



A RARE CASE OF GASTRIC ADENOCARCINOMA PRESENTING AS CHYLOTHORAX, CHYLOUS ASCITES AND LYMPHEDEMA

General Medicine

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ABSTRACT

Chylothorax is the accumulation of chylous fluid in the thorax and chylous ascites is the accumulation of chylous fluid in the abdomen. The association of chylothorax and chyloascites with lymphedema to gastric adenocarcinoma is a very rare manifestation. The present case is a 75 year old male who presented with shortness of breath, abdominal distension and loss of appetite. Thoracentesis and abdominocentesis revealed triglyceride rich milky fluid which was suggestive of chylous effusions. CECT chest and abdomen revealed multiple necrotic lymph nodes in chest, abdomen and pelvis with skeletal metastases. Pleural fluid and ascitic fluid cytology revealed signet ring cells. Upper GI endoscopy revealed polypoidal mass in the antrum which on biopsy showed signet ring cell adenocarcinoma stomach. There was no exact mechanism given in the literature towards the cause of chylothorax and chyloascites in adenocarcinoma stomach. The most likely etiology of chylous effusions in the present case is thoracic duct infiltration of malignant cells.

KEYWORDS

Chylothorax, chylous ascites, signet ring cell adenocarcinoma stomach.

INTRODUCTION:

Chylothorax develops when disruption of the thoracic duct results in passage of chyle into the pleural cavity which is characteristically milky in appearance. Chylothorax develops most commonly due to trauma, possibly iatrogenic. Other causes include malignancy, sarcoidosis, goiter, AIDS, and tuberculosis.^{2,3,4} Chylous ascites refers to the accumulation of lipid-rich lymph in the peritoneal cavity due to disruption of the lymphatic system secondary to traumatic injury or obstruction. Worldwide, abdominal malignancy, cirrhosis, and tuberculosis are the commonest causes of chylous ascites in adults, the latter being most prevalent in developing countries.⁵ Gastric carcinoma with chylothorax and lymphedema as the initial manifestations has rarely been reported. To date, only 18 case reports of gastric carcinoma associated with chylothorax and lymphedema are available. We present a rare case of gastric adenocarcinoma presenting as chylothorax, lymphoedema and chylous ascites.⁶

CASE REPORT:

A 75 year old male with hypertension and chronic obstructive pulmonary disease presented with gradually progressive dyspnea for 2 months. Patient later developed insidiously progressing left upper extremity swelling for 1.5 months upto elbow. He then noticed abdominal distension 1 month later which was slowly progressive and associated with loss of appetite. Physical examination revealed left upper limb edema upto elbow with no palpable lymph nodes. His vitals were stable. Decreased breath sounds were noticed in right mammary area, right interscapular area, and right infraaxillary area on auscultation. Abdominal examination showed shifting dullness with no palpable masses or organomegaly. Cardiovascular system examination was normal.

The laboratory workup revealed the following: Complete blood picture, complete urine examination, renal function tests and liver function tests were normal. Chest radiograph (Figure 1) revealed moderate pleural effusion on the right side. Thoracentesis and ascitic tap revealed milky fluid, which on analysis showed hypertriglyceridemia (pleural – 1012 mg/dl; ascites-1190mg/dl) and normal cholesterol levels indicating chylothorax and chylous ascites. Ultrasonography abdomen and chest showed moderate amount of free fluid in both pleural and peritoneal cavities.

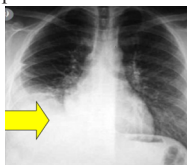


Figure 1: Chest X-ray PA view showing moderate right pleural effusion.

CECT chest showed enlarged subcentimetric nodes in pretracheal, bilateral hilar regions which were suggestive of metastases with moderate right pleural effusion (Figure 2). CECT abdomen revealed multiple lytic and sclerotic lesions in L1, L3 vertebrae and left ischial tuberosity. Few enlarged necrotic lymph nodes were noted in periportal, left gastric regions with largest measuring 14 × 8 mm and an enlarged necrotic lymph node at head of the pancreas measuring 1.8 × 1.3 cm. Few enlarged lymph nodes were noted in the right external iliac regions, largest measuring 12 × 9 mm, in bilateral inguinal regions, which were suggestive of metastases along with ascites (Figure 3) and bulky adrenals with heterogenous enhancement without any evidence of primary.

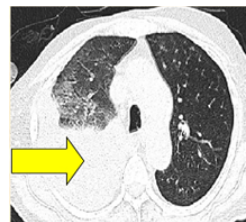


Figure 2: CT Chest showing chylothorax



Figure 3: CT abdomen showing chylous ascites surrounding liver

Further workup was done in search for the primary in view of lymph nodal, bony and adrenal metastases. Cytology of the ascitic and pleural fluid revealed signet ring cells suggestive of gastric adenocarcinoma which was the breakthrough in this case. Upper GI endoscopy showed large sessile polypoidal lesion in the antral region of the stomach (Figure 4), with erythematous mucosa noted near pylorus and biopsy of the lesion turned out to be signet ring cell adenocarcinoma (Figure 5). Patient's ECOG performance score was 3 at the time of referral to medical oncology department. Patient underwent repeated thoracentesis and abdominocentesis for relief of shortness of breath and abdominal distension. On follow up, we came to know that patient couldn't tolerate any further treatment and succumbed to disease four months after discharge.

