

Peritoneal lymphomatosis mimicking peritoneal carcinomatosis – a clinical dilemma: Series of two cases and review of literature

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ABSTRACT

Peritoneal lymphomatosis is a rare presentation of non-Hodgkin lymphoma and most often associated with abdominal organ involvement. The diagnosis is often difficult and needs to be differentiated from peritoneal carcinomatosis and tubercular peritonitis. Imaging, though helpful in disease staging, may not lead to a specific diagnosis; which rests on the histopathological findings. We present two cases of peritoneal lymphomatosis with different presentations and extranodal sites of involvement.

Key words: Burkitt lymphoma, contrast enhanced computed tomography, non-Hodgkin lymphoma, peritoneal lymphomatosis

INTRODUCTION

Diffuse peritoneal seeding can be seen in a number of primary neoplasms including ovary, breast, and gastrointestinal tract tumors.^[1] Peritoneal dissemination as a manifestation of aggressive lymphoma, is rare.^[2,3] We report two cases of non-Hodgkin's lymphoma (NHL) presenting with peritoneal dissemination.

CASE REPORTS

Case 1

An 8-year-old male child presented with gradual onset of painless abdominal swelling since last 6 months, associated with alteration in bowel habit. On examination, he was afebrile, had pallor. No peripheral lymph nodes were palpable. The abdomen was soft and doughy without any tenderness or organomegaly. Contrast enhanced computed tomography (CECT) abdomen was performed

which revealed gross peritoneal thickening (mesentery as well as omentum) and aneurysmal thickening of isolated pelvic ileal loop [Figure 1]. A biopsy of the peritoneal deposit revealed diffused large B cell lymphoma. He was put on high dose chemotherapy and showed symptomatic improvement.

Case 2

A 28-year-old female presented to the gynecology department with menorrhagia for 2 months and a sense of heaviness in bilateral breasts. She also had a vague lower abdominal pain for 1 month. On examination she had pallor; the breast examination revealed a large well-defined mass lesion involving the left breast. No peripheral lymph nodes were palpable. The abdomen was distended and no definite mass lesion was palpable. Her hemoglobin was 6.5 gm/dl, white blood cell count 9,100/mm³ with 70% neutrophils, and platelet count 22,000/mm³. The CA-125 level was 396 U/ml; renal function tests were within normal limits. Imaging included a breast ultrasound which revealed a hypochoic well-defined mass lesion in the left breast. An abdominal CECT was performed; which revealed a large homogenous mass lesion in the left breast [Figure 2a]. There was ascites and omental thickening [Figure 2b]. Bilateral large solid ovarian masses were present [Figure 2c], the uterus appeared normal. A radiological possibility of metastatic breast carcinoma having peritoneal carcinomatosis and ovarian metastases was made.

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A biopsy from the left breast mass revealed multiple small round cells with “starry sky” macrophages. Immunohistochemistry results were positive for CD-20, CD-45, Ki67; negative for CD 34; and fluorescence *in situ* hybridization analysis revealed translocation involving *c-myc*, confirming a diagnosis of Burkitt lymphoma. She was given 4 cycles of CODOX-M/IVAC chemotherapy to which she responded. There was regression of the breast lesion and symptomatic improvement.

DISCUSSION

Lymphoma can involve almost any organs of the body; though ‘peritoneal lymphomatosis’, implying multiple intra-abdominal organ involvement or peritoneal dissemination; is rare.^[2-4] Peritoneal lymphomatosis can be seen either in association with a primary visceral site of involvement, or without any visceral involvement (also called body cavity lymphoma or primary effusion lymphoma). Most cases of peritoneal lymphomatosis have a disseminated lymphoma; primary intra-abdominal site of involvement being the gastrointestinal tract.^[5] Malignant ascites in lymphoma has been reported to have a variable incidence (9% in some series).^[1] Primary effusion lymphoma is seen almost exclusively in HIV-associated lymphoma.^[6]

