Chylothorax-Literature Review and Two Rare Cases

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Abstract
Chylothorax is the accumulation of lymphatic fluid in the pleural space. It can be recognized by the presence of chylomicrons in the pleural fluid. It results from either obstruction or laceration of the thoracic duct. The most common causes are surgical or nonsurgical trauma, neoplasms, tuberculosis and venous trombosis. The most common neoplastic cause is lymphoma, while other malignant causes are very rare. In this article we present two malignant cases which rarely present with chylothorax and review the related literature.

Key words: Chylothorax, mesothelioma, gastric carcinoma

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INTRODUCTION

Chylothorax is the accumulation of lymphatic fluid in the pleural space. It can be recognized by the presence of chylomicrons in the pleural fluid. It results from either obstruction or laceration of the thoracic duct. The most common causes are surgical (mostly cardiovascular or esophageal surgery) or nonsurgical trauma, neoplasms, tuberculosis and venous trombosis. The most common neoplastic cause is lymphoma, while other malignant causes are very rare. In this article we present two malignant cases which rarely present with chylothorax and review the related literature.

CASE 1

A 65 year old male, living in the north western Turkey presented with sweating and left shoulder pain for 15 days. He had a pleural effusion and was cured with diuretics 15 months before his admission. He had a 50 pack-years of smoking history. He reported no asbestos exposure. Physical examination revealed decreased breath sounds over the left chest. There was a homogenous opacity at the base of the left hemithorax with a concave upper border and a 8x8 cm mass on the upper left zone on the chest X ray (Figure 1). Diagnostic thoracentesis was attempted and a cloudy fluid was obtained. Pleural fluid examination showed glucose: 133 mg/dL, LDH:201U/L, protein: 6.1g/dL, albumin: 3.6g/dL, cholesterol: 84mg/dL, and triglyceride: 600 mg/dL. Diagnosis was then confirmed as chylothorax. The cytological examination was reported as chronic inflammation. On bronchoscopic examination we have observed only mucosal irregularity at the distal portion of the left main stem bronchus, and left secondary carena Biopsy cytology was reported as chronic inflammation. Closed pleural biopsy was also undiagnostic. He was then referred to open biopsy. Pleural nodularity and masses were seen and biopsied for frozen section. As malignant cells were seen, pleurectomy was performed. Immunohistochemical staining were negative for CEA (-) and TTF1 (Thyroid transcription factor), positive for Keratin, ESA (Epithelial spesific antigene) focal membranous staining. Pathological diagnosis was confirmed as mixed type malignant mesothelioma. Patient died only six months after initiation of systemic chemotherapy.

CASE 2

A 63 year old female patient referred to our center with dyspnea and swelling of the legs. She presented with bilateral pleural effusion and was treated for heart failure two months before. Fifteen days later her dyspnea recurred and she was reevaluated. By that time left heart failure was excluded and the patient was referred to our clinic. Her biochemical results were normal including CRP and thyroid hormone levels. PPD was negative. Thoracentesis had shown a cloudy, milk-like fluid. Pleural fluid examination showed glucose 143 mg/dL, protein 3,3 g/dL, albumin 0,9 g/dL, LDH 69U/L, cholesterol 85 mg/dL and triglyceride 165 mg/dL. Chylothorax was then confirmed. Pleural fluid cytology showed lymphocytes and atypical mesothelial cells. On CT scan of thorax (Figure 2) bilateral effusion with a bilateral patchy ground glass opacity and reticular lines were seen. Rheumatological examination and markers were normal including C3 and C4 complement factors. Transvaginal pelvic ultrasound, breast examination were also normal. Oral feeding of the patient was discontinued as soon as chylothorax was confirmed. Fiberoptic bronchoscopic examination was normal. The tumor markers were reported as: CA 125:149,5 (N< 35), CEA: 3,65 (N<3,4), CA 15-3: 9,13 (N<25) CA 19-9: 0,6 (N<39): CA 125 and CEA being above normal. Abdominal ultrasound and CT was normal. Video-assisted thoracoscopy (VATS) was then performed to explore the leakage. There was no thoracic duct lesion, and pleural biopsies were taken, which werereported as chronic pleuritis. We planned to have a PET-CT scan to ru-

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le out a malignancy as sreum CA-125 was high and pleural fluid had some atypical cells. But on the sixth day after VATS, she vomitted coffee-like material. Her hematocrit dropped and an emergent gastrointestinal endoscopy was performed. A huge, bleeding ulcerating area was seen. Bleeding was stopped by sclerotherapy and biopsy was reported as low differentiated adenocarcinoma with rignet cells. She was referred to an oncology clinic for further staging and therapy.

