

Chylothorax Associated with Inflammatory Carcinoma

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We report a rare case of a woman with inflammatory carcinoma, an unusual type of cutaneous metastasis, arising from signet-ring cell carcinoma of the stomach, who developed chylothorax as the skin lesion progressed over the chest. No thoracoabdominal lymphadenopathy which can cause obstruction of the thoracic duct was shown by computed tomography. Although a very rare condition, inflammatory carcinoma could be a cause of non-traumatic chylothorax. (Internal Medicine 37: 538–541, 1998)

Key words: chylous effusion, pleural effusion, gastric cancer, signet-ring cell carcinoma, cutaneous metastasis

Introduction

Inflammatory carcinoma, which was first described by Küttner as “erysipelas carcinomatosum” in 1924 (1), is a rare type of cutaneous metastasis from internal malignancy. It is clinically characterized by the development of erythematous skin lesion, which resembles an acute inflammatory process like cellulitis. We herein report a case of inflammatory carcinoma of gastric origin who developed the collection of bilateral chylous pleural effusion late in the course of the disease. Although chylothorax is sometimes caused by malignant neoplasms, mostly lymphoma, the association of inflammatory carcinoma and chylothorax has never been described in the literature.

Case Report

A 58-year-old woman had recognized right inguinal erythema with itching since August 1990, followed by swelling of the vulva one month later. She had a history of stage 0 uterine carcinoma completely resected 10 years before and consulted a gynecologist at her community hospital. No gynecological abnormality was pointed out and she was diagnosed as postsurgical lymphedema. Edema kept spreading peripherally with erythematous lesion at the border with the uninvolved area. When she first consulted the practicing physician in her community in July 1991, lymphedema was present in the right leg as a whole, the vulva and the left thigh, which had progressively worsened in a year. No superficial lymph node swelling was noted.

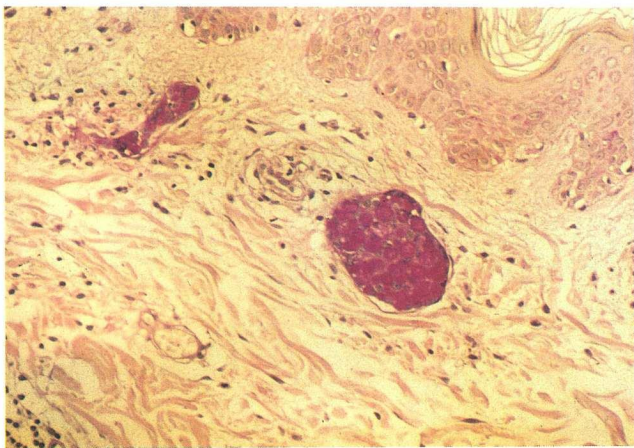
She was admitted to a hospital specializing in the care of lymphedema in August 1992 because lymphedema had spread to the lower back and the abdominal wall. Tentatively diagnosed as lymphedema complicated with cellulitis, antibiotics were administered without any clinical improvement. One month later, a skin biopsy at the left thigh was performed, which demonstrated signet-ring cell carcinoma filling dermal and subdermal lymphatics (Fig. 1). Although upper gastrointestinal fiberoptic, barium enema and thoracoabdominal computed tomography were performed, no primary malignancy was found. After discharge from the hospital, she regularly consulted her family physician. Upper gastrointestinal fiberoptic was performed again because she complained of epigastric discomfort in November 1993, which disclosed a small irregular erosive lesion at the anterior wall of the lower portion of the gastric body. The histological diagnosis of the biopsy specimen was signet-ring cell carcinoma. The lymphedema with erythematous frontier continued to spread gradually. A chest X-ray in June 1994 was normal.

