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Case Report

Peritoneal Lymphomatosis in a case of ascites with portal hypertension: a case report.

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Abstract

Peritoneal Lymphomatosis is rare and should be one of the differential diagnoses in patients with intractable ascites of unknown etiology. Its diagnosis is difficult and has to be differentiated from peritoneal carcinomatosis and tubercular peritonitis which are more common. We present a case of an elderly gentleman who presented with symptoms of abdominal distension, anorexia and weight loss. On examination he had pedal edema with gross ascites and splenomegaly without jaundice Ascitic fluid analysis revealed high SAAG, lymphocyte predominant and negative for malignant cells. Transjugular liver biopsy was normal with elevated HVPG suggestive of portal hypertension. Ascites with portal hypertension can be easily assumed to be due to cirrhosis of liver which is extremely common, if proper evaluation is not done. Subsequently diagnostic laparoscopy revealed ascites, omental thickening, peritoneum studded with deposits and biopsies confirmed Peritoneal Lypmhomatosis due to Follicular Lymphoma.Diagnostic laparoscopy is a useful tool in the accurate diagnosis of peritoneal diseases. With timely and correct diagnosis, Peritoneal Lymphomatosis can be treated with chemotherapy to improve the outcome.

Keywords: Diagnostic Laparoscopy; Follicular Lymphoma; Malignant ascites; Non-Hodgkin`s Lymphoma; Peritoneal Lymphomatosis.

Methods

In India, the most common causes of ascites are cirrhosis of liver (55 to 60 %) followed by tuberculosis (13 to 30 %) and malignancy (8 %) [1]. Malignant ascites is more commonly caused by metastatic spread from ovarian, colorectal, and pancreatic cancers known as Peritoneal Carcinomatosis (PC). Peritoneal Lymphomatosis (PL) defined as the peritoneal spreading of lymphoma, is an extremely rare condition. Lymphoma usually does not invade the omentum, because it is fibro-fatty and lacks lymphoid tissue [2, 3]. PL can occur as a Primary peritoneal lymphoma or secondary involvement from a visceral lymphoma. Even though there is no direct comparison in the literature, secondary is generally much more common than primary peritoneal lymphoma. Primary peritoneal lymphoma also known as body cavity lymphoma or primary effusion lymphoma (PEL) is seen almost exclusively in patients with human immunodeficiency virus (HIV) [4]. PL more commonly occurs with Non-Hodgkin`s lymphoma (NHL) and the association with Hodgkin`s lymphoma is rare. Diffuse large B-cell lymphoma (DLBCL) is the most common NHL associated with PL [5]. The differentiation between PC and PL is crucial since they have different modalities of treatment altogether. We present a case of a patient presenting to us with ascites and also having portal hypertension without cirrhosis of liver, ultimately diagnosed to have PL due to Follicular Lymphoma (FL).

Case Report

A 66-year-old male with diabetes, non-alcoholic presented with complaints of progressive painless abdominal distension, anorexia, significant weight loss of 20 kgs, and bilateral lower limb swelling for 3 months. There was no history of jaundice, hematemesis, melena, or altered sensorium in the past. The patient was started on anti tubercular treatment elsewhere, based on high protein in the ascitic fluid to which he did not respond.

