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Case Report

Primary Small Bowel Non-Hodgkin's Lymphoma - A Study of Clinical Features, Histopathology, Diagnosis and Management

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Abstract

Gastrointestinal lymphoma are lymphomas seen in the digestive tract including esophagus, stomach, small, large bowel and rectum and it account for 10-30% of GI tumors [1].

We are reporting a case of 58 years old female who presented with small bowel obstruction, revealing lymphoma.

Keywords: Intestinal Obstruction; Small Bowel; Lymphoma; Colon

Case Report

- The clinical presentation of small bowel lymphoma varies as the patient may complain of nausea, fatigue, vomiting with vague abdominal pain and/or unintentional weight loss [1].
- In rare conditions it might be complicated by intussusception, perforation or obstruction [1].
- CT scan can help in the diagnosis, but will need histopathology to confirm it, for further management [1].

Case Presentation

A 58-year-old Bahraini female, not a known case of any medical illness, presented to the accident and emergency department with generalized abdominal pain on 18 April 2023. The patient suffered from this abdominal pain for one year, however it increased to the extent she could not tolerate before two days of her presentation. The pain was radiating to the back with severity of ten out of ten. There was a minimal relief with paracetamol. It was associated with nausea, vomiting, bloating and anorexia. She complained of diarrhea for one week in duration followed by four days of constipation. for the last four days. Patient is passing flatus with no history of obstipation. The patient gave history of weight loss (11 kg in four months period). She also mentioned having fever with generalized body ache and malaise in the last two days. The patient's social and family history showed no significance.

It is worth to mention that the patient went for Gastro-colonoscopy in November 2022, in view to her symptoms, it revealed a large non-sliding hiatus hernia, lower esophageal ulcerations, an erosive gastritis, with no active bleeding. According to these laparoscopic findings, Colonoscopy was unremarkable. Omeprazole tablets were prescribed to the patient.

On physical examination, the patient was vitally stable. The blood pressure was 104/73 mmHg, heart rate was high 118 beats per minute, oxygen saturation of 98% on room air and normal body temperature of 37.3. The abdomen was soft with a moderate tenderness over the periumbilical area and there were no palpable masses. No organomegaly appreciated. No palpable inguinal lymph nodes.

Laboratory data showed normal WBC count of 6.8X10^9/L, Hemoglobin of 13.6 g/dL, Mean cell volume of 78.3 fL, Platelets of 249X10^9/L, Amylase was 40 U/L, random glucose was 4.6 mmol/L, Urea of 6.1 mmol/L, elevated creatinine of 74 micromol/L and elevated Lactate dehydrogenase of 283 U/L.

Liver enzymes were within normal range except for Alkaline phosphatase it was elevated 147 U/L.

Normal electrolytes panel, Sodium of 140 mmol/L, Potassium of 3.8 mmol/L, Chloride of 103 mmol/L and Bicarbonate of 22 mmol/L.

Tumor markers were all within normal range CA 15-3 was 18.7 U/mL, CA 19-9 was 31.8 U/mL, Carcinoembryonic antigen (CEA) was 0.9 microgram/L, except for CA 125 it was elevated at 213.5 U/mL. Urine test was normal.

A Gastro-colonoscopy was performed on 14 March 2023 which revealed similar findings to the previous scope which was mentioned earlier except for newly diagnosed internal hemorrhoids.

During this emergency visit, an Abdominal CT scan was performed. It showed multiple heterogenous mesenteric lesions. Some of these lesions are inseparable from the bowel loops, with the largest mesenteric heterogenous hypodense mass seen centrally at the pelvis inseparable from the jejunal loops measuring about 8.3X6X7.4 cm. Some of the lesions showed central reduced attenuation suggestive of necrosis. Moreover, the adjacent small bowel loops showed a narrowing of the lumen due to a diffuse wall thickening. Also, a diffuse peritoneal folds thickening was noted with abdomen/pelvic free fluid, fat stranding and mesenteric congestion. To add, there was multiple enlarged adjacent mesenteric lymph nodes with the largest measuring about 1 cm. A diffuse submucosal wall thickening and edema of the duodenum was noted suggestive of duodenitis. No abnormal thickening of the large bowel or caliber dilatation.

Histopathology examination with immune-histochemical analysis showed Diffuse Large B cell lymphoma (DLBCL) and Follicular large B cell lymphoma Grade 3B.

Histopathology report showed multiple findings. A specimen of the greater omentum was analyzed, it consists of omental fat measuring 35X7X3 cm. Serial sectioning revealed multiple conglomerate tan nodules measures in total 10X6X3.8 cm. The nodules are less than 1 cm away from the omental margin.

Another specimen of the proximal small bowel was taken. It consists of segment of the small bowel measuring totally 20 cm in length with an average diameter of 4 cm. On opening, the lumen is focally narrowed down by a multinodular tan lesion 2.5 cm in diameter with thickening of the wall up to 2 cm. The abnormal area measures 6 cm in length, and it is 5 cm away from the mucosal resection margin and 7 cm away from the other mucosal resection margin. It is also 2 cm away from the mesenteric resection margin. Serial sectioning of the mesenteric fat revealed multiple tan nodules, largest measuring 6.5X5X8.5 cm.

