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PRIMARY ILEO-CECAL LYMPHOMA WITH ILEO-CECAL INTUSSUSCEPTION AND BILATERAL RENAL INVOLVEMENT: A CASE REPORT

Radio-Diagnosis

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ABSTRACT

Extra nodal lymphomas commonly involve the gastro-intestinal tract but primary gastro-intestinal lymphomas are rare, majority originating from B cells. Diffuse large B cell lymphoma, a type of NHL, predisposes the gastro-intestinal tract to develop intussusception as a complication. Renal involvement in DLBCL is unusual. Since the signs and symptoms of primary GI lymphomas are non-specific, PET-CT scan is gaining popularity in baseline staging of the disease and prognosis. The final diagnosis is achieved by histopathology of the tissue sample. RCHOP is considered a better treatment modality in patients with disseminated disease while surgical resection is preferred if complications like intussusception are encountered. In our patient, non specific signs and symptoms along with aneurysmal dilatation in the ileal region with multiple solid lesions in both the kidneys warranted a PET-CT. The PET scan was suggestive of a neoplastic process. Colonoscopy with biopsy of the lesion along with CT guided left kidney biopsy was taken. Diagnosis was confirmed on histopathology and immuno-histochemistry. Treatment in the form of RCHOP chemotherapy regimen was given to the patient.

KEYWORDS

Primary ileo-cecal lymphoma, Non Hodgkin lymphoma, Diffuse large B-cell lymphoma, Intussusception, Bilateral renal involvement,

INTRODUCTION

Lymphomas are a heterogenous group of malignancies that emerge from lymphocytes and present as enlarged non tender lymph nodes but also have extra-nodal presentations[1]. They are of two types- Hodgkin's and Non Hodgkin's lymphoma. Non Hodgkin's lymphomas are a group of lymphoproliferative disorders that arise from the B-, T-, or natural killer T-cells. The gastro-intestinal tract is the most common site for extra-nodal lymphoma involvement however, primary GI lymphoma is a rarer entity and accounts for only 1-4% of all GI malignancies[2]. It also constitutes 10-15% of all NHL and approximately 30-40% of all extra-nodal lymphomas. Studies have shown that stomach is the commonly affected part followed by the ileum of the small intestine(60—65%). Histopathologically, majority of these are of B cell lineage and only 8-10% are T cell lymphomas[3]. Certain risk factors have been found to be associated with GI lymphomas such as- infections with *Helicobacter pylori*, *Campylobacter jejuni*, human immunodeficiency virus (HIV) , Epstein- Barr virus (EBV) , Hepatitis B virus, Human T-cell lymphotropic virus type 1 (HTLV-1) as well as certain inflammatory conditions like celiac disease, inflammatory bowel disease, atrophic gastritis and parasitic infection[4]. In this case report we will be discussing primary GI lymphoma presenting with ileo-cecal intussusception with bilateral renal involvement.

CASE REPORT

A 64 year old female presented to an urban tertiary care centre with complaints of vague abdominal pain which was continuous in nature and poorly localized, since 5-6 months. The patient had no co-morbidities and no significant family history. It was not associated with fever, weight loss, bladder or bowel changes. No significant finding other than generalized abdominal tenderness was found. The laboratory investigations were within normal limits. The patient underwent an ultrasound of the abdomen and pelvis on OPD basis which showed bowel thickening and aneurysmal dilatation in the ileo-cecal part of the bowel. Other findings noted were solid lesions showing mixed echogenicity in the superior and inferior poles of the right and left kidneys.

This was followed by whole body PET-CT which revealed concentric and eccentric thickening with accentuated enhancement of walls of at least 4cm-5cm of the terminal ileum extending up to the ileo-cecal junction and cecum with aneurysmal dilatation of the cecum (ileo-cecal intussusception), latter showing metallic radio-opaque foreign body within high retention index. This was suggestive of a neoplastic process, most likely lymphoma. Metabolically active focal lesions were found in both the kidneys, more so on the left. There was no evidence of metabolically active lymph nodes, lung