From early August 1994, she complained of a productive cough, wheezing and exertional dyspnea, which were resistant to intravenous hydrocortisone and aminophylline and worsened to make her orthopneic. A chest X-ray was taken again and revealed bilateral massive pleural effusion (Fig. 2). She was admitted to our hospital on August 22, 1994. On physical examination, marked non-pitting edema and rough, pale skin in bilateral lower extremities were noted. Abdominal and thoracic skin (Fig. 3) was also edematous with some erythematous change. Neck was erythematous as a whole but not edematous. Superficial lymph nodes were not palpable except for a pea-like

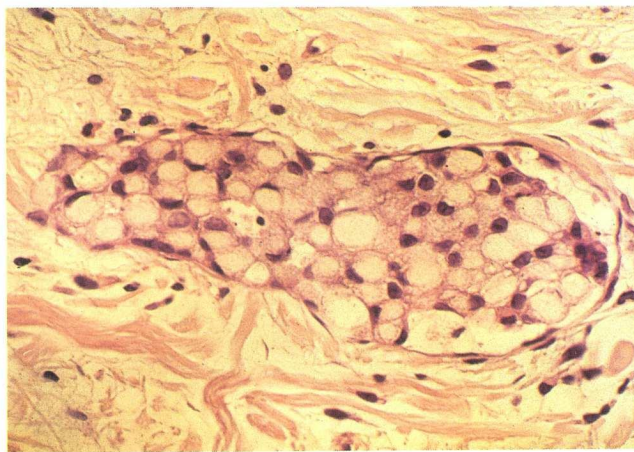
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A



B

Figure 1. A) PAS-positive tumor cell aggregates filling lymphatics in the thickened dermis (PAS stain; original magnification $\times 100$). B) Tumor cells with clear cytoplasm and eccentric nuclei, *i.e.* signet-ring cell carcinoma (HE stain, $\times 400$).

soft right axillar lymph node. Expiratory wheezes were heard over the whole chest and breath sounds were decreased in the lower chest bilaterally. Thoracoabdominal computed tomography demonstrated bilateral massive pleural effusion without ascites and no enlargement of mediastinal or abdominal lymph nodes (Fig. 4). No mass or abnormal opacity in the lung or the pleura was visualized. Thickening of the antral wall of the stomach was suspected. A right-sided thoracentesis yielded a milky yellow fluid which was cleared with ethylether. The effusion contained 300 cells per cubic millimeter, of which foamy macrophages and lymphocytes were predominant, with protein of 3.4 g/dl, glucose 108 mg/dl, lactate dehydrogenase (LDH) 133 IU, amylase 3 IU, triglyceride 673 mg/dl, total cholesterol 132 mg/dl, and chylomicron 793 mg/dl, which gave it the definite diagnosis of chylous effusion (Table 1).

Her symptoms were relieved after the removal of approximately 1,000 ml of right pleural effusion. Her serum protein was

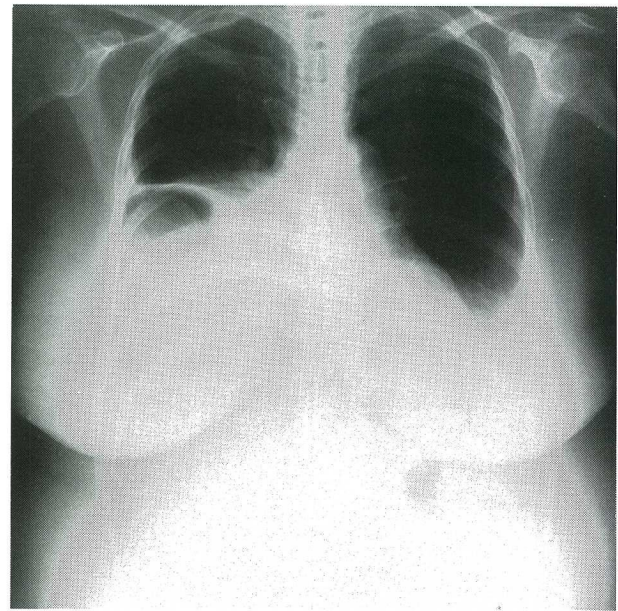


Figure 2. Chest X-ray on admission showing bilateral massive pleural effusion.