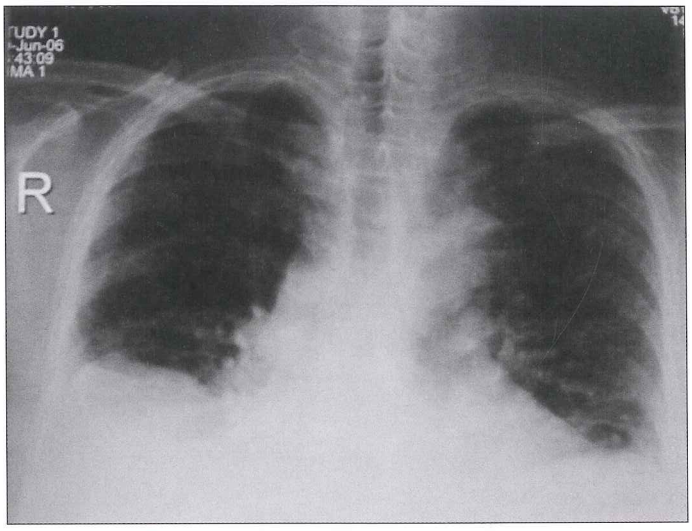


Figure 1.

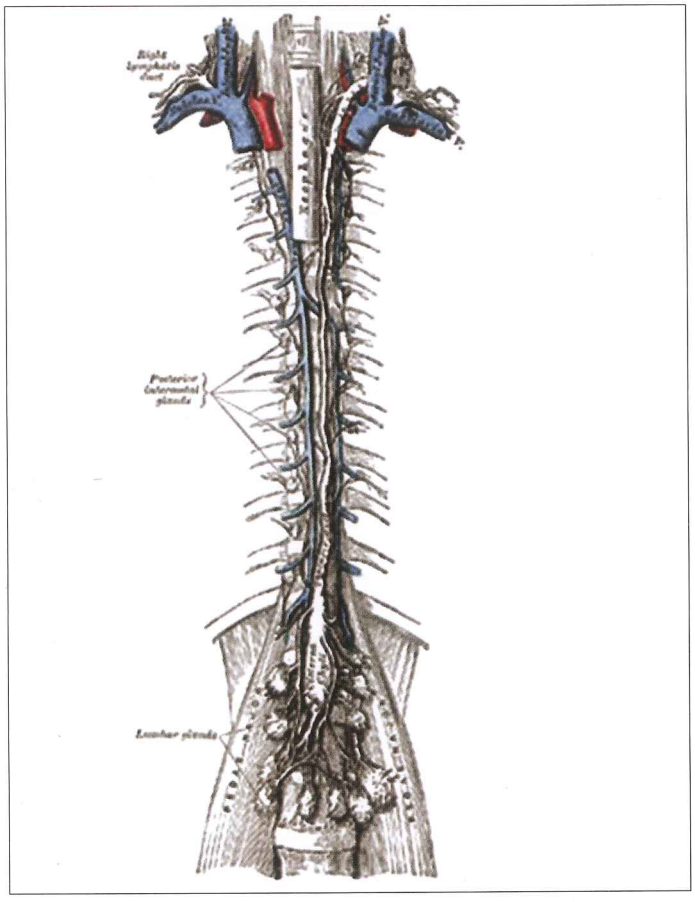
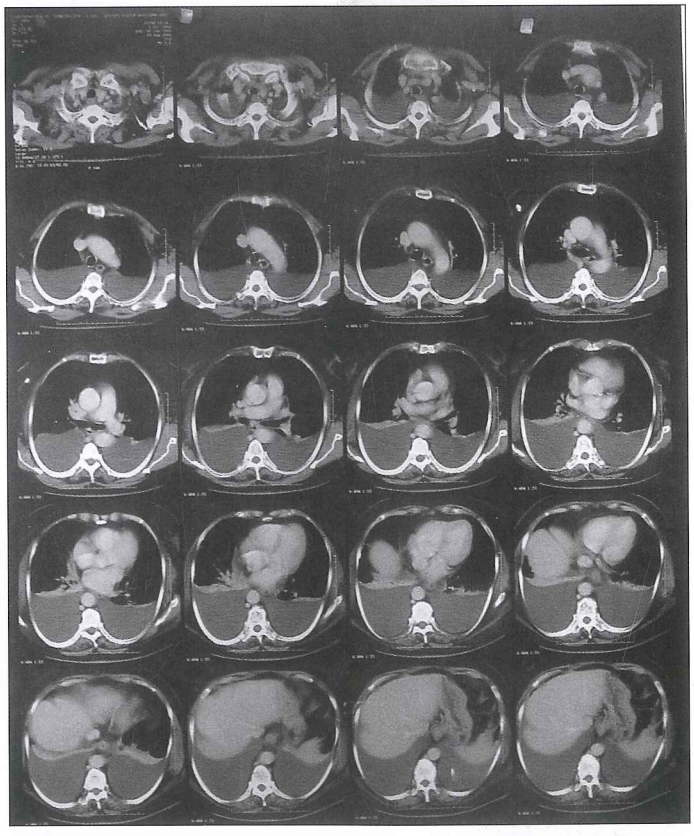