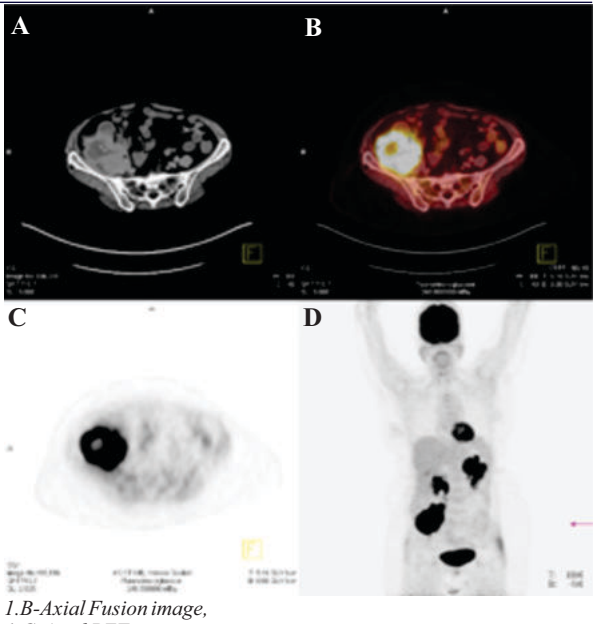

Figure-1*.A- Axial CT image*
1.B-Axial Fusion image,
1.C-Axial PET image
1. D-Coronal PET image

Showing heterogeneously enhancing mass lesion in the ileo-cecal junction with concentric and eccentric thickened walls causing luminal compromise without acute or sub-acute obstruction.

parenchymal/pleural/peritoneal/omental lesions, ascites or pleural effusion. The patient then underwent a colonoscopy which showed an edematous, nodular, ulcerated mucosa with ileal prolapse from the ileo-cecal junction. Multiple biopsies were taken.

Biopsy from the left kidney was taken as well.

Histopathological examination of the biopsy from the ileo-cecal junction described a high grade non-Hodgkin lymphoma of diffuse large B cell type with a 'GCB like' phenotype by Hans algorithm. On immunohistochemistry, the tumour cells expressed CD20, CD10, BCL-6 , MUM-1 and BCL-2. They were immunonegative for CD3, CD5, C-myc, Tdt, CD34, cyclin D1 and SOX-11. The Mib-1 proliferation index was >95%. The trucut biopsy from the left kidney showed a similar histopathological and immunohistochemistry picture.

The patient was treated with 6 cycles of R-CHOP regimen of chemotherapy. Follow up PET-CT scans showed a decrease in metabolic activity in the terminal ileum and cecum and resolution of the

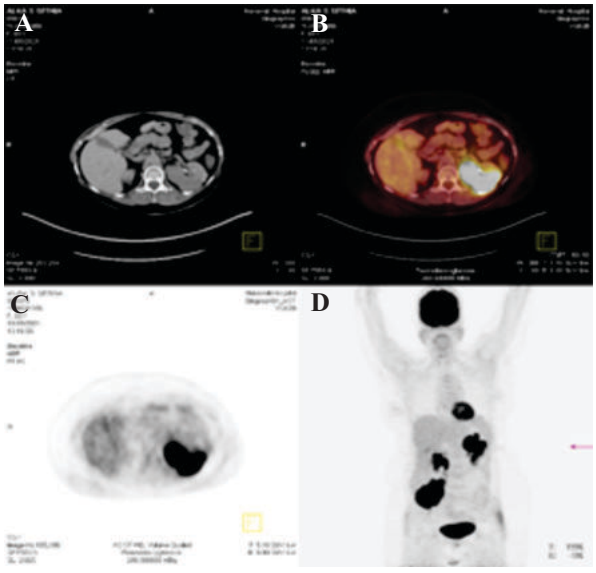

Figure 2*.A- Axial CT image,*
2.B- Axial Fusion image
2.C-Axial PET image
2.D-Coronal PET image

Showing solid heterogeneously enhancing mass lesion in superior pole cortex of left kidney without calcification, not involving the pelvicalyceal system or inferior vena cava.

metabolically active lesions of the right and left kidney with focal cortical scarring. She went on to develop sub-acute intestinal obstruction one and a half months after the last chemotherapy cycle for which a right radical hemicolectomy was performed with ileo-transverse anastomosis. Patient was followed up for one year post surgery with surveillance PET scans.

DISCUSSION

The WHO classification of B cell lineage histological sub types of lymphomas includes-extra nodal marginal lymphoma, follicular lymphoma, mantle cell lymphoma, diffuse large B cell lymphoma and Burkitt's lymphoma[5].

