Brief Report

DOI: 10.5582/irdr.2018.01028

Underlying etiology determines the outcome in atraumatic chylous ascites

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Summary **Chylous ascites is an uncommon entity and infectious etiology is the most common cause in developing countries. However, recently, whether there is any change in trend of etiologies in developing countries is not known. In this study, a retrospective analysis of the data of cases of atraumatic chylous ascites was conducted. Twelve patients of atraumatic chylous ascites with a mean age of 35 years were studied and 6 of them were males. The mean duration of symptoms was 9.6 months and the clinical presentation was abdominal distension (12 cases), pain abdomen (10 cases), loss of appetite and weight (9 cases), peripheral lymphadenopathy (4 cases) and fever (3 cases). Etiologies were tuberculosis (3 cases), malignancy (2 cases), radiotherapy related (2 cases), pancreatitis related (2 cases), lymphatic malformation (2 cases) and multifactorial (1 case). Eight improved with conservative measures, 2 were lost to follow up and 2 died. Our outcomes found infectious etiology still as the most common cause of atraumatic chylous ascites. Benign treatable causes could be managed successfully with conservative measures while malignant etiology had a poor prognosis. Underlying etiology determines the outcome in atraumatic chylous ascites.**

Keywords: Ascites, chylous, tuberculosis, lymphatic malformation, lymphoscintigraphy

1. Introduction

Chylous ascites, an uncommon entity, is characterized by presence of milky peritoneal fluid abundant in triglycerides, nutrients and immunoglobulins resulting from congenital or acquired abnormalities of abdominal lymphatics. Although various etiologies have been described that result in this clinical entity, v.i.z, malignancy, infection, abdominal surgery, trauma, cirrhosis among others, the condition is uncommon and chylous ascites accounts for a very small number of overall ascites cases and as per published literature the current incidence is 1 case per 20,000 admissions (*1*).

Traditionally it has been taught that the most common etiologies for atraumatic chylous ascites in developed countries are malignancy and cirrhosis and that in developing countries it is due to infectious etiologies like tuberculosis and filariasis (*1*). The data for atraumatic chylous ascites is scant as can be seen from a systematic review, which reported that only 21% of the studies in their literature search reported on atraumatic chylous ascites (*2*). Also the data of atraumatic chylous ascites from India is scarce and we do not know whether the trend of etiologies remains the same or has changed in developing countries like India.

Here, we report our experience with patients who presented with atraumatic chylous ascites from North India and report about underlying causes, clinical presentation and outcomes in these patients.

2. Patients and Methods

This is a retrospective analysis of our prospectively maintained database of patients who presented with chylous ascites over a period of 3 years from 2014 to 2017. We evaluated clinical presentation, etiologies, treatment modalities used and the outcome. The diagnosis was suspected in patients who had milky

Released online in J-STAGE as advance publication August 15, 2018.

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hue of the fluid and this was tested for triglyceride level. The cut off triglyceride value to label the case as chylous ascites was taken > 110 mg/dL. We present our experience of these patients who had chylous ascites.

3. Results and Discussion

3.1. *Clinical presentation*

Over a period of 3 years, 12 patients who presented with chylous ascites were seen (Table 1). The mean age was 35 years (range 15-72) and 6 (50%) were males. The mean duration of symptoms was 9.6 months (range 0.5-84 months). The presenting complaints in decreasing order were abdominal distension in 12 cases (100%), pain abdomen in 10 cases (83.3%), loss of weight and appetite in 9 cases (75%), peripheral lymphadenopathy in 4 cases (33.3%) and fever in 3 cases (25%) patients. The mean triglyceride value was 507 mg/dL (range 130-1,600 mg/dL).

3.2. *Etiologies*

3.2.1. Tuberculosis

Three patients had tuberculosis as the etiology of chylous ascites. All 3 were young females aged between 25-26 years and presented with 2-6 months history of abdominal distension along with loss of appetite and weight and fever. While one of them, a 25-year-old, had history of genitourinary tract tuberculosis and was a defaulter and her ascitic fluid examination showed milky fluid with triglycerides of 1,600 mg/dL with low serum ascites albumin gradient (SAAG) of 0.5 and high ADA of 45 IU/L. She had peripheral lymphadenopathy and the FNAC from retroperitoneal lymph nodes showed granulomatous inflammation with AFB stain positive and also was GenXpert positive for tuberculosis. She was started on standard 4 drug antitubercular therapy (ATT) along with octreotide and medial chain triglyceride (MCT) based diet and at 6 months of follow up she had gained weight and the ascites had disappeared.

