CLINICAL VIGNETTE

A Case of New Onset Chylous Ascites

Amar C. Nawathe, MD and Michael Lazarus, MD

A 61-year old male presented to the emergency department (ED) with abdominal pain, weight loss, and loose bowel movements. He noted abdominal distention, progressively increasing over the course of seven months and mild dyspnea related to his increased abdominal pressure. He denied early satiety, fevers, and night sweats. He estimated that he had about seventeen pounds of unintentional weight loss over the prior year. He denied nausea or vomiting but did report up to five loose bowel movements per day. He denied melena, hematochezia, or dysphagia. Bowel movements were watery, non-bloody and did not relieve his abdominal distention. His review of systems was negative for tuberculosis (TB) or TB risk factors, excessive alcohol use and he was not taking any medications or supplements. He had never travelled outside of the continental USA. He had a 32-pack year smoking history but had quit ten years ago. His Family history was notable for a mother with diabetes mellitus and hypertension and a father with prostate cancer and congestive heart failure. He was unmarried with a healthy younger sibling. He denied any prior surgical procedures or prior abdominal trauma. His remaining review of systems was negative. He lived with his elderly mother and younger brother. He worked as a bus driver for most of his adult life but retired recently. On physical exam his vital signs were within the normal range. He had marked, bilateral temporal wasting but no scleral icterus or lymphadenopathy in his neck or axillae. He had no elevated jugular venous pressure nor distended neck veins. His cardiac exam was unremarkable and he had clear bilateral breath sounds. He had reduced resonance over his lower chest bilaterally. Examination of his abdomen was significant for marked distention with positive shifting dullness of 2.5 cm and fluid wave. His abdominal exam was remarkable for distended superficial veins but no periumbilical nodes or hernias (Figure 1). He had moderate pain on palpation of his abdomen without rebound tenderness. There was no hepatomegaly or splenomegaly. The remainder of his physical exam was significant for muscle wasting, and signs of cachexia. His blood analysis included a normal complete blood count and electrolyte panel. His ALT 13 U/L and AST 22 U/L, Total Bilirubin 0.69 mg/dL, alkaline phosphatase 95 U/L, total protein 5.6 g/dL, albumin 3.2 g/d, lactate dehydrogenase 215 U/L, amylase 150.7 U/L, and lipase of 48 U/L. A bedside point-of-care ultrasound confirmed free fluid in the abdominal cavity and a diagnostic paracentesis was performed. Three liters of "light yellow and milky" fluid was removed (Figure 2). The triglyceride level in his ascitic fluid was 600 mg/dL, total protein content was 4.9 g/dL. The serum to ascites albumin gradient (SAAG) was calculated by subtracting the ascitic fluid value of albumin from the serum at 0.7 suggesting his ascites was less likely related to portal hypertension. The differential diagnosis for a SAAG < 1.1 includes Peritoneal Carcinomatosis, TB, and pancreatitis. Ascites was also sent for cell count, culture, Gram stain, total protein concentration, albumin, glucose, lactate dehydrogenase, amylase, and cytology. Magnetic Resonance Cholangiopancreatogram (MRCP) was significant for an enhancing mass involving the inferior pancreatic head and root of the mesentery, encasing the superior mesenteric vein and superior mesenteric artery. Multiple lesions within both hepatic lobes were noted with hypervascularity and retroperitoneal lymphadenoathy. A Positron Emission Tomogram (PET) scan showed a large hypermetabolic pancreatic head mass (4cm with SUV max 7) with evidence of nodal metastases to the mesenteric and bilateral retroperitoneal lymph nodes in addition to ascites and peritoneal carcinomatous. FNA of this mass was consistent with an islet cell tumor. An Octreotide scan, subsequently showed a large pancreatic head malignant mass, a solitary liver metastasis and multiple nodal metastases to the mesenteric and bilateral retroperitoneal lymph nodes all involved with somatostatin receptor rich neuroendocrine tumor. Measurement of urine serotonin and HIAA were both elevated and likely accounted for his watery diarrhea. He was treated with octreotide and everolimus but ascites and renal function worsened. He eventually opted for hospice care and died four months later.

Discussion

Chylous Ascites is defined as abnormal peritoneal fluid accumulation with a triglyceride level above 200 mg/dL, although some use a cutoff value 110 mg/dL.[1,2] In first world countries, over two-thirds of all cases are caused by intra-abdominal malignancy, lymphatic derangements, or cirrhosis. In the developing world, infection, most notably tuberculosis and filariasis is the most frequent culprit. Rarer conditions include congenital and inflammatory disorders, iatrogenic intraoperative lymphatic damage or trauma. Chylous ascites develops when there is a disruption of the lymphatic system, due to three underlying mechanisms. First, obstructed lymph flow due to malignancy, with leakage from lymphatics into the peritoneal cavity. Secondly, exudation of lymph through the walls of congenitally dilated retroperitoneal vessels (congenital lymphangiectasia).Thirdly, acquired thoracic duct obstruction from trauma or surgery, causing direct leakage of chyle through a lymphoperitoneal fistula.[3]