The omental nodules along with the small intestine mesenteric nodules showed lymphoid tissue with predominantly neoplastic expanded lymphoid follicles that are composed of mixture of small centrocytes and large centroblasts. These cells of these follicles are diffusely positive for CD20, CD10 and Bcl-6. They are also diffusely positive for Bcl-2 supporting the diagnosis of follicular lymphoma. Both CD21 and CD23 highlight the follicular dendritic cell meshwork in these follicles. The follicles showed a proliferation index ranging from 10-20%. Areas mentioned above are best classified as diffuse large B cell lymphoma (DLBCL) most likely arising from areas of follicular lymphoma grade 3B. These areas constitute more than 50% of the section studied.

Sections from the obstructing mass in the small intestine (jejunum) showed complete replacement of the muscular wall by neoplastic lymphoid follicles with associated extensive sclerosis of the

surrounding stroma and involvement of the overlying mucosa. The proximal resection margin of the jejunal segment showed lymphoid follicles with suspicious involvement of in situ follicular neoplasia.

After revealing the histopathology, further workup was done for the patient including enhanced Thorax/Cervical CT scan showing the following findings. Bilateral basal atelectatic changes seen in both lower lobes. No intrapulmonary nodule is identified on either side. Prominent mediastinal lymph nodes are seen, the largest in the superior mediastinum adjacent to the esophagus and measures almost 10 mm.

Sub centimeter axillary lymph nodes seen bilaterally. A lower cervical lymph node adjacent to the left thyroid gland was visualized which measures 11 mm.

Liver appears normal except for tiny hypodense lesion in the left lobe of the liver. Stable juxta diaphragmatic, mesenteric and retroperitoneal lymph nodes in the upper abdomen (Figure 1 and 2).

Figure 1: Coronal contrast enhanced CT Abdomen and Pelvis: showing a Heterogeneous hypodense mass seen centrally at the pelvis inseparable from the jejunal loops measuring about 8.3 x 6 x 7.4 cm.

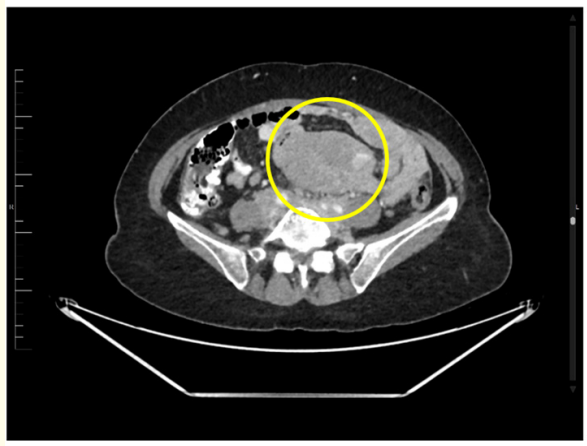

Figure 2: Axial contrast enhanced CT Abdomen and Pelvis: showing a Heterogeneous hypodense mass seen centrally at the pelvis inseparable from the jejunal loops measuring about 8.3 x 6 x 7.4 cm.

Initially, patient was kept for conservative management including keeping her NPO, Nasogastric tube insertion, Intravenous hydration maintenance with replacement of NGT 1:1 with Normal saline for 24 hours, when the decision was made to proceed with surgical intervention. Exploratory laparotomy was performed with small bowel resection and anastomosis with loop colostomy along with biopsy of mesenteric lymph nodes under general anesthesia.

Intraoperative findings included the first mass in the transverse colon omentum, not attached to the colon, large in size measuring 20X15 cm irregular in shape. The second mass was 50 cm away from the DJ attached to the jejunum measuring 20X15 cm extending to the mesentery. Finally, the third mass was 10X10 cm irregular in shape seen in the rectosigmoid region. There was no collapsed bowel seen (Figure 3).

Multiple enlarged lymph nodes appreciated, in addition to a hard nodule in the right lateral wall. Other structures like Liver,

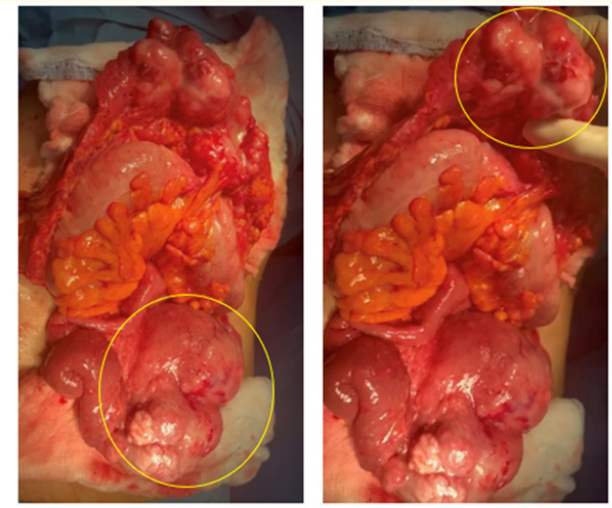

Figure 3: Intraoperative findings of three mesenteric lesions found in Transverse colon omentum not attached to the colon, jejunum and rectosigmoid region.