5.0 g/dl, albumin 2.6 g/dl, LDH 315 IU, triglyceride 79 mg/dl, total cholesterol 142 mg/dl, chylomicron 20 mg/dl; carcinoembryonic antigen (CEA) was below the detectable limit and the CA19-9 was elevated at 606 U/ml (normal range: <37 U/ml). Because the effusion rapidly reaccumulated, oral intake was discontinued and total parenteral nutrition was initiated at the fifteenth hospital day. A few days later the triglyceride level in the effusion decreased to 22 mg/dl, but the rate of fluid accumulation showed no substantial change. Continuous drainage and subsequent chemical pleurodesis was attempted but resulted in failure. Repetitive thoracentesis from both sides of the chest was necessary for relief of dyspnea. Cytological examination for malignant cells was negative at first and became positive at the third thoracentesis with small numbers of atypical cells of signet-ring cell type, which increased as the procedure was repeated. The erythema proceeded upward and peripherally to involve the face, the ears and the upper extremities. Gastrofiberscopy in September revealed reddened, edematous and erosive mucosa with spotty hemorrhage from the cardia to the angle. The biopsy specimen again demonstrated signet-ring cell carcinoma of the stomach.

Subsequent computed tomography in November showed enlarged paratracheal and subcarinal lymph nodes and a moderate amount of ascites in addition to massive bilateral pleural effusion. Her general condition gradually worsened and she died on December 8. Autopsy was not permitted, but necropsy of the right subclavian skin was performed with her family's permission. The histology was the infiltration of dermal lymphatics with aggregates of tumor cells of signet-ring cell type.



A



B

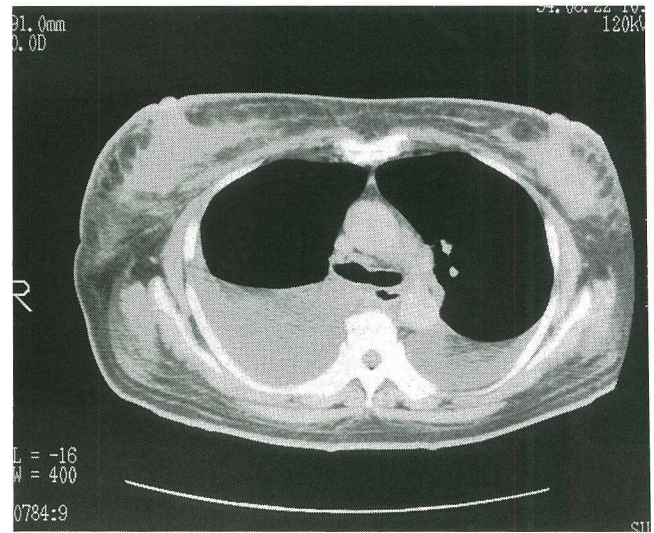
Figure 3. A) Edematous skin of the upper thorax. B) The skin had erythematous change which was most prominent in the neck.

Table 1. Laboratory Findings of the Pleural Effusion at the First Thoracentesis

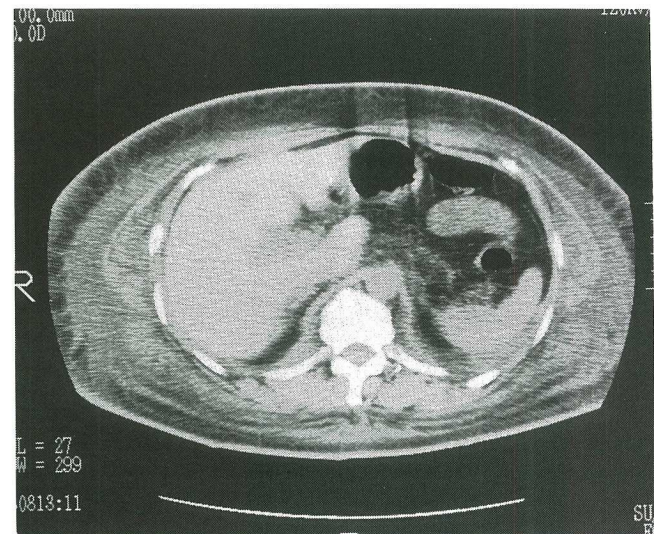
	Pleural effusion	Serum
Total protein	3.4 g/dl	5.0 g/dl
LDH	133 IU	315 IU
Triglyceride	673 mg/dl	79 mg/dl
Total cholesterol	132 mg/dl	142 mg/dl
Chylomicron	793 mg/dl	20 mg/dl