Another 25-year-old female had a ascitic fluid triglyceride of 323 mg/dL with low SAAG of 0.3 and ADA of 18 IU/L. She too had peripheral lymphadenopathy, and chest x ray showed military shadows and abdominal CT scan showed gross ascites with omental nodularity with a sheet like lymph nodal mass encasing the inferior vena cava with calcification. The FNAC from this lymph nodal mass showed granulomatous inflammation with AFB stain positive. She was started on standard 4 drug ATT with MCT, however, subsequently she died of ATT induced liver failure.

The third patient a 25 year old female presented with above the complaints and ascitic fluid triglyceride was 400 mg/dL with low SAAG of 0.2 and high ADA of 74 IU/L. Her abdominal CT scan showed gross ascites with multiple mesenteric lymph nodes. She had cervical lymphadenopathy and its FNAC showed granulomatous inflammation and AFB stain was positive. She was started on standard 4 drug ATT with MCT based diet and after completion of 6 months of ATT and a further 5 months follow up is doing fine with disappearance of ascites and weight gain. All 3 patients were HIV negative.

Table 1. Summary of the cases of atraumatic chylous ascites

Characteristics	Summary
Age mean (range)	35 years (15-72)
Males	6 cases
Mean duration of symptoms	9.6 months (range 0.5-84)
Clinical presentation	Abdominal distension: 12 cases (100%) Pain abdomen: 10 cases (83.3%) Loss of weight and appetite: 9 cases (75%) Peripheral lymphadenopathy: 4 cases (33.3%) Fever: 3 cases (25%)
Mean triglyceride value	507 mg/dL (range 130-1600)
Etiologies	Tuberculosis: 3 cases Malignancy: 2 cases Radiotherapy related: 2 cases Pancreatitis related: 2 cases Lymphatic malformation (Milroy disease and primary intestinal lympangiectasia): 2 cases Multifactorial: 1 case
Outcome	Improved with conservative measures: 8 Lost to follow up: 2 cases Died: 2 cases

3.2.2. *Malignancy*

Two patients had malignancy associated chylous ascites. A 72-year-old female with history of diabetes mellitus and coronary artery disease presented with a 1 ½ year history of loss of weight of around 25 kg and abdominal pain and distension for 2 weeks. The CECT abdomen showed a 1.2 × 1.3 cm hypodense lesion in the body of pancreas with multiple hypodense lesions in both lobes of liver with gross ascites. The ascitic fluid triglyceride was 130 mg/dL with high SAAG of 2.0, however, the malignant cytology was negative. The FNAC from the pancreatic lesion showed features of adenocarcinoma. She was discharged on MCT based diet and asked to follow under the department of radiotherapy, but was lost to follow up.

The second patient was a 16-year-old male who presented with 3 months history of abdominal distension, generalized weakness and weight loss of 10 kg. The ascitic fluid triglycerides were 241 mg/dL and ascitic fluid malignant cytology was positive. He also complained of nausea and postprandial vomiting. We did an esophagogastroduodenoscopy that revealed thickened gastric fold to the extent that the scope was not negotiable beyond the antrum (Figure 1). The histopathology from the stomach biopsy showed poorly differentiated signet ring cell carcinoma. We advised MCT based diet and explained the prognosis of this metastatic disease to the family after which they took discharge and were lost to follow up.

3.2.3. *Radiation induced*

Two patients developed radiation induced chylous ascites. A 40-year-old female who was a known case of carcinoma of the cervix had received chemo radiotherapy for the disease. After completion of her radiotherapy cycles she was asymptomatic for 4 months after which she noticed gradually progressive abdominal distension over a period of 6 months. Her ultrasound abdomen showed gross ascites with fluid triglyceride of 600 mg/dL, however, the malignant cytology was three times negative. She was started on MCT based diet with octreotide and improved with it.

Another patient a 39-year-old male had unresectable carcinoma of the esophagus for which he received radiotherapy, however, later developed constrictive pericarditis and then presented with abdominal distension and had gross ascites with fluid triglyceride of 512 mg/dL. He was started on MCT based diet, but developed recurrence of the primary malignancy and later on succumbed to it.