DLBCL is known to originate from the germinal centre or post germinal centre B cells, majority expressing the B-cell lymphoma gene(bcl-6) found to be associated with DLBCL

Dawson's criteria used to describe primary GI lymphoma requires the absence of peripheral lymphadenopathy during presentation, no mediastinal lymph node involvement, total and differential WBC count to be within normal limits, predominant bowel lesion during laparotomy with only a single lymph node being affected in immediate proximity and no lymphomatous spleen or liver involvement[5].

Primary GI lymphomas are commonly seen in adults older than 55 years with a male predominance[6]. It usually presents with non-specific signs and symptoms like abdominal pain, decreased appetite, weight loss, paralytic ileus, diarrhoea, palpable mass and bleeding from the GI tract[3]. According to a study conducted by Suresh B, et al. in Bengaluru, India, dull aching abdominal pain and discomfort were the common symptoms experienced by the patients of gastro-intestinal lymphoma, similar to our case[7]. Intussusception is a rare cause (<5%) of adult intestinal obstruction and 52-55% occur in the small intestine[8]. DLBCL of the GI tract can predispose patients to develop intussusception which may present as colicky abdominal pain, nausea, vomiting or even ulceration and perforation[3]. About 14% of these are associated with malignant lesions[8]. A case similar to ours was reported by Wagh A, et al. which describes a 50 year old male presenting with ileo-colic intussusception due to DLBCL[9].

Renal parenchymal involvement is rare in DLBCL and is mainly secondary to direct extension from a retroperitoneal mass or due to a disseminated disease leading to hematogenous spread[10]. A study conducted by Nuguri S, et al. included 12 patients who presented with AKI and the kidney biopsies showed interstitial lymphomatous infiltration. 10 out of these 12 patients were diagnosed as secondary lymphomas of the kidney. Of these, 6 were DLBCL[11].

Since the clinical presentation of GI-DLBCL can be non-specific, imaging techniques hold significant importance in diagnosis as well as in picking up associated complications like bowel perforation, intussusception and fistulas[4]. Typical imaging features for intussusception are target sign, doughnut sign in transverse view with sensitivity being 98-100% and specificity being 88%, however CT is still considered gold standard in such cases[9]. ^{18}FDG-PET is attaining increasing importance in baseline staging of lymphoma and provides prognostic information with contribution in determining the best initial treatment modality. It is also useful in evaluation of treatment response as it can delineate residual viable malignant lymphomatous lesions with greater accuracy compared to other treatment modalities[12].

The most commonly encountered patterns of small bowel lymphoma on CT/MRI are polypoid, infiltrative, aneurysmal, exophytic and stenosing. The aneurysmal pattern (luminal dilatation more than 4cm) represents 31% of small bowel lymphomas. The patient described in this case report also showed aneurysmal dilatation of the cecum. Certain theories postulate that destruction of the myenteric plexus and muscle layers along with stretching of the muscle fibres leads to loss of contractility eventually causing aneurysmal dilatation.

Secondary renal lymphomas or deposits can be unilateral or bilateral or even present as a solitary mass, multiple focal masses or diffuse bilateral nephromegaly in the absence of any focal lesions[13]. The most common finding in renal lymphoma is multiple solid parenchymal masses (50-60%).

Radiology alone cannot be used for diagnosis of small intestinal lymphoma therefore warranting the need for pathological assessment of tissue sample. In our case this was done by taking tissue samples through multiple biopsies during colonoscopy as well as trucut biopsy of the left kidney under CT scan guidance to establish the diagnosis of DLBCL.

Surgical resection of the segment suspected to have NHL of the small intestine is the treatment modality of choice especially if the patient has presented with the underlying complication of intussusception. This is also supported by the fact that 14% of all adult intussusceptions were associated with malignancy[8]. It has also been found by a retrospective cohort study conducted by Kim JS, et al. using clinical data of 345 patients with primary intestinal DLBCL that chemotherapy with surgery has the advantage of improved 3 year survival rate and lower relapse rate compared to surgery alone in localised DLBCL. It was also found by the study that R-CHOP is a better treatment modality for disseminated disease whereas R-CHOP showed no statistically significant difference from CHOP therapy alone for localised disease[14].This was supported by a study conducted by Hwang SH, et al[15].

CONCLUSION

Primary small intestinal lymphomas with non-specific signs and symptoms make clinical diagnosis challenging. Usually, multiple differential diagnoses are considered along with small intestinal lymphoma. This is usually aided by radiological findings and confirmed with histopathological and immunohistochemical assessment especially in cases of disseminated disease such as in our case where multiple lymphomatous renal deposits were found bilaterally without any symptoms or signs involving the genitourinary system.

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