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Jejunal Perforation: A Rare Presentation of Extra Nodal Follicular Lymphoma in Patientwith Hepatitis B

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Abstract: *The non-traumatic perforations of the small intestine are pathological entities.The primary sources of the perforation might be foreign bodies, inflammatory diseases, tumors, infectious diseases, etc. In most cases intestinal perforation is discovered only by laparotomy and the definitive diagnosis is made only after histopathologic examination. Small bowel malignancies are rare; among them, lymphomas rank third in frequency, being mostly B-cell non Hodgkin lymphomas.Primary non-Hodgkin's lymphoma of the small bowel with hepatitis B presenting as a perforated viscus entity with peritonitis is extremely rare.*

A 58 year old man presented with pain abdomen and distension. An emergency laparotomy revealed a perforation in jejunum with enlarged mesenteric lymph nodes and mesenteric thickening which required segmental resection and anastomosis. Histopathological examination of the resected segment of jejunum revealed extranodal follicular lymphoma of small intestine. The nonspecific clinical manifestations of intestinal lymphomas make from diagnosis a difficult procedure. Lymphoma of the small intestine has been reported to have a poor prognosis. Early diagnosis and treatment are important to improve the prognosis of bowel perforation in patients with non-Hodgkin's lymphoma.

This case illustrates a rare complication of follicular lymphoma of (large B-cell non-Hodgkin's lymphoma) the jejunum associated with hepatitis B that was responsible for small-bowel perforation and following peritonitis.

Keywords: *perforation, small intestine, hepatitis B, extra nodal follicular lymphoma, peritonitis*

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I. Introduction

The nontraumatic perforation of the smallintestine is a rare pathological entity, which hasparticular aspects in respect to the clinical diagnosis and subsequent therapeutic conduct. The primary sources of the perforation includeforeign bodies, inflammatory diseases, tumors,infectious diseases, etc. Despite the fact that the small bowel represents 75% of the length and over 90% of the mucosal surface of the intestinal tract, malignant tumors of the small bowel account for less than 1% of intestinal malignances and primary lymphomas of the small intestine are rare[1,2].The gastrointestinal tract is the most common organ for primary extra nodal non-Hodgkin lymphoma (NHL) with most cases occurring as single lesions in the stomach and less commonly in the intestine[3]. Ileum and ileocecum are the most frequent locations for primary intestinal lymphomas; with diffuse large B-cell lymphoma (DLBCL) as the most common histological type and representing approximately one third of all cases[4].

Several epidemiological studies have demonstrated a positive association betweenpersistent, hepatitis B surface antigen (HBsAg)-positive hepatitis B virus (HBV) infection and B-cell non-Hodgkin lymphoma (NHL), with HBV-infected patients having a 2-3-fold higher risk to develop NHL than non-infected patients[5].

II. Case Presentation

A 58 year old man presented with colicky pain abdomen since last 1 month which aggravated over last three days associated with nausea and vomiting. There is abdominal distension and not passing stool and flatus since last 2 days. Patient is known case of Diabetes mellitus and on Insulin.During this disease course,patientdiagnosed as Hepatitis-B positive. The patient's vital signs on admission were as follows: temperature 39.2°C, heart rate 107 bpm, respiratory rate 18 breaths/minute, blood pressure 104/73 mm Hg and oxygen saturations of 97% on room air. On physical examination,tense, distended,generalised tenderness and umbilicus stretched with tympanic abdomen and no palpable lump.Loss of liver dullness and sluggish bowel sounds.

Laboratory investigations demonstrated Total leukocytecount-11×109/L, haemoglobin-10.2 g/dl, Blood sugar(Random)-205mg/dl, blood urea-16 mg/dl, serum creatinine-1mg/dl, Liver Function Test -within normal limits, HBsAg positive. An erect chest X-ray demonstrated free air underthe diaphragm. An urgent CECT scan was performed and this confirmed intestinal perforation. Emergency laparotomy performed and a 6 mm perforation found in jejunum with mesenteric thickening and enlarged mesenteric lymph nodes. Segmental resection and end to end anastomosis of intestine done. Specimen of resected jejunal segment sent for histopathologic examination.Post operative period was uneventful and patient discharged under satisfactory conditions. Patient followed up regularly.