Discussion

The causes of chylothorax are divided into four major categories by Light (2): tumor, trauma, idiopathic, and miscellaneous. The most common cause of neoplastic chylothorax is lymphoma, but metastatic cancer from virtually every organ in the body is included as a cause. A chylous effusion caused by



A



B

Figure 4. Computed tomography of the thorax (a) and abdomen (b) showing bilateral massive pleural effusion without intrapulmonary opacification, ascites, or lymphadenopathy. The extracavitary soft tissue was extraordinarily thickened due to lymphedema.

malignancy usually develops as the result of extrinsic compression or direct invasion of the thoracic duct by a tumor mass. However, simply blockage of the thoracic duct by neoplasm is not enough to cause chylothorax (2), due to the presence of collaterals or lymphovenous channels (3).

In the present case, a bulky mediastinal or abdominal mass capable of obstructing the thoracic duct was absent on computed tomography when the effusion accumulated. The chylothorax did not develop until the erythematous lesion, representing the frontier of cutaneous infiltration of the tumor cells, proceeded over the chest. Although there was no direct evi-

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dence, extensive obstruction of the dermal and subdermal lymphatics of the chest wall due to intraluminal tumor extension may have played a role in the development of chylothorax. It was speculated that the collateral lymph flow from the tumor-filled thoracic duct into the posterior intercostal lymphatics could not escape through the cutaneous lymphatic network and subsequently the intraluminal pressure of the posterior intercostal lymphatics increased. Finally, the chyle refluxed through the intercostal lymphatics into the parietal pleural lymphatics and leakage of the chylous lymph occurred into the negative-pressured pleural space. The inflammatory cutaneous metastases may have promoted the development of the chylothorax rather than being the direct cause. Accumulation of the bilateral chylous effusion suggested the extensive obstruction of the thoracic duct from the lower to the upper thorax, because obstruction of the right-sided duct in the lower thorax and the left-sided duct in the upper thorax usually causes right-sided and left-sided chylothorax, respectively.

The association of pleural effusion and inflammatory cutaneous carcinoma has been mentioned in only two reports as far as we could search. In one case (4) metastasized from lung cancer, pleural effusion preceded the emergence of inflammatory carcinoma and its nature was not chylous but with adenocarcinoma cells. In another case (5) with the histology of signet-ring cell carcinoma, the condition was quite similar to the present case. The cutaneous manifestation occurred at the groin at first and then progressed to the leg. As the cutaneous lesion worsened, bilateral pleural effusion appeared, and finally the patient died with increased pleural effusion. However, in that report the nature of the pleural effusion was not mentioned. Consequently, the present report is the first one which clearly referred to the association of the inflammatory cutaneous carcinoma and the chylothorax.

The reason for the absence of the chylous ascites was not clear. Neoplastic chylous ascites usually occurs as a result of the obliteration of the chyle flow near the cisterna chyli under the diaphragm due to the abdominal mass (6) which was absent in our case. In addition, although the abdominal collateral lymphatics must have elevated the intraluminal pressure in the present case due to the extensive tumorous infiltration of the cisterna chyli, the positive pressure of the abdominal cavity may have not allowed seepage of the chyle from the peritoneal lymphatics.

This case indicated that inflammatory cutaneous metastasis of internal malignancy, although it is very rarely encountered in the clinical setting, should be included in the causes of nontraumatic chylothorax.

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