Uncommon Diagnosis of Common Presentation: Diffuse Large B-cell Lymphoma of Ileocaecal Region

Upama Sharma, MBBS; Jagdish L Baidya, FRCS, FCPS
Department of General Surgery, B&B Hospital, Gwarko, Lalitpur, Nepal

Address for Correspondence:
Upama Sharma, MBBS
Department of General Surgery, B&B Hospital, Gwarko, Lalitpur, Nepal
Email:upama43@gmail.com

Received, 12 May, 2017
Accepted, 21 May, 2017

Extranodal lymphoma occurs in about 40% of all patients with lymphoma and has been described in virtually all organs and tissue. However, diffuse large B-cell lymphoma (DLBCL), which is the most common histological subtype of non-Hodgkin's lymphoma (NHL), primarily arising in the ileocaecal region has been rarely reported. In this article, we report a rare case of young male, who virtually was misdiagnosed as the symptoms were unspecific and histopathology later confirmed the diagnosis. Unfortunately, due to aggressive nature of the disease and the operation itself, the patient couldn't survive.

Keywords: appendicitis, diffuse large B-cell lymphoma, gastrointestinal lymphoma, Intestinal perforation.

Gastrointestinal tract is the most common extranodal site involved by lymphoma accounting for 5-20% of all cases.[1]Extranodal disease is more common with non-Hodgkin's lymphoma (NHL) and diffuse large B-cell lymphoma (DLBCL) is the most common histological NHL subtype in adults, accounting for about 25% of all NHL cases. Thus, it is known that gastrointestinal DLBCL is the most frequent form of extranodal lymphoma.[2]

The primary sites of origin in decreasing order of frequency include the Stomach (50– 70%), Small bowel (20–35%), Colon (especially the cecum) (5–10%) and the Esophagus (<1%). The most common site of involvement are the ileocaecal region and caecum, probably because of more lymphoid tissue present normally in the ileocaecal region than any other part of the colon.[3,4]

Case Report

A 22-year-old gentleman presented with a history of vague pain abdomen mostly at umbilical and right iliac region, which had increased in severity since 2-3 days prior to presentation associated with low grade evening rise of temperature, loss of appetite and vomiting. He also had history of significant but unquantified weight loss.

He was a regular IV drug abuser, smoker and consumed alcohol.

On arrival in Emergency department, he was found dehydrated, anxious but vitals were normal. There were no enlarged or palpable lymph nodes. Abdomen was soft with tenderness over the right iliac fossa and hypogastric region.

The peripheral blood count was unremarkable (Hemoglobin 11.2 g/dl, white blood cell count 11,500/mm^3, and platelet count $3.79\times10^5/mm^3$). The peripheral blood smear revealed no immature cells (89% neutrophils, 11% lymphocytes). Liver and renal functional tests were normal.

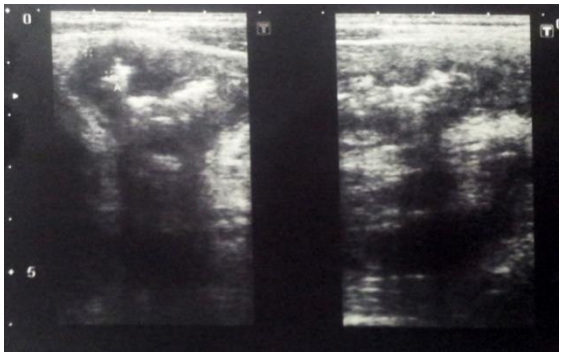

Figure 1: Ultrasonography (USG) of abdomen showing thickening of caecum

Ultrasonography of abdomen showed long segment bowel wall thickening in RIF measuring 5.2 mm with mesenteric lymphadenopathy largest one measuring 21.1 mm in size. Appendix could not be visualized separately **(Figure 1)**.

Contrast-enhanced computed tomography (CECT) of chest and abdomen revealed patchy nodular right upper chest, hepatomegaly and pelvic ascites.

So, the patient underwent diagnostic peritoneoscopy. Intraoperatively, appendix was found inflamed, edematous, about 6cm in length, retrocaecal, with purulent and thick peri-appendicular collection. Caecum and ileo-caecal junction bowel wall was edematous and was covered with omentum. There were also multiple large mesenteric lymph nodes up to 1 cm in diameter along the mesentery of ileo-caecal region. Appendectomy was done and peritoneal fluid sent for PCR to rule out tuberculosis and later found negative.

Patient did well and was discharged on 2nd day. Histopathology reported as acute suppurative appendicitis with no evidence of tuberculosis or neoplasia.