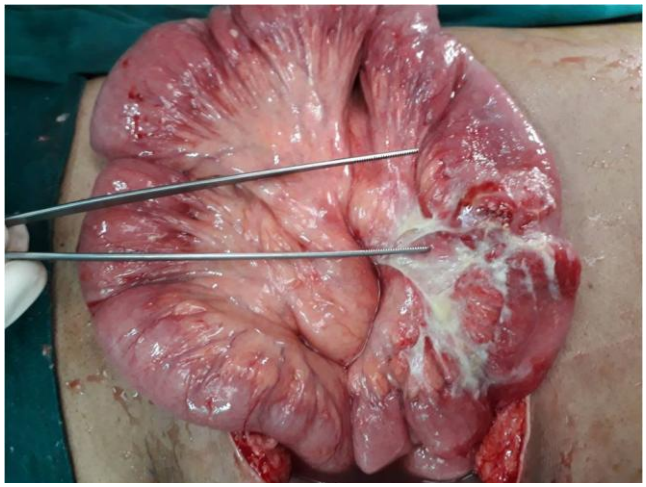

Fig 1Intra-operative view of specimen, perforation in jejunum

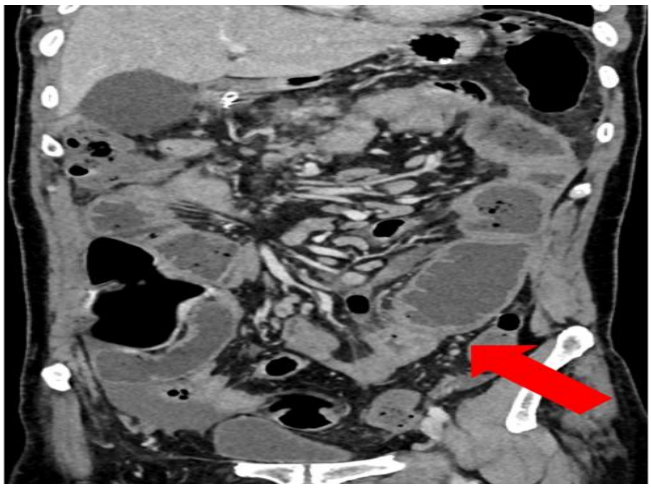

Fig 2 CECT abdomen showing small bowel thickening

Histopathological Examination

Histopathological examination revealed Sheets of tumour cells identified which were present in sub mucosal, muscular layer and reaching up to the serosa. However, mucosa was free of tumour infiltration.The tumour cells were large having high N/C ratio, prominent to inconspicuous nucleoli, moderate to scant amount of eosinophilic cytoplasm and indistinct cell boundaries. Low power photomicrograph shows Sheets of tumour cells which were present in submucosal, muscular layer and reaching up to the serosa.

Immunohistochemical examination revealed leukocyte common antigen(LCA) positive, CKIT negative, CD30 negative, CD79a(B cell) positive,BCL6 and BCL2 positive

Histopathological aspect and immunohistochemically tests were compatible with the diagnosis ofextra nodal follicular lymphoma of small intestine**.**

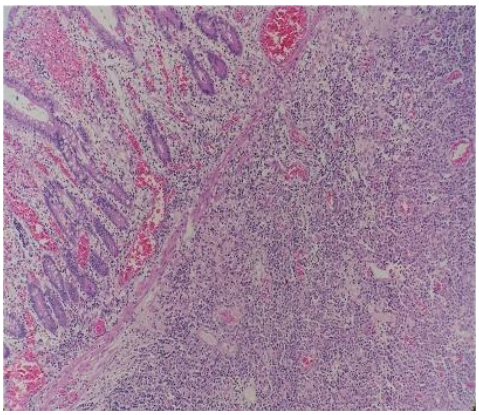

Fig 3-10 x submucosa showing tumor infiltration

III. Discusion

The primary lymphomas of digestive tract remain rare entities, even though in the last decades a slight increase of their incidence (with percentage between 3 and 5) is reported. It is possible because of the improvement of the immunohistochemical diagnosis, which facilitates their diagnosis[6, 7]. Almost 90% of the gastrointestinal lymphomas derive from B lymphocytes and very few from T lymphocytes[8]. Gastrointestinal lymphoma can be classified into three main categories: (1) immunoproliferative small intestinal disease (IPSID), (2) enteropathy-associated T cell lymphoma (EATL) which arises mostly in gluten-sensitive enteropathy and (3)diffuse large B cell lymphoma, mantle cell lymphoma, Burkitt's lymphoma and follicular lymphoma[9].