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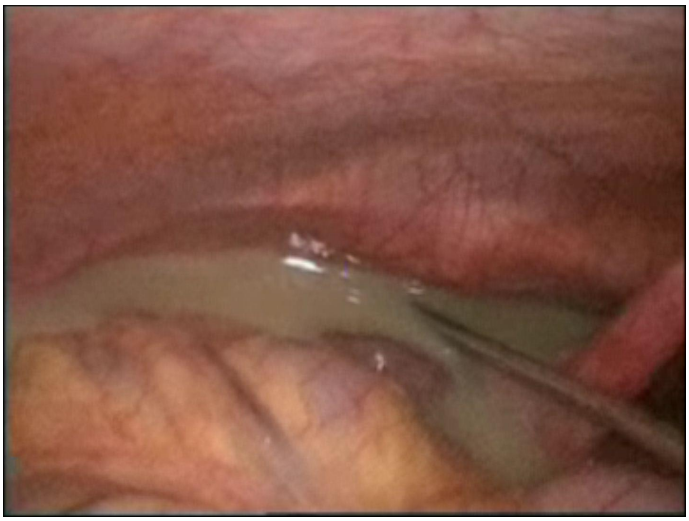
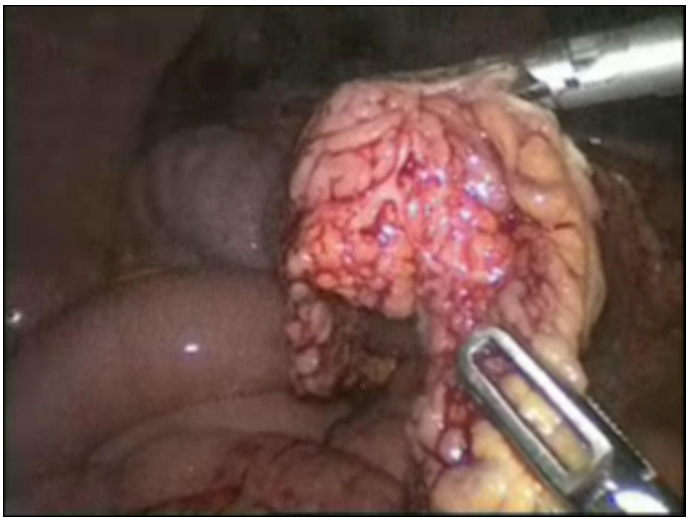
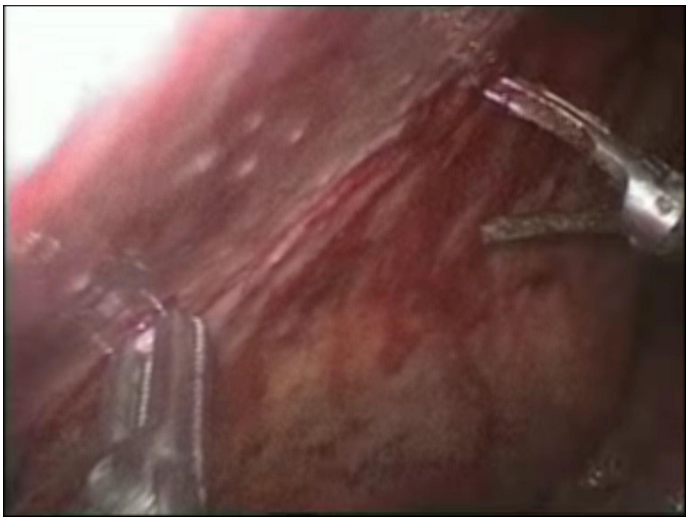
On examination, the patient had pallor, edema feet, gross ascites, splenomegaly , no icterus, no lymphadenopathy. Laboratory investigations revealed Haemoglobin 9.7g/dL, Total leucocyte count 8400/mm3, Platelets 1.5 lacs/mm3. T.Bilirubin-0.3 mg/dL, SGPT/SGOT- 23/16 U/L, Alkaline phosphatase-112 IU/L,T. Proteins-7.5 g/dL,Albumin-2.6 g/dL,Globulin-4.9 g/dL, INR-0.99,Creatinine-1.9 mg/dL, electrolytes were normal, LDH-156 U/L(normal 140-280 U/L),Uric Acid-14.9 (normal 3.4-7.2 mg/dL). Elevated uric acid and LDH can be due to the rapid growth of tumors. HIV, HBsAg, and Anti HCV were non-reactive. Ultrasound scan of the abdomen showed a normal sized liver with altered echotexture without any biliary obstruction, overdistended Gall Bladder with echogenic sludge, gross splenomegaly and gross ascites. Color Doppler of abdominal veins showed narrowing of hepatic veins and high velocity flow most likely to be stenosis, multiple intrahepatic collaterals likely Budd Chiari syndrome; patent and prominent portal and splenic veins with changes of portal hypertension. Upper GI endoscopy showed no esophageal varices. The Ascitic fluid analysis: Total cells: 495 cells/mm3 with 80% lymphocytes, Proteins: 3.9 g/dL, Albumin: 1.5 g/dL, and serum-ascites albumin gradient (SAAG) 2.1. No microorganisms were detected, and acid-fast stain of ascites was negative. Cytology: Scattered Lymphocytes with mesothelial cells and no malignant cells. A CT Abdomen with contrast could not be done because of raised creatinine. Our first Differential Diagnosis was Budd Chiari Syndrome. For confirmation, a Transjugular liver biopsy, hepatic angiography and pressure measurements were done. On angiography, all hepatic veins were patent and the liver biopsy was normal. The Wedged hepatic venous pressure (WHVP) =28 mmHg, Free hepatic venous Pressure (FHVP) =18 mmHg, and Hepatic venous pressure gradient (HVPG) =10 mmHg. HVPG is equal to WHVP minus FHVP. An HVPG of > 5 mmHg defines portal hypertension [6]. Diagnostic Laparoscopy (DL) revealed omental caking with rolled up omentum, peritoneal surface studded with deposits (shown in Fig. 1- 3). Liver was enlarged with irregular surface and Splenomegaly (shown in Fig. 4). Biopsies were obtained from multiple peritoneal sites. The histopathological examination (HPE) revealed NHL of follicular type, grade II (WHO 2008). On Immunohistochemistry (IHC) the tumor cells expressed CD 20, CD 10, bcl-6 & bcl-2 and were immunonegative for CD 3, CD 5, cyclin D1, MUM-1 & C-MYC. CD 21 demonstrated remnant follicular dendritic cell meshworks in the lesional follicles. After creatinine stabilized positron emission tomography (PET CT) of the whole body was done for staging the disease. Hypermetabolic enlarged multiple supra and infra diaphragmatic nodes with enlarged spleen and multifocal marrow involvement was reported favoring a possibility of a lymphoproliferative disorder. Bone Marrow aspiration and biopsy showed hypercellular marrow with erythroid hyperplasia with the differential diagnosis of 1.Chronic myeloproliferative disorder with trilinear hyperplasia 2.Polycythemia vera. The patient was then referred to an oncologist and started on Rituximab based chemotherapy.



