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# Peritoneal Lymphomatosis: The Failure of Occam's Razor

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## BACKGROUND

- During the last two decades the incidence of lymphomas has increased with extranodal lymphomas increasing more rapidly than nodal.<sup>1</sup>
- Primary Extranodal-Non Hodgkins lymphoma (PE-NHL) account for about 25%–40% of non-Hodgkin's lymphoma (NHL) among which primary gastric non-Hodgkin's lymphoma account for the most.
- Peritoneal lymphomatosis (PL) is a very rare type of PE- NHL seen in the setting of aggressive NHL.
- It is characterized by diffuse intra-abdominal peritoneal involvement and can easily be misdiagnosed as carcinomatosis especially when presenting as the sole initial manifestation.
- Early diagnosis is paramount in these patients to have effective and potentially curative management.

## CASE PRESENTATION

- A 72-year-old male presented with a 2-week history of abdominal swelling, discomfort and constipation.
- He denied fever, night sweats, nausea, vomiting, weight loss or jaundice. His medical history included hypertension and dyslipidemia.
- He denied having alcohol or tobacco use, or illicit drug use.
- Physical exam revealed tight, distended abdomen with shifting dullness.
- A CT scan demonstrated ascites in the presence of multiple intra-abdominal masses, omental caking appearing as nodular enhancement along the omentum, and enhancement of the peritoneal lining extending into the pelvis (Figure 1. A,B,C).
- Carcinoembryonic antigen and CA19-9 levels were tested and reported normal. His colonoscopy was unrevealing with only extrinsic compression seen.
- He underwent a diagnostic and palliative paracentesis which provided immediate symptom relief.
- Ascitic fluid studies showed white blood cell count (WBC) of 5100 cells / mm<sup>3</sup>, polymorphonuclear leukocyte (PMN) count 102 cells/mm<sup>3</sup>, lactate dehydrogenase level (LDH) 509 U/L (serum LDH 264 U/L), glucose 64 mg/dL, total protein 4.5 g/dL, albumin 2.9 g/dL and triglyceride 80 mg/dL. Serum-to-ascites albumin gradient (SAAG) was 0.9 g /dL.
- He was admitted for exploratory laparotomy and potential bowel resection.
- By this time, ascitic fluid cytopathology was resulted (Figure 2). Flow cytometry showed a monoclonal B-cell population within the large cell gate which demonstrated lambda light chain restriction, dim-moderate CD19, dim CD20, dim CD10 and dim CD38 (Figure 3).
- The immunophenotypic findings combined with the cytologic features were diagnostic for an aggressive diffuse large B-cell lymphoma (Figure 4).
- Surgery was aborted and he subsequently received 6 cycles of chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone.
- Post chemotherapy CT scan showed complete resolution of peritoneal nodularity and ascites (Figure 1. E,F,G) and PET-CT scan confirmed complete response (Figure 1. D & H).
- He continues to be in remission at 1 year follow-up.

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## DISCUSSION

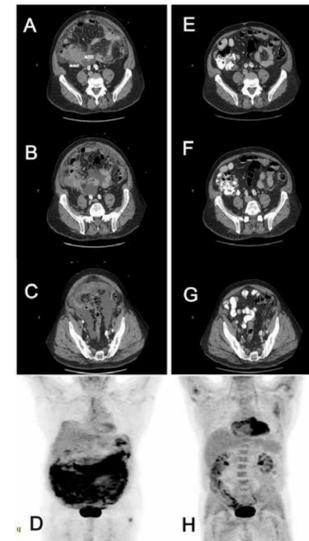
- PL is a rare clinical-pathological entity, defined by the presence of intraperitoneal spread of lymphoma, that closely mimics peritoneal carcinomatosis.
- Peritoneum is predominantly fibro-fatty tissue and is devoid of lymphoid tissue, which makes the existence of PL intriguing.
- CT findings that point to lymphoma includes bulky homogeneous masses or smooth peritoneal soft tissue thickening, diffuse lymphadenopathy, variable extranodal lymphomatous involvement manifesting as peritoneal enhancement or peritoneal thickening, which can be linear or nodular.<sup>4,5</sup>
- Primary diagnosis of lymphomas from peritoneal involvement and ascitic fluid is extremely challenging, hence a high index of clinical suspicion is warranted to diagnose this entity prior to embarking on cytoreductive surgeries.
- A routine ascitic fluid cytology is highly specific but the diagnostic sensitivity is only in the range of 40-60%.
- Ascitic fluid findings that point to peritoneal lymphomatosis include elevated protein level, disproportionately elevated WBC compared to PMN count, high LDH, low glucose and sterile ascites.<sup>1</sup>
- The association of lymphoma to chylous ascites (usually with triglyceride level > 200mg / dL) is well known, however lymphoma can be present with a normal ascitic fluid triglyceride level, as in our patient.<sup>2</sup>
- With a SAAG < 1.1 g/dl, carcinomatosis and tuberculosis were considered in the differential diagnosis. Tuberculous peritonitis was not suspected in our case with a negative history of tuberculosis exposure. Ancillary studies, such as flow cytometry and immunohistochemistry are often invaluable in these cases in establishing an accurate diagnosis.<sup>2,3</sup>

## CONCLUSION

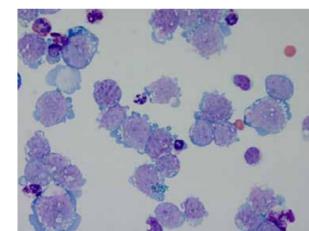
- PL presenting with extensive peritoneal involvement is extremely rare.
- The case highlights the importance of awaiting cytology and immunophenotyping in the presence of peritoneal disease and ascitis of unknown primary as this could have potential implication in management.

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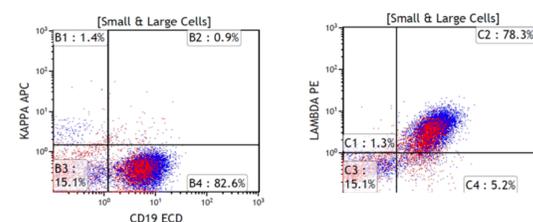
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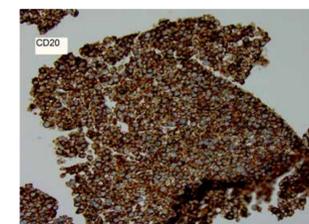
**Figure 1:** A, B, C CT scan showing multiple intra-abdominal masses, omental caking and enhancement of the peritoneal lining extending into the pelvis. E, F, G post chemotherapy CT scan showing complete response. D, H – Pre and post treatment PET – CT scan.



**Figure 2:** Ascitic fluid examination showing medium and large lymphoid cells with vacuolated cytoplasm, round nuclei with vesicular chromatin and inconspicuous nucleoli.



**Figure 3:** Flow cytometry showing monoclonal B-cell population within the large cell gate.



**Figure 4:** Paraffin immunohistochemistry performed demonstrating the lymphoma cells to be: CD20+.