Case Report

Retroperitoneal mass lesion leading to massive chylothorax and chylous ascites. An extrathoracic pathology

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Abstract

Most common causes for chylous ascites include trauma, malignancy and infections. Amongst pediatric age group congenital lymphatic anomalies such as lymphangectasia are most common described cause for chylous ascites. We are presenting a case of retroperitoneal mass lesion leading to massive chylothorax and chylous ascites.

Keywords: Retroperitoneal mass, Chylothorax, Chylous ascites, Extrathoracic pathology

(*Thoracic Endoscopy 2016; 1: 139-143)*

Introduction

Chylous ascites is accumulation of peritoneal fluid that is rich in triglycerides, generally a triglyceride level more than 200 mg/dl give rise to chylous ascites .Estimated incidence in United States is approximately 1 per 20, 000.[1] Most common causes for chylous ascites include trauma, malignancy and infections. Amongst pediatric age group congenital lymphatic anomalies such as lymphangectasia are most common described cause for chylous ascites.

Case Report

A 61 years old man presented in emergency with progressive shortness of breath for 7 days associated with diffuse pain abdomen and gradually progressive abdominal distention. It was not associated with any pedal edema, chocking sensation or air hunger during night. There was no history of smoking, addiction or high risk behavior. On examination patient had pulse rate 88 beats per minute, regular normovolemic and blood pressure of 118/80 mm hg. There was no clubbing, cyanosis, pedal edema or elevated jugular venous pressure. On systemic examination air entry was reduced on right side with decreased tactile vocal flemitus and vocal resonance and on abdominal examination he had ascites with no palpable organomegaly.

Chest x ray revealed a large pleural effusion on right side, on aspiration pleural and ascitic fluid were milky white in color (Figure 1) and had triglyceride content of 537 mg/dl and 1242 mg/dl respectively with exudative features on fluid analysis, fluid was negative for malignant cytology.

A high resolution CT was ordered in view of high suspicion of malignancy in the background of rapidly developing polyserositis. CT Abdomen (High Resolution Computer Tomography) revealed retroperitoneal mass and FDG (18 Flouro Deoxy Glucose) avid mass lesion on PET (Positron Emission Tomography) scan (Figure 2).

Medical thoracoscopy was done for right sided massive effusion and to obtain pleural biopsy suspecting Lymphoma (Diagnostic pleural fluid cytology was

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showing atypical lymphoid cells). Around 3000 ml of pleural fluid was aspirate slowly and quick inspection of pleural cavity was done with flexirigid olympus pleuroscope. Parietal pleura were looking thickened and inflamed and neovascularization noted on parietal and visceral pleura. Lots of fibrin strands floating in pleural space and on collapse lung. Multiple biopsies taken from parietal pleura and sent for histopathology. Pleurodesis was done with slurry talc via Talc poudrage method.

Figure 1. Milky white fluid

Pleural biopsy showed fragments of benign fibro collagenous and fibro adipose soft tissue infiltrated by reactive lymphoid cells. Patient was diagnosed to have Follicular Lymphoma – Stage II and immunochemistry was positive for CD 23,CD 20 ,CD 10, CD3 and Bcl6,Ki 67 index was <10% and a diagnosis of low grade lymphoma possibly follicular or small cell lymphoma carried forward.

Patient was started on R – Bendamustine (R – CHOP), Rituximab and octreotide. Gradual resolution of tumor size, pleural fluid and ascites was noted on therapy. 3 month after starting Chemotherapy Patient presented with massive hydrothorax progressive disease leading to obstructive jaundice and cholangitis. On reimaging there was progressive disease leading to malignant biliary infiltration and obstruction was not amenable for therapeutic endoscopic or percutaneous drainage. Patient was resuscitated and managed in intensive care but we unfortunately lost the patient due to progression into multiple organ dysfunction state.

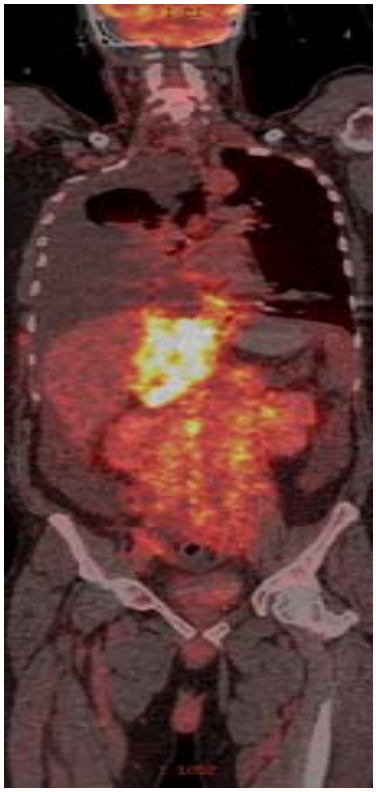

Figure 2. PET scan showing retroperitoneal mass

Discussion

A Chylothorax occurs when lymph fluid from the thoracic duct or its tributaries accumulates in the pleural space. Chylous ascitic fluid can also flow into the pleural space. The etiologies of chylothorax can be categorized as non-traumatic or traumatic. Malignancy is the leading cause of nontraumatic chylothorax. Chyle (lymphatic fluid of

intestinal origin) has a high content of triglycerides in the form of chylomicrons, which produce the milky, opalescent appearance of lymphatic fluid. This milky appearance may clear during a fast and rapidly return after ingestion of dietary fat.