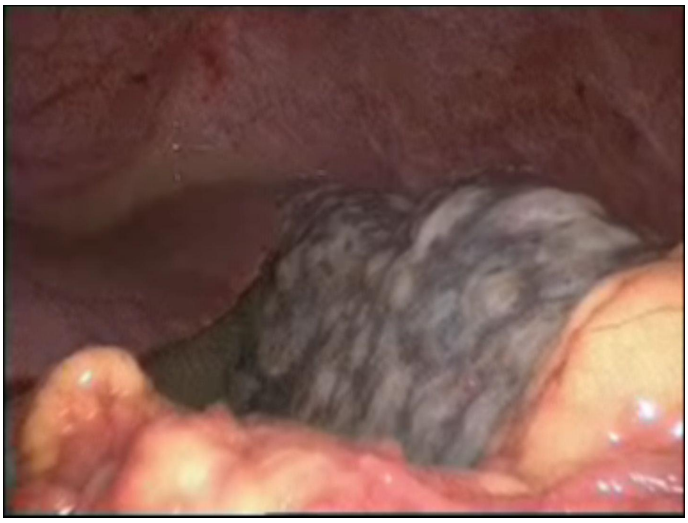
(Fig. 1. Ascitic fluid seen in peritoneal cavity)



(Fig. 2. Omental thickening)



(Fig. 3. Peritoneal surface studded with deposit)



(Fig. 4. Gross Splenomegaly. (Arrow shows enlarged spleen)