Risk factors like Celiac disease, Helicobacter pylori infection, Campylobacter jejuni, virus infection like HIV, EBV, HBV and HTLV-1, inflammatory bowel disease, Wegener's granulomatosis, rheumatoid arthritis, Wiskott-Aldrich syndrome and immunosuppression, have been found to be taking part in the pathogenesis of gastrointestinal lymphoma[10, 11]. Primary GI tract lymphoma is defined as a tumor that predominantly involves the GI tract with lymph node involvement confined to the drainage area of the primary tumor site. There is no liver or spleen and chest involvement and palpable lymph nodes are not present. Peripheral white blood cells are normal[12]. Primary lymphoma of GI tract accounts for only 0.9% of all GI tract tumors. Differentiation between primary GI lymphoma and systemic lymphoma with GI tract involvement has important implications because the prognosis is better in the primary form when diagnosed early on, with five-year survival rates as high as 62 to 90%[12]. GI tract lymphoma, whether primary or systemic, most often involves the stomach (50%), followed by the small intestine (33%), colon (10–16%), and esophagus (1%). NHL of the GI tract is more common in men than in women (3:2)[12]. There is a double peak, with the first peak in patients less than 10 years of age and the second at a mean age of 53[12].

Symptoms will depend on the site of involvement, but may include dysphasia, abdominal pain, nausea, vomiting, anorexia, diarrhea, GI bleeding malabsorption, and diarrhea, all of which are nonspecific. B symptoms (fever, weight loss and night sweats) are also nonspecific which occurring in the context of a lymphoma get a prognosis value[12]. There may be a palpable mass[12]. Uncommonly, there may be small-bowel obstruction. Treatment modalities for GI tract lymphomas include chemotherapy, radiation therapy, stem cell transplant, and antibiotic treatment for H. pylori[12]. Considering the standard treatment protocol, segmental resection of involved intestine and adjacent mesentery should be the treatment for localized small-intestinal lymphoma, as in our case. If the small intestine is diffusely affected by lymphoma, chemotherapy rather than surgical resection should be the primary treatment[13]. However, the value of adjuvant chemotherapy after resection of localized lymphoma is controversial[13].

As early as in 1970s, the detection of hepatitis B surface antigen (HBsAg) in the hepatocytes of some patients with lymphoproliferative disorders was reported and first suggested this association[14, 15]. Data from two hospital-based case-control studies in South Korea are among the studies supporting the possibility that chronic HBV infection increases NHL risk[16, 17]. A most recent meta-analysis, in which over 40,000 cases of NHL and 1,660,000 cases of control were included, showed that HBV-infected individuals had an odd ratio of 2.24 (95% confidence interval 1.80–2.78; $P \leq 0.001$) of developing NHL.[18]

Some authors found in their studies that HBVinfection was associated with a significantlyearlier disease (NHL) onset. Thus, Kim et al[16]found that the HBsAg positive rate was consistentlyhigher for NHL patients in every agegroup, but the risk of NHL was most evident inthe younger HBsAg positive groups.

IV. Conclusion

Our case illustrates a rarely reported complication of Extra Nodal Follicular Lymphoma (large B-cell non-Hodgkin's lymphoma) of the jejunum that may be responsible for small-bowel perforation and following peritonitis. This clinical pattern may appear as a leading presentation of small-intestinal neoplastic disease in which surgical resection needs to be considered. Morbidity and mortality increases significantly in primary gastro intestinal lymphomas presenting as perforation, but with proper understanding of the disease and timely intervention, management will yield good results as we saw in our patient.

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