Unusual association of diseases/symptoms

Idiopathic chylothorax in a young man

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Summary

Chylothorax refers to the presence of chyle in the pleural space owing to disruption or obstruction of the thoracic duct or one of its tributaries. We present a case of non-traumatic, idiopathic chylothorax in an 18-year-old man. Lymphoscintigraphy was used to identify the site of leak. We tried the full armamentarium of available non-surgical therapy on him, including a modified diet, chest tube insertion, total parenteral nutrition and octreotide. Yet, despite optimal medical therapy, his effusion continued to persist. He eventually required thoracic duct ligation, which resulted in resolution of the effusion.

BACKGROUND

Chylothorax is an important cause of unilateral, massive pleural effusion, along with others like malignancy and parapneumonic effusions.

Idiopathic chylothorax accounts for 5–10% of the cases of chylothorax, and often needs lymphangiography or lymphoscintigraphy to establish the diagnosis.

One should always check pleural fluid triglyceride levels on a grossly milky-appearing tap.

It is important to treat this condition, as untreated cases are almost always fatal.

There are a few treatment options available for treatment; however, a chyle leak of greater than 1 litre per day, or a persistent leak for more than 2 weeks, warrants surgical treatment.

CASE PRESENTATION

An 18-year-old Indian man, presented with a 3-day history of right-sided pleuritic chest pain and shortness of breath with even minimal exertion. He did not sustain any trauma to his chest, and did not complain of any constitutional symptoms. There was no history of fever/chills/cough with expectoration/haemoptysis/wheezing. Nor did he complain of dizziness/syncope/palpitation/swelling in his feet.

He had no significant medical or surgical history.

He lived with his parents, was a student at school and was unmarried. He denied use of tobacco/alcohol/illicit drugs.

There was no history of any significant lung conditions or premature heart disease in his family.

At presentation, he was afebrile with normal vital signs. Apart from the sharp pain, he was in no distress. On respiratory examination, he had fullness of the right side of his chest, with the trachea deviated to the left, reduced air entry on the right, and reduced tactile vocal fremitus with dullness to percussion on the right side, all suggestive of a massive right-sided pleural effusion. Cardiovascular examination revealed a normal S1 and S2, with a regular rate

and rhythm, with no audible murmurs or friction rubs. He did not have jugular venous distension or pedal oedema.

INVESTIGATIONS

Haemoglobin was 12.3 g%, with a white cell count of 12 300/mm³ (polymorphs: 78%, lymphocytes: 22%). Erythrocyte sedimentation rate (ESR) was 16 mm (by the Westergren method). A Mantoux test revealed <5 mm induration at the end of 48 h. Chest x-ray confirmed a massive right-sided pleural effusion (figure 1). It was decided to do a diagnostic thoracentesis, which revealed a grossly white, milky fluid (figure 2), with 4/mm³ polymorphs, 102/mm³ lymphocytes, protein level of 4.112 g%, lactate dehydrogenase (LDH) of 301 IU/l, and normal adenosine deaminase (ADA) levels. Gram stain and culture were negative. Serum protein level was 7.2 g/dl, while

Figure 1 Chest x-ray with complete opacification of the right hemithorax.

Figure 2 Milky white pleural fluid.

serum LDH was 352 IU/l; suggestive of an exudative effusion by Light's criteria. Diagnostically, the pleural fluid triglycerides were elevated at 302 mg%, compared to serum triglyceride level of 104 mg%. This picture was highly suggestive of a chylothorax. Pleural fluid and peripheral smear for microfilaria was negative.

CT scan of the chest and abdomen, performed to identify any abnormal masses or lymph nodes compressing the thoracic duct, revealed no other abnormality apart from the unilateral effusion. Lymphoscintigraphy, performed to identify the site of leak, revealed leak of dye into the right pleural space. Thus, a diagnosis of non-traumatic, idiopathic chylothorax was made.

DIFFERENTIAL DIAGNOSIS

Chylothorax of possible following aetiology:

- ▶ Filariasis
- ▶ Tuberculosis
- ▶ Thoracic duct cysts
- ▶ Idiopathic
- ▶ Malignancy

TREATMENT

Initially, the patient was provided with a modified diet, restricted in fat with supplementation of medium-chain fatty acids. A chest tube inserted into the right pleural space drained 1400 ml of fluid daily. Because of the unsatisfactory response, he was then kept Nil Per Os, provided with total parenteral nutrition (TPN), and started on octreotide (100 µg three times a day). Subsequently, over the next few days, the drain output reduced to 800 ml, and then to 600 ml per day. But, despite optimal medical management, the effusion continued to persist. Therefore, a right thoracotomy with ligation of the thoracic duct was performed, which resulted in resolution of the effusion.

OUTCOME AND FOLLOW-UP

There was complete resolution of the pleural effusion, and complete relief of the shortness of breath, a few days following the thoracic duct ligation. He has had no recurrences to-date (3 years after his surgery).

DISCUSSION

Causes are divided into non-traumatic and traumatic. Malignancy is the most common, with lymphomas accounting for 60% of these. Non-malignant causes are divided into idiopathic, congenital and miscellaneous. Traumatic causes include iatrogenic and non-iatrogenic (secondary to trauma).

As opposed to empyema fluid, the supernatant of the pleural fluid from a chylothorax typically fails to clear after centrifugation.¹ A pleural fluid triglyceride concentration greater than 110 mg/dl (1.24 mmol/l) strongly supports the diagnosis, whereas a level less than 50 mg/dl (0.56 mmol/l) almost certainly excludes a chylothorax.² An intermediate triglyceride level between 50 and 110 mg/dl (0.56 and 1.24 mmol/l) warrants lipoprotein electrophoresis of the fluid to detect chylomicrons.² A ratio of pleural fluid cholesterol to triglyceride of less than 1 is also diagnostic.

CT of the thorax and abdomen is typically needed to identify the presence of lymphadenopathy or masses and the site of thoracic duct rupture. If the site of leak still remains uncertain, lymphangiography or lymphoscintigraphy may be helpful.

Conservative management should be tried first because the thoracic duct leak closes spontaneously in nearly 50% of patients. Tube thoracostomy or repeated thoracentesis should be performed. Chyle production can be reduced by instituting TPN^{3 4} or a fat-restricted oral diet supplemented with medium-chain triglycerides.^{3 4}

Octreotide reduces the splanchnic blood flow, reduces the absorption of triglycerides, inhibits serotonin, motilin, vasoactive intestinal peptide (VIP) and gastrin, and decreases gall bladder contractility and bile flow, and has been used with great success.^{3 5}

Chyle leak greater than 1 litre per day⁶ for 5 days, or a persistent leak for more than 2 weeks despite conservative management, should be treated surgically. In difficult cases, pleurodesis is the only option.⁷

Learning points

- ▶ Consider chylothorax in the differential of massive unilateral pleural effusion.
- ▶ Consider lymphoscintigraphy if CT scan of the chest and abdomen fails to reveal the aetiology.
- ▶ Always start with conservative treatment.
- ▶ Octreotide is an effective agent in chylothorax.
- ▶ Timing of surgical intervention is essential.

Competing interests None.

Patient consent Obtained.

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