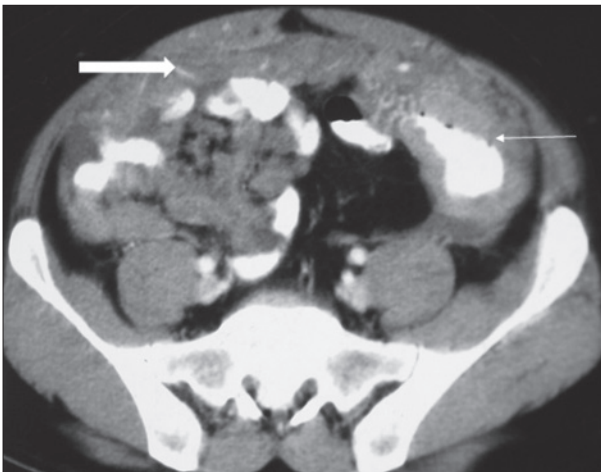


Figure 1: Axial CECT abdomen reveals aneurysmal dilatation and wall thickening of a small bowel loop (arrow) associated with gross omental thickening (block arrow)

Diffuse large B cell lymphoma, is the commonest histologic type of NHL seen in adults and a large proportion of patients present with extranodal involvement.^[7] Burkitt lymphoma is a highly aggressive form of NHL first described by Dennis Burkitt in 1958.^[8] It often presents as an extranodal lymphoma or as an acute leukemia. Previously they were classified as small noncleaved cell NHL (in patients presenting with solid tumor or nodal disease). In the presence of more than 25% bone marrow involvement, it was classified as French-American-British L3 ALL. Because of the shared molecular features, both of the entities are recognized as a single pathology of mature B cell neoplasm by World Health Organization classification of lymphoid diseases,^[9] which can present as Burkitt cell lymphoma or Burkitt cell leukemia. Peritoneal dissemination is an extremely uncommon manifestation on both the histologic subtypes.

Clinical presentation of diffuse peritoneal lymphomatosis is nonspecific and often detected on imaging. Cases are reported of peritoneal lymphomatosis associated with increased CA-125 titer, mimicking epithelial carcinoma of ovary.^[10] Ascitic fluid adenosine deaminase levels can also be high in cases of lymphoma, making the differentiation from a tubercular peritonitis difficult.^[11] Our first patient had alteration in bowel habits suggesting bowel involvement, whereas the presentation of the second patient was with menorrhagia and breast mass, which is even more unusual. CT is often the initial and optimum modality of investigation, though the imaging features on CT are also nonspecific. Close imaging mimickers are peritoneal carcinomatosis, tubercular peritonitis, mesothelioma, infiltrating fibromatosis, and round cell desmoplastic tumor of the mesentery.^[3,12] There is no specific imaging pointer for differentiating peritoneal lymphomatosis from carcinomatosis; as omental caking and lymphadenopathy can be observed in both. Visceral involvement in lymphoma on imaging is the most important pointer for lymphomatosis. Some of the imaging features including aneurysmal dilatation of the bowel loop with wall thickening, lymphadenopathy, hepatic and splenic enlargement with or macroscopic involvement can help the diagnosis of lymphoma and exclude other differentials, but most often, histology is the mainstay of diagnosis.

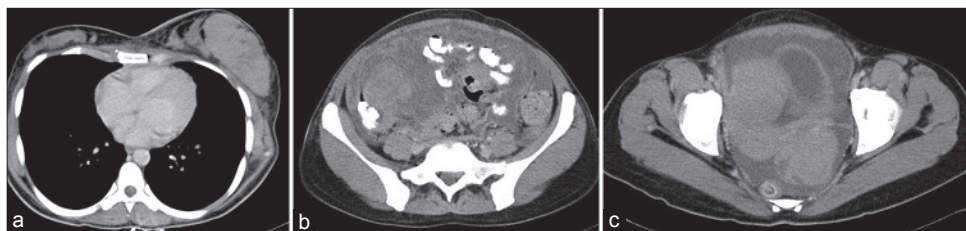


Figure 2: (a) CECT thorax soft tissue window axial section reveals a large soft tissue mass in the left breast. (b) CECT of abdomen axial section soft tissue window showing ascites and diffuse omental thickening. (c) CECT of pelvis axial section reveals ascites and bilateral solid ovarian masses; the uterus appears normal

To conclude, peritoneal lymphomatosis is an extremely uncommon presentation of NHL, which needs to be differentiated from the commoner causes like peritoneal carcinomatosis and tuberculosis.

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