead to improvement, but success rates are limited up to 33%. (7)

Chylous pericardial effusions are extremely rare but have been described in literature, originally noted to occur in 3% of patients with nontraumatic chylothoraces. (8) Currently, the management of chylopericardium and more commonly chylothoraces is based on the etiology and consists of a strict dietary fat restriction, surgical intervention if indicated, and treating the underlying cause. (9) The potential causes of chylous effusions are extremely broad but primarily fall into 4 categories: malignancies, trauma, idiopathic, and miscellaneous with lymphoma being the most common nontraumatic cause. (10)

CONCLUSION

This case is unique as it is one of the cases to document bilateral chylothorax with chylopericardium due to Thymoma. As mentioned, studies have documented transformation of thymoma to lymphoma and small risk to develop secondary malignancies were mentioned in small population-based studies. Long term follow-up is necessary since thymoma is a rare malignancy with variable presentation, aggressive profile should be monitored especially to young patients to prevent life threatening complications and to provide adequate therapy.

Recommendation of this case report includes to document thymoma cases whether with benign or malignant features. Since only a few cases were reported along with also a few nontraumatic etiologies which makes developing evidence-based guidelines more challenging.

References