Chylothorax and Chylous Ascites(Rare Presentation of a Rare Disease)



Medical Science KEYWORDS:

Dr.Indresh Dixit	BJ Medical College,CivilHospital,Ahmedabad
Dr.Naim M Kadri	BJ Medical College,CivilHospital,Ahmedabad
Dr.Karan M Desai	BJ Medical College,CivilHospital,Ahmedabad
Dr.MiteshChandarana	BJ Medical College,CivilHospital,Ahmedabad
Dr. Ravi Patel	BJ Medical College,CivilHospital,Ahmedabad

Chylothorax and chylous ascites are uncommon and usually associated with trauma , neoplasms or infections. To the best of our knowledge, Brachiocephalic/IJV thrombus leading to thoracic duct obstruction

leading to chylothorax and chylous ascites is very rare. We are presenting the case of a 30 year old female (2 months post LSCS) with complaints of abdominal distension, pedaledema and dyspnea.

Introduction

ABSTRACT

Chylothorax refers to the presence of lymphatic fluid in the pleural space secondary to leakage from the thoracic duct or one of its main tributaries.

A tear or leak in the thoracic duct causes chylous fluid to collect in the pleural cavity, which can cause acute or chronic alterations in the pulmonary mechanics. In a normal adult, the thoracic duct transports up to 4 L of chyle per day, allowing a rapid and large accumulation of fluid in the chest.

Chylous ascites is the extravasation of milky chyle into the peritoneal cavity. This can occur de novo as a result of trauma or obstruction of the lymphatic system. Moreover, an existing clear ascitic fluid can turn chylous as a secondary event.

True chylous ascites is defined as the presence of ascitic fluid with high fat (triglyceride) content, usually higher than 110 mg/ dL.(>200 mg/dL as per some guidelines)

Milky ascites is subdivided into 3 groups as follows:

- True chylous ascites Fluid with high triglyceride content
- Chyliform ascites Fluid with a lecithin-globulin complex due to fatty degeneration of cells
- Pseudochylous ascites Fluid that is milky in appearance due to the presence of pus

Case Report

A 30 year old 2 months post-LSCS female from presented to us with complains of progressive abdominal distension since last 45 days and pedal edema and dyspnea since the last 30 days.

Her past medical history includes 2 abortions before full term and is otherwise insignificant.

She gives history of being admitted in a private setup before 10 days with similar complaints where she underwent therapeutic pleural fluid tapping of 1 liter milky pleural fluid.

On examination, her BP was 114/82 mm Hg bilaterally symmetrical, pulse 88/min with normal force,volume,tension and bilateralsymmetry and a respiratory rate of 18/min.

Her general examination was insignificant except mild pitting edema in lower limbs. There was no evidence of icterus, enlarged neck nodes or engorged neck veins.

Systemic examination findings included decreased chest movement bilaterally and a decreased air entry bilaterally in lower zones. Also, she had atense abdomen with dull note on percussion.

Her lab values viz. CBC,LFT,RFT,SERUM ELECTROLYTES and ESR were within normal limits.

Serum Protein was 6.72 g% with reduced Albumin levels : 2.9g% and Globulin level : 3.82

She was negative for ANA,RA, HIV, HBsAG,APLA.

Her lipid profile was suggestive of a reduced Cholesterol and Triglyceride level.

Her Chest X-ray was suggestive of bilateral moderate pleural effusion.

Her 2D ECHO findings were normal.



She underwentan abdominal Sonogram which revealed a moderate to gross ascites with enlarged left ovary $(2 \times 4 \text{ cm})$.

So we ordered a CECT Thorax+Abdomen/Pelvis which was suggestive of a Right sided IJV and Brachiocephalic trunk thrombus and also of bilateral pleural effusion, gross ascites and a bulky left ovary.

A venous Doppler findings were additive for presence of IJV and Brachiocephalic Trunk thrombus.

At this stage we referred her to CTVS department. They advised oral anticoagulants plus LMWH for her thrombus as invasive thrombolyis or filter placement was not required in their opinion.

