Case Report
Spontaneous Metachronous Chylothorax—Presenting As a Neck Lump

Ramanan Daniel, Ashvin Paramanathan, Fiona Hill, and Patrick Walsh
Department of ENT, Western Health, Gordon St, Footscray VIC 3011, Australia
Address correspondence to Ramanan Daniel, ramanan.daniel@gmail.com

Received 31 August 2015; Accepted 16 May 2016

Copyright © 2016 Ramanan Daniel et al. This is an open access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Abstract
Chylothorax is defined as an accumulation of chyle-containing lymphatic fluid within the pleural space. Most adult chylothoraces are related to surgery or malignancy. Spontaneous idiopathic chylothorax in adults is an extremely rare occurrence with very few reported cases. We present the first described case of spontaneous metachronous chylothorax presenting as a neck lump in a 48-year-old Caucasian female and its associated workup, diagnosis, and management. It is an extremely rare condition and efforts should be made at excluding more common causes of pleural effusion in adults. Definitive diagnosis should be made through thoracocentesis and fluid analysis. Its management remains variable however conservative management is usually advocated as first line therapy.

Keywords spontaneous chylothorax; recurrent; metachronous; neck lump

1. Introduction
Chylothorax is an accumulation of chyle-containing lymphatic fluid in the pleural space [1]. While common in the neonatal and fetal population, it is responsible for only 3% of pleural effusions in adults [1]. Malignancy and iatrogenicity are the leading differentials for adult chylothoraces with a prevalence of approximately 90% [2]. Spontaneous idiopathic chylothorax is extremely rare and in particular there have been no reported cases of recurrent idiopathic chylothorax presenting as a neck lump.

2. Case report
A previously well 48-year-old female nonsmoker initially presented two years ago with spontaneous left sided neck swelling associated with left sided pleuritic chest pain and mild dyspnea. There was no history or symptoms indicating trauma, infection or malignancy. Clinical examination revealed a diffuse left sided nonspecific swelling throughout the supraclavicular fossa with no evidence of any oral pathology, cervical or axillary lymphadenopathy. There were no other distinct palpable neck masses. Auscultation showed reduced breath sounds in the left lower zone with dullness to percussion.

Ultrasoundography demonstrated a 2.7 cm × 2.0 cm anechoic fluid collection lateral to the internal jugular vein. Subsequent computed tomography (CT) of the neck revealed extrinsic compression of the left jugular vein by a 2.7 cm diameter subtle low-density lesion at the root of the neck (Figure 1) associated with an extensive left sided interstitial soft tissue edema of the neck, mediastinal interstitial edema, and a left sided pleural effusion. A pleural tap was performed revealing milky fluid. Biochemically, the fluid had triglycerides of 26 mmol/L, cholesterol of 2.6 mmol/L, total protein of 32 g/L, and albumin of 30 mmol/L. The fluid was consistent with the diagnosis of a chylothorax.

Further investigations including a full blood count, biochemical and inflammatory markers were normal. Sputum microscopy; cultures and sensitivities; and Ziehl-Neelsen staining revealed no growth. She clinically improved and was discharged after four days. A repeat
Figure 2: Second presentation: posterior-anterior erect chest radiograph showing a left-sided pleural effusion.

Figure 3: Second presentation: CT scan with contrast showing soft tissue swelling within the left supraclavicular region of the neck (white arrow) with terminal dilatation of the thoracic duct.

Figure 4: Second presentation: posterior-anterior erect chest radiograph showing a complete resolution of effusion after one week with conservative management.

CT scan performed two months later revealed no evidence of any remaining neck mass or suspicious lung lesion or fluid accumulation.

She subsequently represented two years later with identical symptoms and signs. A chest X-ray (Figure 2) and CT scan (Figure 3) demonstrated a left sided pleural effusion with extensive soft tissue swelling centered around the left supraclavicular region extending upward along the left side of neck with terminal dilatation of the thoracic duct measuring 2 cm × 1.1 cm × 1.4 cm. She was conservatively managed on a low fat diet and discharged home. She was reviewed in outpatient clinics one week later at which time she was symptom free. A repeat chest X-ray at this time (Figure 4) did not show any residual pleural effusion. The patient elected for conservative management and did not wish for any further investigations unless there were further episodes.

3. Discussion

Spontaneous chylothorax is extremely rare with only sporadic case reports occurring in the adult population. In one case series of 203 patients with chylothorax in a tertiary center, 6.4% of cases were found to have unknown causes [3]. Another 30-year case series of 18 patients with chylothoraces demonstrated two cases with spontaneous idiopathic chylothoraces [4]. Given its rarity, such a condition represents a diagnostic conundrum. The above discussed patient described having a high fat diet during the month preceding her second presentation but there is no current evidence suggesting this may be a cause for spontaneous chylothorax. The symptoms can be nonspecific, as chyle is not irritating to the pleura and symptoms generally relate only to the presence of fluid in the chest cavity. The patient presented with sudden onset nonspecific neck swelling most apparent in the supraclavicular region associated with mild dyspnea.
and mild pleuritic chest pain without any associated constitutional symptoms. In chronic presentations, electrolyte disturbances, reduction of venous return, lymphocytic depletion, and weight loss can occur.

The diagnosis of a chylothorax is made through both radiological and biochemical testing. In this case, computed tomography was useful to rule out other sinister causes of pleural effusion [1]. Chylothoraces can also occur bilaterally given the normal course of the thoracic duct in the chest traverses from right to left at the vertebral level of T5. The most specific diagnostic test remains a thoracocentesis and biochemical evaluation of the fluid. Triglyceride levels greater than 1.24 mmol/L (in this case it was 26 mmol/L) is shown to be specific in 99% of cases [5]. Triglyceride levels lower than 0.56 mmol/L are associated with a less than 5% chance of chylothorax [6]. Lymphoscintigraphy and lymphangiography are useful in identifying the site of leak and any anatomical malformations in the thoracic duct with a recent study demonstrating localization of leaks in approximately 79% of patients [7]. However, its usefulness is unclear in patients with spontaneous chylothoraces where resolution of symptoms occurs quickly.

Management of spontaneous chylothorax depends on the etiology and premorbid status of the patient and is highly variable given its paucity. The principles of management include addressing any underlying etiology such as malignancy or infection and interventions that may aid with immediate symptom management. In the above case, given the suggestion of a neck lesion on initial imaging, the patient was investigated for possible infectious or mitotic causes. The repeat CT scan showed resolution of these changes, suggesting they represented a collection of chyle.

In idiopathic cases, common practice normally includes conservative management with low fat diets and observation of symptoms. Dietary control measures including a low fat diet decreases the flow of chyle through the thoracic duct and may allow spontaneous closure of a duct leak. If symptoms persist, more aggressive therapy is required which includes symptom control through thoracocentesis [5]. Definitive surgical management strategies include thoracic duct embolization, thoracic duct ligation or surgical closure of any leak. Medical management can include somatostatin and octreotide administration, however there is no definitive opinion on its efficacy in adults. A recent case series has also reported a therapeutic benefit from lymphangiography due to the sclerosing effects of the contrast agent [8]. In our reported case, the patient was clinically stable and had minimal discomfort from her symptoms; hence we elected for a conservative approach.

In conclusion, we present an unusual case and presentation of a recurring spontaneous idiopathic chylothorax. It remains an extremely infrequent event and efforts should be made at excluding more common causes of pleural effusion with definitive diagnosis made through thoracocentesis and fluid analysis. Its management remains variable however conservative management is usually advocated as first line therapy in clinically stable patients.

Conflict of interest The authors declare that they have no conflict of interest.

References