In our patient, malignancy was the cause of his marked ascites. Lymphoma accounts for at least one-half to one-third of the cases.[2] In most malignancies causing chylous ascites, direct obstruction and invasion into the lymphatics causes disrupted lymphatic flow.[4] Rarer oncologic causes include breast, esophageal, pancreatic, colon, renal, testicular, endometrial, cervical, ovarian, and prostate cancer; Kaposi sarcoma; carcinoid tumors; and lymphangiomyomatosis.[2] Carcinoid tumors should be excluded in patients with chylous ascites and secretory diarrhea as in our case.

Chylous ascites frequently presents as progressive and painless abdominal distention, occurring over weeks to months, depending upon the underlying cause. The most common presenting symptom was abdominal distension (81 percent) in a systematic review that included 131 studies (with a total of 190 patients) who had atraumatic chylous ascites.[4] Patients who have undergone abdominal or thoracic surgery may present with a more acute onset.

Patients may complain of weight gain and/or dyspnea resulting from increased abdominal pressure. Other features include nonspecific abdominal pain, weight loss, diarrhea and steatorrhea, malnutrition, edema, nausea, enlarged lymph nodes, early satiety, fevers, and night sweats.[5] However, in the majority of cases, the diagnosis is not suspected before performing a diagnostic paracentesis. When ascites is found clinically, the patient should be asked about weight loss or gain, symptoms of malignancy, family history, prior abdominal surgery, foreign travel, prior abdominal injury, and existing hepatic or renal disease. Typical physical examination findings include a fluid wave, shifting dullness, pleural effusions, lower extremity edema, lymphadenopathy, wasting, abdominal masses, and periumbilical hernias. Other clinical features of severe hepatic decompensation such as icterus, palmar erythema, spider angiomas, and encephalopathy should be sought. Abdominal paracentesis is the most important study in making the diagnosis. The total protein content can vary. A range of 2.5 to 7.0 g/dL has been suggested.[6] The serum to ascites albumin gradient is also helpful in making the diagnosis.[7] A complete ascitic fluid analysis should be sent for cell count, culture, Gram stain, total protein concentration, albumin, glucose, lactate dehydrogenase, amylase, and cytology.[8] A tuberculosis smear, culture, and adenosine deaminase (ADA) should be performed in selected cases when tuberculosis is suspected. Computed tomography (CT) of the abdomen can identify intraperitoneal adenopathy and masses. Lymphangiography and lymphoscintigraphy can detect abnormal retroperitoneal nodes, dilated lymphatics, fistulae, and thoracic duct anomalies.[9] The best study for diagnosing obstructed lymphatics is Lymphangiography (LAG), however tissue necrosis and fat emboli are serious adverse effects.[10]

Specific treatment of chylous ascites is determined by the etiology. Symptom control is aimed at reducing the ascites formation. Initial approaches for patients who have progressive ascites, a high-protein and low-fat diet with medium-chain triglycerides (MCT) is helpful. Dietary restriction of long-chain triglycerides (LCT) avoids their conversion into monoglycerides and free fatty acids (FFA), which are transported as chylomicrons to the intestinal lymph ducts. Low-fat diet with MCT supplementation reduces the production and flow of chyle.[11] Case reports have suggested that both somatostatin and subcutaneous octreotide are also effective in the management of chylous ascites.[12] This may be due to inhibition of lymph fluid excretion through specific receptors found in the normal intestinal wall of lymphatic vessels.[13] In patients with a large amount of ascites, a total paracentesis to relieve abdominal discomfort and dyspnea should be performed and repeated as needed.

In conclusion, based on a literature review, chylous ascites in patients with carcinoid tumors have a universally poor outcome.[14] Although neuroendocrine tumors are usually less aggressive, particularly in those patients with carcinoid tumors of midgut origin, it is definitely more aggressive in those patients presenting with chylous ascites.

Figures

Figure 1.