We treated the Patient conservatively with Aspirin,Clopidogre l,Dicoumarone,LMWH,antibiotic prophylaxis plus Diuretics for the edema.Also she was advised a low fat and high protein diet.

She required ascitic tapping on two separate occasions to relieve the respiratory embarrassment and abdominal distension. Albumin was replaced on both occasions.

Ascitic Routine-Micro revealed raised Proteins(1.4 gm%) and Raised Cells(270 PMN/cu cm) with Triglyceride Levels 210 mg% hence confirming chylous ascites.

Ascitic fluid culture came back negative.



Ascitic fluid ADA was normal and Ascitic Cytology did not reveal the presence of any abnormal cells.

Her serum CA-125 Levels were 910 IU/ML(normal <35) which is highly raised.LDH levels were slightly raised. β -HCG and α -FP levels were normal.

We transferred the patient to Oncology Department for further workup where aCytoblock of her Ascitic fluid came back negative and her Ovarian size was deemed too small to get a confirmatory biopsy.So after undergoing conservative treatment and bilateral therapeutic pleural tapping she was discharged and asked to come back for follow up after 2 months to check for increase in ovarian size.

Meanwhile we referred the patient back to CTVS department for further workup of the IJV thrombus where she was advised to continue with a conservative line of management for the thrombus.

Discussion and Conclusion

Chyle is described as being white, milky, or opalescent in appearance. This characteristic color is seen in less than one half of patients with chylous effusion^{1,2}. Pleural-ascitic fluid triglyceride level of more than 110 mg/dl(>200 mg/dl as per some guidelines) generally confirm the diagnosis of chylothorax or chylous ascites^{1,2}. Chylous pleural effusion (Chylothorax) is usually secondary to disruption of the thoracic duct or derangement of lymphatic flow within the thorax ^{1,3,5}. Chylous ascites is associated most frequently with malignant conditions such as lymphomas and disseminated carcinomas from primaries in the pancreas, breast, colon, prostate, ovary, testes, and kidney⁸. In some patients, chylothorax occurs in the setting of chylous ascites, which in turn is related to a primary abdominal process such as nephrotic syndrome, hypothyroidism, cirrhosis of the liver, abdominal operations,

and pancreatitis1.

Table 1. Causes of Chylous Ascites

Neoplastic (common in adult population) Lymphoma Other cancers (see text) Lymphangiomyomatosis Carcinoid tumors Kaposi's sarcoma Cirrhosis (common in adult population) Infectious Tuberculosis Filariasis (Wuchereria bancrofti) Mycobacterium avium intracellulare Congenital (more common in pediatric population) Primary lymphatic hypoplasia Yellow Nail Syndrome Klippel-Trenaunay Syndrome Primary lymphatic hyperplasia Bilateral hyperplasia Intestinal lymphangiectasia Inflammatory Radiation Pancreatitis Constrictive pericarditis Retroperitoneal fibrosis Sarcoidosis Celiac sprue Whipple's disease Retractile mesenteritis Postoperative Abdominal aneurysm repair Retroperitoneal node dissection Catheter placement for peritoneal dialysis Inferior vena cava resection Traumatic Blunt abdominal trauma Battered Child syndrome Other causes Right heart failure Dilated cardiomyopathy Nephrotic syndrome

Chylothorax and Chylous Ascites are extremely rare clinical entities on their own.

So,can the presence of and IJV/Brachiocephalic vein thrombus be dismissed as a coincidence?

Pregnancy is a hypercogulable state on its own so that could explain the thrombus part. $^{9}\,$

Chylothorax could be seen in upto 5% cases of IJV thrombosis¹⁰

Also, there are causes other than gynaecological malignancy of a raised CA-125 with chylous a scites. $^{\rm 11}$

So, it is currently not possible to point out the exact cause of the chylous effusion and ascites.

All in all,this case doesn't fit in the classic 3 T'S of chylous effusion/ascites as in trauma,TB or Tumour or atleast one of them can't be stamped just yet.

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