Discussion

FL and DLBCL account for more than half of the cases of NHL. FL falls in the group of indolent lymphomas which are slow-growing, whereas DLBCL is an aggressive type of lymphoma and Burkitt lymphoma is included in the highly aggressive group. The peritoneal surface may be secondarily affected by 3 cell lines: epithelial (carcinomatosis), mesenchymal (sarcomatosis) and lymphoid (lymphomatosis) [5]. Although lymphoma can occur at any site of the body, diffuse and extensive involvement of the peritoneal cavity is rare and only a few cases have been reported . Kurtz RC et al. reported lymphomas as the underlying cause for ascites in 8% of cases among 101 cases of malignant ascites. Das et al. in an excellent review cited T-cell lymphomas to be associated with serous effusions much more frequently than did B-cell neoplasms. We present an unusual case of Follicular lymphoma with PL without any obvious primary site. The typical scenario of PL is the occurrence of primary tumors in the gastrointestinal tract, most commonly the stomach and terminal ileum [3]. PL is more commonly associated with high-grade lymphomas, but in our case it was a low-grade lymphoma . The patient presented to us with ascites and non-specific symptoms without any obvious pointers to its etiology on history and physical examination. The initial reports of ultrasound abdomen and venous doppler were hinting towards Budd Chiari syndrome but angiography confirmed that all the hepatic veins were patent. The patient had portal hypertension as suggested by elevated HVPG. The high SAAG ascites could have been caused by portal hypertension or possibly mixed ascites, though the ascitic fluid cytology was negative for malignancy. Ascitic fluid cytology has an overall sensitivity of around 60% to 70% in diagnosing malignant ascites. Additionally, lymphoma can evoke mesothelial hyperplasia, affecting the cytology results. Therefore, a tissue specimen must be obtained for histopathological examination (HPE) if possible, which is the gold standard for diagnosis of PL Hence, a decision of diagnostic laparoscopy (DL) was made. In a study by the same author, Ascites of unknown cause was the commonest indication for DL. The advantage of DL is that lesions less than 1 cm and the parietal peritoneum which cannot be assessed by the imaging techniques can be adequately visualized. DL can identify lesions as small as 1 to 2 mm in size which can be biopsied precisely under direct vision and provides the capability to obtain large histological specimens as compared to imaging-directed biopsies which are more of a cytological than histological examination . Also, like in our situation where a contrast-based imaging could not be done due to renal failure, DL can serve as a very useful modality in diagnosing peritoneal diseases. Omental involvement in the course of lymphomas is uncommon because the omentum does not contain lymphoid tissue. Although, the route of this dissemination is not completely clear, the presumed way of spread is believed to be via the pathways like gastrocolic ligament, transverse mesocolon and visceral peritoneal surface [14]. The basic mechanisms of ascites formation in PL are due to lymphatic obstruction and increased permeability of peritoneal vessels [3].The HPE and IHC confirmed the diagnosis of FL. FDG-PET/CT was done to stage the disease. It is also helpful in evaluating the chemotherapy response. On CT, PL is characterized by diffusely thickened peritoneal surfaces with multifocal nodules and masses on that mimic PC. Other CT features of PC include ascites, peritoneal enhancement and thickening, omental caking and infiltration of the small bowel mesentery. The presence of extensive lymphadenopathy exclusively in the retrocrural region and small bowel mesentery may suggest lymphomatosis over carcinomatosis [3]. Development of ascites is a manifestation of the end-stage disease of lymphoma, a poor prognostic feature and associated with reduced overall survival . PL does not require surgery and is amenable to chemotherapy in contrast to other malignant peritoneal diseases which usually require surgery [2]. The patient was started on Rituximab based chemotherapy by the oncologist after ruling out occult hepatitis B infection. The response to treatment could not be determined as the patient lost to follow up.

Conclusion

PL is considered a rare manifestation of NHL, characterized by systemic involvement and rapid clinical deterioration often leading to death. Therefore, it is extremely important to provide correct and early diagnosis in order to provide treatment and prolong survival. In lymphoma cases surgical intervention can contribute to disease progression and additional clinical deterioration. Image-guided needle biopsy or laparoscopic peritoneal and omental biopsy appears to be the gold standard method for diagnosing PL. Therefore, they should be used in order to make the diagnosis preoperatively and every effort should be made to avoid unnecessary laparotomy or massive surgery whenever possible.

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