Figure 2.

DISCUSSION

Chylothorax is the accumulation of chyle in the pleural space. Chyle is a milky, opalescent fluid that contains chylomicrons, triglycerides and lymphocytes. Because of its bacteriostatic and nonirritating property, it does not cause fibrothorax. It is the lymphatic fluid occuring in the gastrointestinal system and the left main collecting vessel of the lymphatic system. The anatomic pathway of the duct is variable. The most common variation is shown in the Figure 3. It originates from the cisterna chyli in the abdomen. Cisterna chyli is a globular structure that is 3-4 cm long and 2-3 cm wide. It may be found along the vertebral column anywhere between T10 and L3 on the right side of aorta. The duct ascends and enters the thorax through the aortic hiatus at the level of T10 to T12 still in the right side of the aorta. It passes to the anterior face of the vertebral column, posterior to the eosophagus. When ductus thoracicus reaches the level of T5 to T7, it crosses the dorsal face of aorta, to the left posterior side of the mediastinum and goes along the left side of the esofagus. It drains into the venous system at the junction, where the left subclavian vein and internal jugular vein unite. If we asses its journey in the body,



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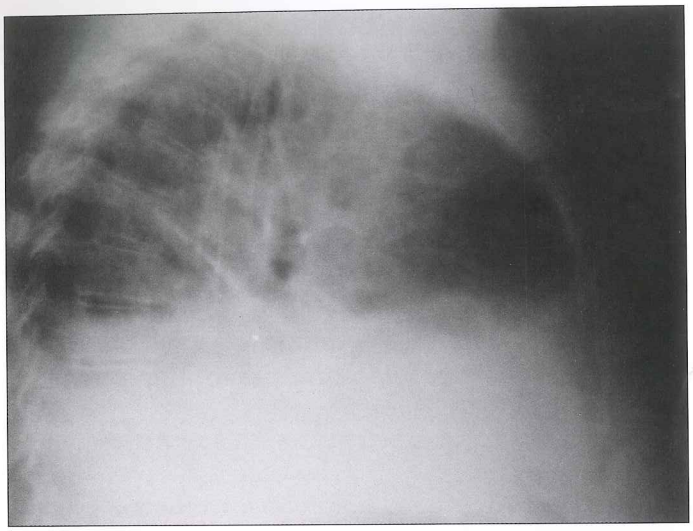