Chylothorax generally results in an exudative pleural effusion; however, transudative chylothorax has been reported in small numbers of patients with amyloidosis, cirrhosis, nephrotic syndrome, superior vena cava obstruction, heart failure, and chylous ascites that has crossed the diaphragm into the pleural space.

Computed tomography (CT) of the thorax and abdomen is typically performed in most patients with a chylothorax, looking for mediastinal and retroperitoneal lymphadenopathy or masses, and also lung parenchymal diseases that may provide a clue to the etiology (eg. lymphangioleiomyomatosis).

Combination of R – bendamustine and rituximab keeps two uncommon types of non hodgkins lymphoma, indolent (slow growing) lymphoma and mantle cell lymphoma from worsening longer than standard chemotherapy. For a long time, the standard treatment for NHL has been rituximab plus chemotherapy with Cyclophosphamide (cytoxan), Doxorubicin (Adriamycin), Vincristine (Oncovin, Vincasar) and Prednisolone called as R CHOP.

Chylous ascites is accumulation of peritoneal fluid that is rich in triglycerides, generally a triglyceride level more than 200 mg/dl give rise to chylous ascites. Estimated incidence in United States is approximately 1 per 20,000 per ascites cases.[1]Most common causes for chylous ascites include trauma, malignancy and infections. Amongst pediatric age group congenital lymphatic anomalies such as lymphangectasias are most common described cause for chylous ascites.

Chylous ascites is milky appearance of peritoneal fluid due to excessive accumulation of triglyceride content in peritoneal fluid. Threshold limit for appearance of chylous ascites is triglyceride content more than 200 mg/dl. In a review from developing and developed countries that had atraumatic chylous ascites, the most common causes in adults were malignancy, cirrhosis and mycobacterium infection[2].In developing countries mycobacterium infection and fialariasis are the leading causes of chylous ascites[3].

Principle mechanism for formation of chylous ascites is discrepancy between formation and absorption of lymph either due to decreased absorption or increased production, in cirrhosis with portal hypertension, increased systemic venous pressure along with increased lymph formation is the purposed mechanism for development of chylous ascites[4] and decompression of portal venous system has been shown to reverse the ascites[5]

Abdominal paracentesis is important diagnostic tool in evaluation of patients with ascites and chylous ascites must be distinguished from pseudochylous ascites, in which the turbid appearance is due higher cellular content without actually containing high levels of triglycerides[6]. Fluid with an absolute neutrophil count less than 1000/mm^3 may be nearly clear. Ascitic Fluid with a count greater than 5000/mm^3 appears cloudy, and fluid with a count greater than 50,000/mm^3 resembles mayonnaise. SAAG (serum ascites albumin gradient) is an important diagnostic aid to differentiate ascites due to liver associated pathology or other and calculated SAAG >1.1 generally favors the hepatic etiology but elevated SAAG is not exclusive feature of hepatic

ascites it can be elevated in hypothyroidism and cardiac ascites also. Elevated protein >2.5 gm./dl and high SAAG clues towards cardiac ascites as opposed to hepatic ascites which generally contains low protein content except Budd Chiari Syndrome.

Computer tomography (CT) is an important adjunctive noninvasive imaging tool that may delineate the differentials of chylous ascites and can be helpful in management also. Ascites of unknown etiology is an indication for laparoscopy in patients with ascites when tuberculosis or malignancies are highly suspecious[7]. Positron emitting tomogram (PET) scan may give additional information regarding underlying metabolic activity of lesion but only drawback with PET is that it cannot differentiate between inflammation and tumor as both appear FDG avid and appear hot lesion on PET.

There are no definitive consciences on management of chylous ascites. Treatment of underlying cause is one of the most accepted options. Nutritional management in persons with chylous ascites include dietary modification to low fat, high protein and Medium chain triglyceride rich diet as medium chain triglycerides are directly taken up by blood capillaries rather than being transported to the lacteals. Orlistat has been shown to reduce ascites and triglyceride levels in ascetic fluid in a patient of cirrhosis [8].Octreotide is effective in the management of chylous ascites due to different causes because of its principle property of universal negative secreatalogue[9].

Large volume paracentesis and TIPS are another option in persons with refractory ascites due to cirrhosis[10]. Chylothorax associated with chylous ascites in background of cirrhosis or transudative chylothorax generally do not require drainage until it causes respiratory distress. It generally resolves by itself as ascites vanes off. Massive chylothorax associated with malignancy may require intercostal tube drainage for symptomatic benefit.

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