3.2.4. *Pancreatitis related*

A 15-year-old male presented with abdominal pain for 3 months and distension for 1 month. His Serum amylase was 1,160 U/L and CT scan revealed dilated main pancreatic duct (MPD) with gross ascites. Ascitic fluid was milky with triglyceride of 281 mg/dL and amylase of 7,000 U/L. All etiologic work up for chronic pancreatic disease was negative so the case was labeled as idiopathic chronic pancreatitis with pancreatic chylous ascites and was started on MCT based diet and octreotide, however ascites persisted so the patient was taken up for ERCP which revealed hugely dilated, tortuous MPD with contrast extravasation at the head region of pancreas and a 5 F stent was placed across the disruption. Patient showed clinical improvement and 4 weeks later a repeat CT scan revealed disappearance of ascites with stent in situ.

Another patient a 35-year-old chronic alcoholic male presented with abdominal pain and distension over 15 days and CT scan revealed non-enhancing pancreas with gross ascites. The Fluid appeared milky with triglyceride of 220 mg/dL and amylase of 19,950 U/L. The patient

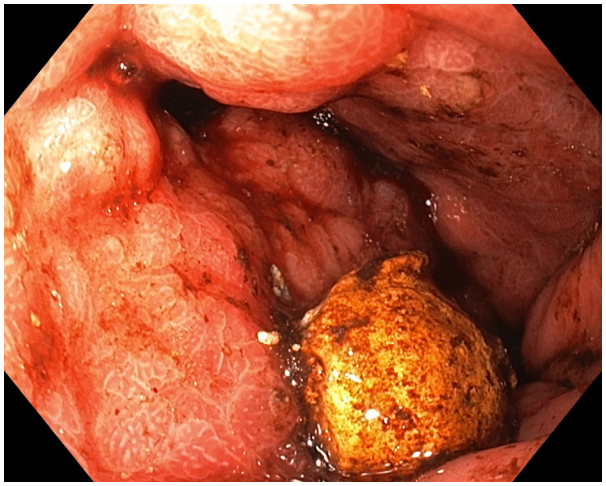

Figure 1 showing unhealthy edematous gastric folds with narrowing in the antral region with a food bolus.

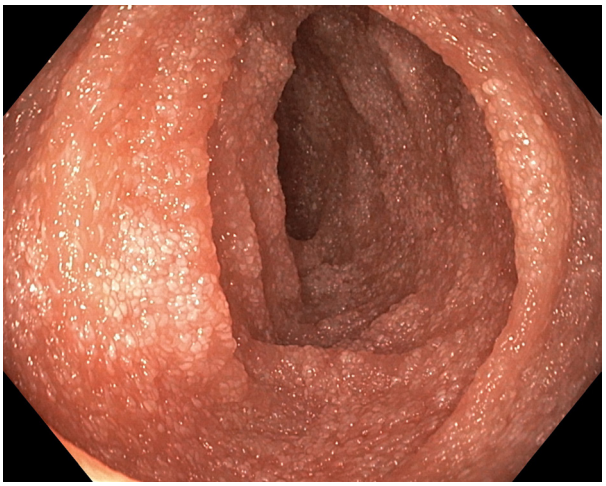

Figure 2 showing granular edematous mucosal surface of duodenum with scattered whitish specs throughout the duodenum.

was febrile with total leucocyte count of 23,600/dL so was started on antibiotics and pigtail drainage was done and was started on MCT based diet with octreotide and improved with the above measures.

3.2.5. *Lymphatic malformation*

A 34-year-old male with prolonged history of bipedal edema over 10 years presented with gradually progressive abdominal distension over 2 months. The fluid triglyceride was 206 mg/dL and MR lymphangiogram revealed lymphatic channels absent above the ankle suggestive of Milroy's disease. Patient was started on MCT based diet and improved gradually with disappearance of ascites in 2 months.

Another patient a 42-year-old male with history of diarrhea with malabsorption over 7 years presented with abdominal distension over 3 months. Ascitic fluid had low SAAG of 0.6 and triglyceride of 560 mg/dL and esophagogastroduodenoscopy revealed a granular edematous mucosal surface of duodenum with scattered whitish specs throughout the duodenum (Figure 2). The biopsy was suggestive of lymphangiectasia. Patient was started on MCT based diet and improved.