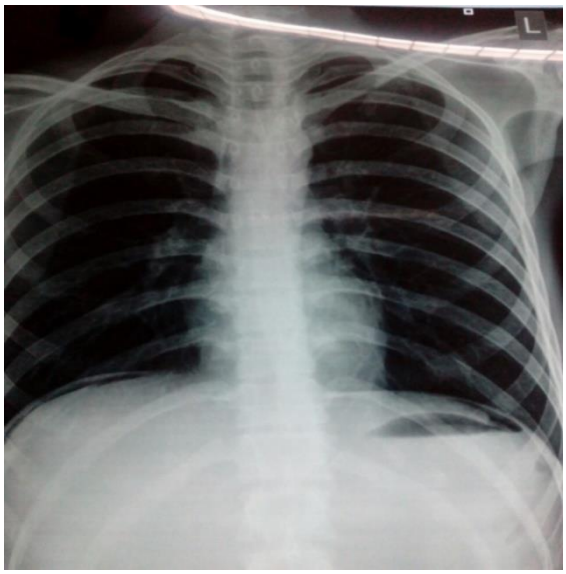

Figure 2: Chest X-ray showing free gas under both domes of diaphragm

However, one month later patient presented at other centre with fever and burning abdominal pain over the epigastrium. Upper GI endoscopy showed fleshy mucosal tag at 32 cm with surrounding scar. Biopsy was taken which revealed mild dysplasia with esophagitis. All routine laboratory investigations were inconclusive, hemoglobin was 9.8gm/dl with total WBC count raised up to 13,100/mm^3. Empirical treatment for tuberculosis was started in view of clinical suspicion of abdominal tuberculosis and discharged.

He again presented to emergency department of the same hospital after five days with the complaints of loose motion for 1 day. Chest X-ray done then showed gas under the diaphragm **(Figure 2)**.

Ultrasonography done showed collection in the deep pelvic cavity measuring 5.9x5.8x5.8cm, loculated in right lumbar region with moderate ascites and air noted in the collection. Reactive lymph node present **(Figure 3)**. Patient was then referred to our center.

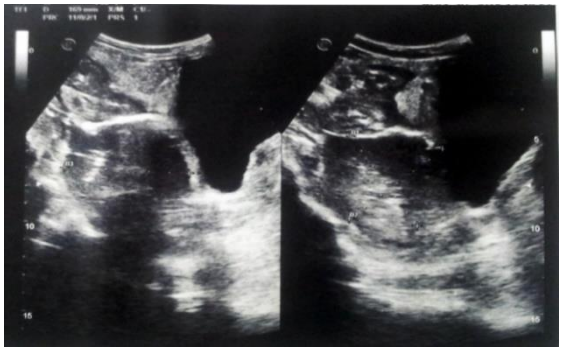

Figure 3: USG showing loculated collection in pelvis showing thickening of caecum

On arrival at the ER department, patient had generalized abdominal pain associated with loose stool and fever with chills and rigors.

Clinically, patient was ill-looking, anxious, pale, dehydrated and tachycardic, but no lymphadenopathy. There was mild tenderness over the RIF, rebound tenderness absent. On per rectal examination, rectum was empty with tender bogginess over the rectovesical pouch.

So, with high suspicion of pelvic abscess probably due to Koch abdomen and post laparoscopic appendectomy status patient was taken to operating theatre with the plan for laparoscopic drainage of pelvic abscess.

Intraoperative findings are listed below:

- About 300ml purulent bilious fluid in the pelvis with adhesions between the small bowel and abdominal wall.
- Multiple perforations in distal segment of ileum covered with omentum. All of the perforations were positioned at the anti-mesenteric border with circular, healthy margin, largest one measuring about 2x4cm **(Figure 4)**.
- Another large perforation at the base of the caecum measuring 3 cm in diameter.
- Total 11 perforation sites identified.

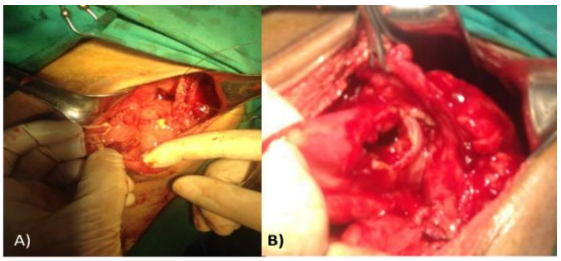

Figure 4: Intraoperative picture showing A) perforation in ileum, B) large caecal perforation

Diagnostic peritoneoscopy was converted to Laparotomy and primary repair of all the perforations were done with diversion loop ileostomy about 25 cm proximal to the ileo-caecal junction. Biopsy from the perforated bowel wall mucosa was taken.

Histopathological report revealed diffuse Non-Hodgkin's Lymphoma and immunohistochemistry was sent to differentiate the type. Anti-tubercular drugs stopped and patient was planned on starting chemotherapy once the wound healed.

Postoperatively, patient developed pancytopenia with hemoglobin 7.6gm/dl, WBC total count 1800/mm^3 and platelets 95,000/mm^3 which could not be corrected despite of multiple transfusion of blood product. He ultimately developed

disseminated intravascular coagulopathy with septic shock, started bleeding from ileostomy site and died.