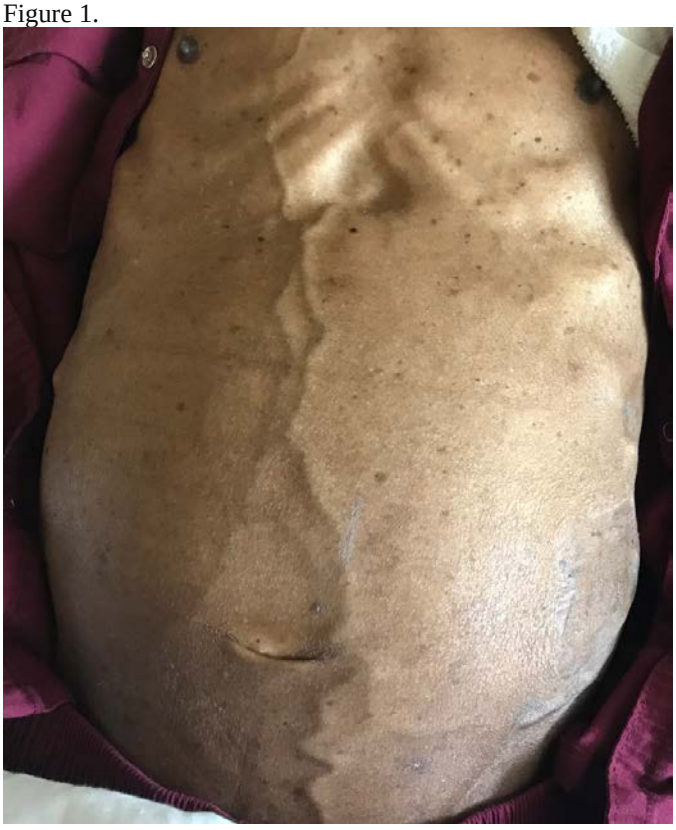

Figure 2.

REFERENCES

1. **Cárdenas A, Chopra S**. Chylous ascites. *Am J Gastroenterol*. 2002 Aug;97(8):1896-900. Review. PubMed PMID: 12190151.
2. **Press OW, Press NO, Kaufman SD**. Evaluation and management of chylous ascites. *Ann Intern Med*. 1982 Mar;96(3):358-64. PubMed PMID: 7059101.
3. **Browse NL, Wilson NM, Russo F, al-Hassan H, Allen DR**. Aetiology and treatment of chylous ascites. *Br J Surg*. 1992 Nov;79(11):1145-50. PubMed PMID: 1467885.
4. **Almakdisi T, Massoud S, Makdisi G**. Lymphomas and chylous ascites: review of the literature. *Oncologist*. 2005 Sep;10(8):632-5. Review. PubMed PMID: 16177287.
5. **Var T, Güngor T, Tonguc E, Ozdener T, Mollamahmutoğlu L**. The conservative treatment of postoperative chylous ascites in gynecologic cancers: four case reports. *Arch Gynecol Obstet*. 2012 Mar;285(3):849-51. doi: 10.1007/s00404-011-2050-3. Epub 2011 Aug 12. PubMed PMID: 21837421.
6. **Runyon BA, Hoefs JC, Morgan TR**. Ascitic fluid analysis in malignancy-related ascites. *Hepatology*. 1988 Sep-Oct;8(5):1104-9. PubMed PMID: 3417231.
7. **Runyon BA, Montano AA, Akriviadis EA, Antillon MR, Irving MA, McHutchison JG**. The serum-ascites albumin gradient is superior to the exudate-transudate concept in the differential diagnosis of ascites. *Ann Intern Med*. 1992 Aug 1;117(3):215-20. PubMed PMID: 1616215.
8. **Runyon BA**. Care of patients with ascites. *N Engl J Med*. 1994 Feb 3;330(5):337-42. PubMed PMID: 8277955.
9. **Archimandritis AJ, Zonios DI, Karadima D, Vlachoyiannopoulos PG, Kiriaki D, Hatzis GS**. Gross chylous ascites in cirrhosis with massive portal vein thrombosis: diagnostic value of lymphoscintigraphy. A case report and review of the literature. *Eur J Gastroenterol* Hepatol. 2003 Jan;15(1):81-5. Review. PubMed PMID: 12544699.
10. **Pui MH, Yueh TC**. Lymphoscintigraphy in chyluria, chyloperitoneum and chylothorax. *J Nucl Med*. 1998 Jul;39(7):1292-6. PubMed PMID: 9669413.
11. **Ohri SK, Patel T, Desa LA, Spencer J**. The management of postoperative chylous ascites. A case report and literature review. *J Clin Gastroenterol*. 1990 Dec;12(6): 693-7. Review. PubMed PMID: 2266250.
12. **Leong RW, House AK, Jeffrey GP**. Chylous ascites caused by portal vein thrombosis treated with octreotide. *J Gastroenterol Hepatol*. 2003 Oct;18(10):1211-3. PubMed PMID: 12974913.
13. **Karaca S, Gemayel G, Kalangos A**. Somatostatin treatment of a persistent chyloperitoneum following abdominal aortic surgery. *J Vasc Surg*. 2012 Nov;56(5): 1409-12. doi: 10.1016/j.jvs.2012.05.004. Epub 2012 Sep 20. PubMed PMID: 22999046.
14. **Onaitis MW, Kirshbom PM, Hayward TZ, Quayle FJ, Feldman JM, Seigler HF, Tyler DS**. Gastrointestinal carcinoids: characterization by site of origin and hormone production. *Ann Surg*. 2000 Oct;232(4):549-56. PubMed PMID: 10998653; PubMed Central PMCID: PMC 1421187.

Submitted April 22, 2019