3.2.6. *Multifactorial*

A 51-year-old female with previous co morbidities in the form of hypertension and hypothyroidism presented with progressive abdominal distension over 2 months. Four years back she was diagnosed as a case of chronic pancreatitis and had developed a symptomatic large pseudocyst that required cystogastrostomy and also had extrahepatic cholestasis due to common bile duct stricture resulting from chronic pancreatitis and had undergone choledochojejunostomy. This time along with abdominal distension she had pedal edema, pallor with hemoglobin of 8.2 gm % and deranged renal function with creatinine of 6.2 mg/dL and 24-hour urine protein of 7 gm. Her ultrasound abdomen revealed a shrunken liver with irregular outline and raised echotexture and esophagogastroduodenoscopy revealed portal hypertensive gastropathy. Her ANA titer was 1:80 and SMA titer 1:40. All other work up for etiology of chronic liver disease was negative. Ascitic fluid showed low SAAG 0.2 with triglycerides of 1,010 mg/dL and the renal biopsy showed features suggestive of membranous lupus nephritis. The etiology of chylous ascites was thought to be multifactorial with contributions from cirrhosis and lupus and she was started on steroids with octreotide, MCT based diet and diuretics and improved dramatically with disappearance of ascites and improvement of renal function.

3.2.7. *Discussion*

Our series is a compilation of cases of atraumatic chylous ascites seen in the medical gastroenterology unit from 2014 till 2017. In our series infection due to tuberculosis was the most common etiology suggesting that the trend still has not changed and the traditional teaching that malignancy and cirrhosis are the causes of atraumatic chylous ascites in developed and infection due to tuberculosis/filariasis in developing countries still remains the same (*1*). Our study highlights the fact that a benign treatable etiology for atraumatic chylous ascites has good prognosis with most recovering with conservative measures along with treatment of the underlying etiology, however, malignancy will have a poor prognosis as 1 of the 3 patients with malignancy died (one who had constrictive pericarditis due to radiotherapy ultimately died due to recurrence of the primary esophageal malignancy) and the other 2 patients who also had a disseminated malignancy were lost to follow up but the prognosis in such a situation is expected to be adverse. Two of our patients had lymphatic malformation (Milroy disease and primary intestinal lymphangiectasia) and both of them had resolution of the chylous ascites with conservative measures suggesting a good prognosis. In lymphatic malformations where conservative measures fail one can get a pre-operative lymphoscintigraphy which can show back flow and/or a lymphangiography that can clearly show dysplastic areas and sites of lymphatic leaks. Presence of localized abdominal dysplasia, lymphatic blocks, evidence of back flows, ectatic lymphatic vessels and tissue areas and lymphangiomatosis can be dealt with surgery. The lymphangiectasias and lymphangiomatosis are sclerosed with polidocanol and then are surgically debulked. The incompetent lymph vessels can be surgically ligated in a "stair like" manner. The ectatic vessels with low degree of dilatation can be welded with the help of a CO_2 laser. In presence of proper chylous and lymphatic vessels, Chylovenous or lymphovenous microsurgical shunts can be performed in iliac or mesenteric areas (*3*).

None of our patients required surgery as most of our patients had benign treatable causes and all resolved with conservative therapy. The role of surgery comes when we have refractory chylous ascites not responding to the conservative measure in the form of low fat, high protein, medium chain triglyceride based diet with or without somatostatin or octreotide and diuretics and in some cases total parenteral nutrition in which case a pre-operative or intra operative lymphangiogram helps to localize the leak and then a fistula closure or bowel resection can be done (*4*). Where intervention radiology facilities with expertise in lymphangiography and embolization are available the need for surgery can be circumvented with good results, however, sometimes the conventional lymphangiography may not pick up the leak in such cases an MR lymphangiogram and recently balloon occluded retrograde abdominal lymphangiography and embolization have been found useful (*5,6*). The peritoneovenous shunt has been

abandoned due to high rate of shunt block with repeated replacement and also serious complications (*7*).

To conclude infective causes still lead the list of etiology of atraumatic chylous ascites in developing countries like India. A benign treatable cause in a majority of cases recovers with conservative measures with dietary modification with or without octreotide/ somatostatin and diuretics and treating the underlying cause while malignancy portends a poor prognosis.

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(Received March 27. 2018; Revised May 29, 2018; Accepted June 21, 2018)