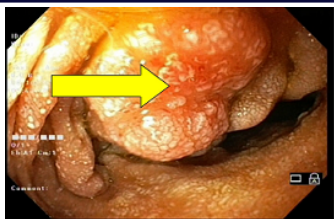


Figure 4: Upper GI endoscopy showing polypoid lesion in the antral region of the stomach

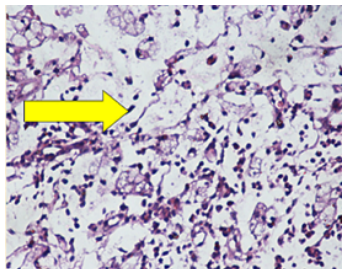


Figure 5: Signet ring cell pattern of adenocarcinoma in which the cells are filled with mucin vacuoles that push the nucleus to one side, as shown in the arrow.

DISCUSSION:

Chylothorax is defined by the accumulation of chylous, lymphatic fluid mainly from the gastrointestinal tract in the pleural space. Chylothorax occurs with disruption of normal lymphatic flow through the thoracic duct and is confirmed when the pleural fluid triglyceride levels exceed 110 mg/dL or in the presence of chylomicrons. Most non-traumatic etiologies of chylothorax are caused by lymphoma and very rarely by metastasis of other malignancies. Chylous ascites is also related to obstruction or disruption of the lymphatic ducts with a triglyceride concentration exceeding 110 mg/dL. Malignancies, mainly lymphomas, have been the common cause of chyloperitoneum and gastric carcinoma is an uncommon cause of this entity. In this situation, the chylothorax is likely the result of an overflow of the chyloascites into the pleural cavity. Chylothorax is a late manifestation of occult gastric adenocarcinoma, as evidenced by the fulminant course and death within months in the reported patients.^{7,8,9}

Information is limited on the mechanism of gastric carcinoma-induced chylothorax in the literature. The thoracic duct is primarily located at the right side of the pleura; this may help explain the development of predominantly right sided effusion. Previous case reports had various explanations for chylothorax: (i) compression of left jugular vein caused by enlarged supraclavicular nodes thereby causing increase in thoracic duct pressure. However, a large number of collateral and lymphovenous networks exist in the backflow of the thoracic duct. Shibata *et al*¹¹ stated that merely the compression of the thoracic duct was insufficient to cause chylothorax. (ii) invasion of the metastatic mediastinal lymph nodes into the thoracic duct as the cause of chylothorax. (iii) invasion of metastatic tumor cells into the thoracic duct as the direct cause of chylothorax. In our study there was mediastinal lymphadenopathy but there were no enlarged supraclavicular lymph nodes. The most probable cause of chylothorax in our case may be the invasion of tumour cells into the thoracic duct.^{9,11,12,13}

Lymphedema was proposed to be caused by the micrometastasis of gastric carcinoma cells obstructing or infiltrating into the lymphatic networks located close to the skin. The close association between lymphedema/chylothorax and gastric carcinoma in the cases described in literature was attributed to the infiltration of gastric carcinoma cells into the lymphatics.¹⁴ Primary lymphatic fibrosis, originated by malignant obstruction of the lymphatic or chyle cistern, which generates a subserosal dilation with extravasation into the peritoneal cavity is the most likely reason for chylous ascites in the present study.¹⁵

Signet cell adenocarcinoma is a diffuse, mucinous type of adenocarcinoma with more than 50% mucus producing cells. Tumour cells produce large amounts of mucus and are secreted into interstitium. It is termed "signet cell" when the mucus remains inside the tumor cell, pushing the nucleus to the periphery. Signet cell

adenocarcinoma occurs more often in males and in young patients (but our case was seen in elderly). It is known to infiltrate lymph vessels very early in the course of the disease, associated with a worse prognosis. The major risk factors are *Helicobacter pylori* infection, smoking and some dietary factors such as smoked foods. Our patient was a smoker and tested negative for *Helicobacter pylori* infection on Rapid Urease test. Systemic chemotherapy is the treatment modality of choice in patients with metastatic disease as seen in the present case. Palliative surgery, radiation and/or endoscopic procedures are the other treatment modalities available.¹⁴

CONCLUSION:

Chylothorax, chylous ascites and lymphedema combination is a rare presentation of gastric adenocarcinoma. Among the reported cases of 18 in which patients with adenocarcinoma presented with chylothorax, chyloascites was seen in very few cases. Differential diagnosis of non traumatic chylothorax should consider the possibility of occult gastric adenocarcinoma and cytology of pleural or ascitic fluid can give a lead towards the primary lesion as seen in this case.

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