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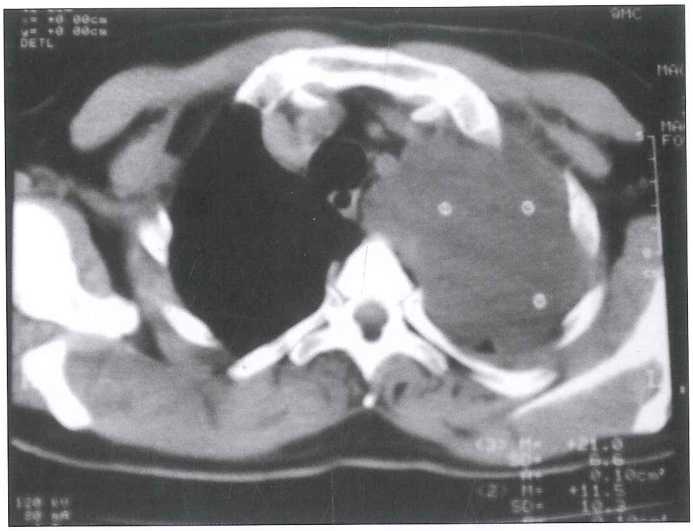

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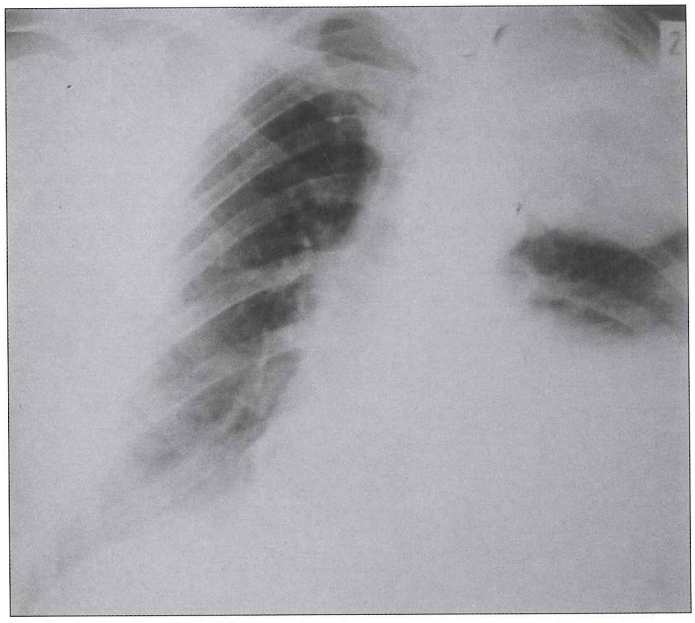

Figure 5.

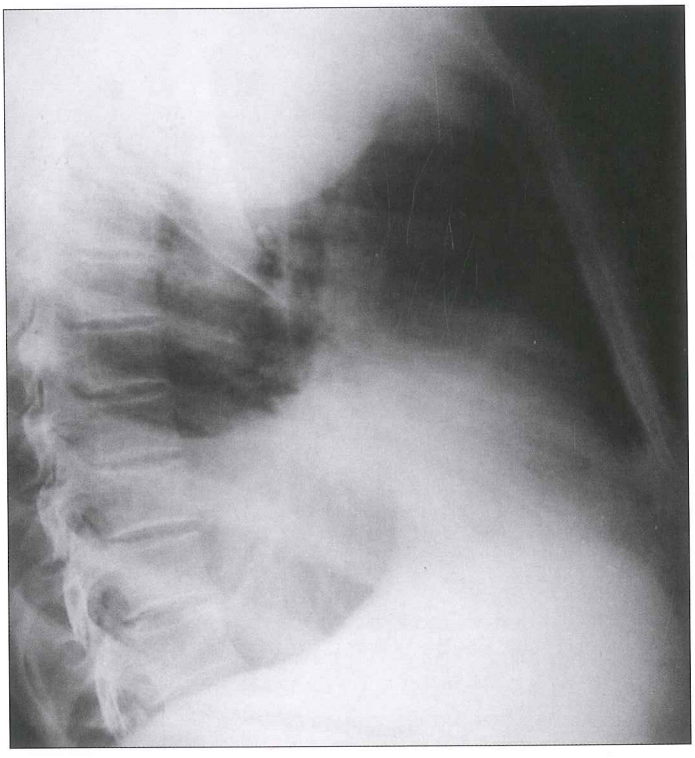

Figure 7.

we can easily understand why an injury at the level below T5, results in a right sided chylothorax while an injury above this level causes left sided. There are many collateral veins along the way through, and so the ligation of the duct doesn't impair the delivery of the lymph.