Uterus and Ovaries looked normal.

During the operation, the mass in the transverse colon was detached from the omentum, the second mass was resected and anastomosis was done. The rectosigmoid mass was left and loop colostomy was created.

Furthermore, the case was discussed in the National Tumor Board where Adjuvant Chemotherapy was suggested.

Patient is currently treated with Adjuvant Chemotherapy and will undergo reversal colostomy once she completes her Chemotherapy cycles.

Discussion

Small bowel lymphoma considered 10-30% of all GI tumors, which is commonly seen in the ileum (60-65%) followed by jejunum lymphoma (20-25%) [1]. The most common histopathology seen was the B- cell lymphoma that usually causes the obstruction, followed by Follicular lymphoma as a rare cause of obstruction [1].

Table 1: Summary of the laboratory findings before surgical intervention.

Laboratory investigation	Result	Unit	Reference range
White blood cell count	6.81	$\times 10^9$/L	3.6-9.6
Hemoglobin	13.6	g/dL	12.0-14.5
Mean cell volume	78.3	fL	80.0-97.0
Platelets	249	$\times 10^9$/L	150.0-400.0
Neutrophils%	81.5	%	42.2-75.2
Lymphocytes%	9	%	20.5-55.1
Amylase	40	U/L	30-118
Random Glucose	4.6	mmol/L	3.6-8.9
Urea	6.1	mmol/L	3.2-8.2
Creatinine	74	µmol/L	40-66
Albumin	47	g/L	34-48
Total Bilirubin	9	µmol/L	5-21
Alkaline phosphatase	147	U/L	46-116
Alanine Aminotransferase	10	U/L	<33
G-Glutamyl Transferase	10	U/L	<38
Sodium	140	mmol/L	132-146
Potassium	3.8	mmol/L	3.5-5.5
Chloride	103	mmol/L	99-109
Bicarbonate	22	mmol/L	20-31
Creatine Kinase	79	U/L	34-145
Lactic Dehydrogenase	283	U/L	135-214
CA 125	213.5	U/mL	<35
CA 15-3	18.7	U/mL	<31.3
CA 19-9	31.8	U/mL	<37
Carcinoembryonic Antigen	0.9	µg/L	1.8-3.5

Furthermore, composite lymphoma is one of the rare types seen as we encountered in our case. The term "composite lymphoma" refers to the presence of two or more morphological different types of lymphomas in single anatomical site [2].

Risk factors includes HIV, Hepatitis B, positive *H. Pylori*, Celiac disease, EBV, IBD, Human T cell lymphotropic virus-1 and immunocompromised [3]. In our case no risk factors were noted.

Clinical presentation of small bowel lymphoma can vary or as in our case it presented as partial obstruction with constitutional symptoms.

Computed tomography demonstrated a mass in the jejunum along with diffuse small bowel wall thickening and multiple enlarged mesenteric lymph nodes.

However, upon discussing the CT findings with the radiologist, another mass in the mesentery of the transverse colon was suspected. Moreover, the differential diagnosis at that time was limited to Gastrointestinal stromal tumors (GISTs) and Desmoid tumors and that explains the patient partial obstruction.

Both Gastro-colonoscopies which were done previously in (November 2022) and (March 2023) showed no masses nor thickening or any kind of suspicion.

In our unique case, the surgical intervention was the only approach to relieve the patient symptoms and reveal the final diagnosis of Lymphoma.

According to the American Cancer Society our patient is categorized as high – intermediate risk with three poor prognostic factors including being stage III disease, elevated LDH level with extra-nodular involvement, making her overall 5-year survival rate of DLBCL is 58%. While the low-grade Follicular lymphoma 5-year survival rate is up to 87% [4].

Table 2: Description of the types of lymphomas that can cause small bowel obstruction [5].

*Three cases were reported as plasmablastic lymphoma, one case as anaplastic lymphoma kinase-positive, large B-cell lymphoma, and one case as immunoblastic lymphoma associated with Crohn's disease.

MALT: mucosa-associated lymphoid tissue [5].

Type of lymphoma	N (%)
Diffuse large B-cell lymphoma*	35 (42.7)
Burkitt lymphoma	18 (22.0)
Mantle lymphoma	11 (13.4)
MALT lymphoma	5 (6.1)
Follicular lymphoma	4 (4.9)
Anaplastic large cell lymphoma	4 (4.9)
Enteropathy associated T-cell lymphoma	2 (2.4)
Post-transplantation lymphoproliferative disorders	2 (2.4)
Lymphoblastoma (Hodgkin's type)	1 (1.2)

Conclusion

In conclusion, the Prescence of small bowel obstruction secondary to lymphoma is a rare presentation. Note to mention that it is uncommon to see Composite lymphoma with multiple masses as in our case.

Human Ethics

Consent was obtained by all participants in this study.

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