Discussion

Primary malignant tumors of the small intestine are very rare, accounting for less than 2% of all gastrointestinal malignancies. Lymphoma constitutes 15-20% of all small intestine neoplasms and 20-30% of all primary gastrointestinal lymphomas. Ileum is the most common site (60-65%) involving small intestine lymphoma followed by jejunum (20-25%), duodenum (6-8%) and other sites (8-9%).(1)

Dawson's criteria are used for labeling primary gastrointestinal lymphoma which are true in our case too. This include (1) absence of peripheral lymphadenopathy at the time of presentation; (2) lack of enlarged mediastinal lymph nodes; (3) normal total and differential white blood cell count; (4) predominance of bowel lesion at the time of laparotomy with only lymph nodes obviously affected in the immediate vicinity; and (5) no lymphomatous involvement of liver and spleen.[1]

The age of presentation varies with the histological subtype of lymphoma. The clinical presentation of small intestinal lymphoma is non-specific and the patients have symptoms, such as colicky abdominal pain, nausea, vomiting, weight loss and rarely acute obstructive symptoms, intussusceptions, perforation or diarrhea.[1]For most gastrointestinal tract lymphomas, no specific association with a preexisting disease or pathological lesion have been reported.[5] However, in our case, patient presented with features of acute appendicitis. Probably the reason was due to obstruction in the lumen of appendix causing the inflammation. However, primary appendiceal lymphomas presenting as acute appendicitis are very rare: they occur in only 0.015% of all gastrointestinal lymphoma cases.[6]So, histopathology report of appendix was acute suppurative appendicitis without evidence of neoplasia.

The distal ileum is classically thought to be the most common site of small bowel B-cell lymphoma because of the greater amount of lymphoid tissue in this portion of the bowel.[7] There were multiple large mesenteric lymph nodes up to 1 cm in diameter along the ileo-caecal region during the appendectomy. Appendicitis is frequently associated with lymphadenopathy, most commonly in the mesentery of the right lower quadrant.[8] So, this mislead us and we thought it to be a straight forward case of appendicitis only.

The tumor may involve a relatively long segment of bowel and may ulcerate and perforate into the adjacent mesentery, resulting in the formation of a confined, usually sterile abscess.[7]

The lack of specific complaints probably accounts for the delay in the diagnosis.

The treatment outcome of intestinal lymphoma is relatively poorer than that of gastric lymphoma depending on their histologic subtypes. Lymphoma primarily located in the small intestine usually warrants laparotomy with the affected segment removed both for its diagnosis and for its treatment.[1]

Combined modality of approach that includes surgical debulking and systemic chemotherapy is the preferred treatment. Currently, the procedure of choice for

intestinal DLBCL is widely considered to be a combination of surgery followed by cyclophosphamide, doxorubicin, vincristine, and prednisolone (CHOP) or rituximab plus CHOP (R-CHOP) chemotherapy, primarily because the preoperative diagnosis is difficult and risk of complications requiring surgery is relatively high during chemotherapy.[9] Surgery alone can be considered as an adequate treatment for patients with low-grade NHL disease that does not infiltrate beyond the sub mucosa.

Conclusion

Although gastrointestinal lymphomas may be common, it may be very difficult to diagnose preoperatively. Many-a-times, it mimics other abdominal pathologies like acute appendicitis in our case. Abdominal tuberculosis, which is very common in our part of the world, also has similar features thus misleading and delaying the right treatment.

References

1. Ghimire P, Wu G-Y, Zhu L. Primary gastrointestinal lymphoma. World J Gastroenterol 2011;17:697-707.
2. Cai YL, Xiong XZ, Lu J, Lin YX, Cheng NS. Non-Hodgkin's lymphoma with uncommon clinical manifestations: A case report. Oncology letters 2015;10:1686-8.
3. Doolabh N, Anthony T, Simmang C, Bieligk S, Lee E, Huber P et al. Primary colonic lymphoma. Journal of surgical oncology 2000;74:257-62.
4. Gonzalez QH, Heslin MJ, Dávila-Cervantes A, Alvarez-Tostado J, de Los Monteros AE, Shore G et al. Primary colonic lymphoma. The American Surgeon 2008;74:214-6.
5. Lewis JD, Bilker WB, Brensinger C, Deren JJ, Vaughn DJ, Strom BL. Inflammatory bowel disease is not associated with an increased risk of lymphoma. Gastroenterology 2001;121:1080-7.
6. Guo J, Wu G, Chen X, Li X. Primary appendiceal lymphoma presenting as suspected perforated acute appendicitis: clinical, sonography and CT findings with pathologic correlation. International journal of clinical and experimental pathology 2014;7:7068.
7. Levine M, Rubesin S, Pantongrag-Brown L, Buck J, Herlinger H. Non-Hodgkin's lymphoma of the gastrointestinal tract: radiographic findings. AJR American journal of roentgenology 1997;168:165-72.
8. Lucey BC, Stuhlfaut JW, Soto JA. Mesenteric Lymph Nodes Seen at Imaging: Causes and Significance 1. Radiographics 2005;25:351-65.
9. Hwang HS, Yoon DH, Suh C, Park C-S, Huh J. Intestinal diffuse large B-cell lymphoma: an evaluation of different staging systems. Journal of Korean medical science 2014;29:53-60.