Chylothorax results from either an obstruction or a laceration of the thoracic duct. The cause in our cases is thought to be the lymphatic obstruction similar to the cases reported by Ito , Liote and Bautz et al [2,4,10]. In Ito's case, lymphoscintigraphy suggested an obstruction of the right parasternal lymphatic vessel [2]. In other cases the cause of the chylothorax is not discussed. The passage of chylous ascites from the diaphragm also causes chylothorax. We did not find a case occuring this way.

The etiology of the chylothorax is listed in Table 1. More than 50 % of chylothoraces are because of tumors lymphoma being the most common cause (75 %). Trauma is the second cause of chylothorax. It can be surgical or non surgical. Especially cardiovascular operations are very important in the etiology of chylothorax. Eosophageal procedures must also be taken into account as a common cause. In the literature review since 1962 chylothorax is seen in only six cases of mesothelioma [1-4] and three cases of gastric carcinoma [5-13] as the initial manifestation like our cases.

Table 1. Etiology of chylothorax (16)

Surgical trauma	Cardiovascular surgery	**Nontraumatic**	Lymphoma
	Eosophageal surgery		Other malignancies
	Pulmonary surgery		Retrosternal goiter
	Thoracic sympathectomy		Sarcoidosis
	Costovertebral surgery		Tuberculosis
	Neck surgery		Lymphangioleiomyomatosis
	Diafragm surgery		Lymphangiomatosis
Nonsurgical trauma	Blunt chest tauma		Lymphangiectasis
	Strong cough		Tuberosclerosis
	Vomiting		Yellow nail syndrome
	To give birth		Amyloidosis
	Hold something heavy		Filariasis
			Central venous trombosis
	Vertebra hyperextension		Heamangiomatosis (Gorham syndrome)
	Vertebra fractures		Behcet disease
	External cardiac massage		Congenital
	Aortic arteriography		Congestive heart failure
	Subclavian vene catheterization		Subclavian vene stenosis
	Gun shot	**Idiopathic**	

The chylous pleural fluid must be differentiated from pseudochylothorax and empyema. In the former one the milkiness is due to leukocytes while in the latter, due to cholesterol or lecithin-globulin complexes. Pseudochylothorax occurs in prolonged persistence of exudative pleural fluid and most commonly seen in tuberculosis or rheumatoid arthritis in majority. We suspected chylothorax in our cases after seeing the cloudiness of the fluids.

The diagnosis of chylothorax is confirmed by measuring the triglyceride levels of the effusion. Levels above 110 mg/dL (1,24 mmol/L) is considered that chylothorax is probably present. The levels below 50 mg/dL exclude the diagnosis but the levels between 50 and 110 mg/dL necessitate a protein electrophoresis. The demonstration of chylomicrons confirms the diagnosis of chylothorax.

A detailed history and physical examination is very important for clarifying the etiology. CT examination of mediastinum and abdomen is useful for the demonstration of a nontraumatic chylothorax. Lymphangiography can be a guide for demonstrating the leakage and for deciding about the type of therapy. We were unable to perform a lymphangiography in our cases.

Patients with chylothorax can be treated by conservative methods or surgery. Certain principles are common for both treatment options, including treating the underlying cause, decreasing chyle production by modifying (giving medium chain triglicerides) or cessation of oral intake and giving parenteral hiperalimentation, draining and obliterating the pleural space (by pleuroperitoneal shunt or tube drenage) and instituting necessary respiratory care. In the first case the characteristics and the appearance of the fluid changed immediately after nearly one week after discontinuation of enteral feeding. It was shown in animal models that the thoracic duct leak closed spontaneously in 10 or 14 days by a conservative management. (14,15). Somatostatin, or its analogue, octreotide, has been used with success in some pediatric cases of postoperative and iatrogenic chylothorax. If lymphoma or metastatic carcinoma is detected, radiotherapy is recommended before pleuroperitoneal shunt or tube thoracostomy drainage. The most effective therapy for chylothorax is ligation of the duct. This does not cause lymphatic stasis because of the rich collateral veins and can be performed by the help of open thoracotomy and thoracoscopy. Our surgeons did not prefer to ligate the duct. In the first case the pleurectomy was done, as the operation frozen sections were reported as mesothelioma.

CONCLUSION

In short chylothorax is a condition in which fluke diagnosis can be made, and as diagnosis is made, rare malignant causes must also be kept in mind.

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