Colonic lymphoma presenting acutely with perforated colo-splenic fistula

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**Abstract**

**Introduction:** Primary colonic lymphoma is rare. It comprises less than 1% of large bowel malignancies. Affected patients often present with non-specific vague symptoms with subsequent delays in diagnosis and management.

**Presentation of Case:** An immuno-competent 35-year-old male presented with left iliac fossa pain, fever and constipation. Clinical examination revealed left-sided abdominal peritonism. After the initial radiological and endoscopic investigations, a provisional diagnosis of a localized perforation of a splenic flexure diverticulum was made and ultrasound-guided percutaneous drainage of the abscess was performed. The patient failed to settle on conservative treatment and therefore exploratory laparotomy was carried out. An inflammatory phlegmon consisting of a left paracolic gutter abscess, the spleen and the splenic flexure of the colon was resected en-bloc and a primary colo-colic anastomosis was performed. His operative recovery was complicated by wound infection which was treated conservatively. The histopathology revealed colo-splenic fistula secondary to a perforated colonic non-Hodgkin’s lymphoma. The spleen contained multiple metastatic lymphomatous deposits. He was started on chemotherapy and remained well at 5-year follow up.

**Discussion:** Colon non-Hodgkin’s lymphoma may present initially with an acute abdomen due to perforation. It mimics any acute surgical condition. Perforation and fistulization into the spleen is very rare.

**Conclusion:** This highlights the delay and difficulty in diagnosing primary colonic lymphoma without resorting to surgical resection.

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**1. Introduction**

Primary colonic lymphoma is rare comprising 10–20% of all gastrointestinal lymphomas and less than 1% of large bowel malignancies which makes it the third most common large bowel malignancy after adenocarcinoma and carcinoid. It is often associated with inflammatory bowel disease and immunosuppression, and males are predominantly affected with highest incidence at the age of 50–70 years. Patients often present with vague and non-specific symptoms that subsequently lead to delay in diagnosis which is often made after laparotomy and surgical resection. This delay in diagnosis also results in delayed medical management with detrimental effect on survival.

We report a case of an immuno-competent young male who presented with a colo-splenic fistula and a localized colonic perforation with paracolic abscess formation due to a primary colonic lymphoma. This case highlights yet another an unusual and rare initial presentation of non-Hodgkin’s lymphoma (NHL). The diagnosis was delayed by this an unusual presentation and was only made after surgical resection of the affected colon.

**2. Case report**

A 36-year-old immuno-competent male presented to the emergency room with 4-day history of severe colicky peri-umbilical pain which was radiating to the right flank and back with no relation to food. It was associated with intermittent fever and constipation but there was no associated vomiting or weight loss. He denied any history of urinary symptoms, but he admitted to joint pains and headache. On examination: he looked toxic with a temperature of 38.7°C, pulse 100/min, with normal blood pressure and respiratory rate. Abdominal examination revealed left-sided abdominal tenderness with guarding and rigidity mostly in the left iliac fossa. It was difficult to appreciate any masses or organomegaly, but bowel sounds were audible. Clinical impression of acute diverticulitis was made and therefore he was started on intravenous fluids and antibiotics after full septic work up. His laboratory investigations revealed normal haemoglobin and platelet count but leucocytosis of 11.3 (80% neutrophils, 10% lymphocytes) and high ESR of 70 and C-reactive protein of 32.6 (0–0.3) mg/dl. The urea, electrolytes and amylase were normal and sickle cell test was negative. The liver function test (LFT) was slightly disturbed with raised transaminases [GCT 90 (7–64), ALP 290 (50–136), ALT 122 (30–65), and AST 46 (15–37)] but normal LDH [144 (up to 190 n)]. HIV, Widal test, VDRL and brucella titres were all negative. His stool analysis was negative for ova and parasites. Skin tuberculin test (PPD) was

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negative, and tumor markers and echocardiography were normal. Chest and plain abdominal X-rays were unremarkable. Ultrasonography (US) of abdomen and pelvis showed enlarged heterogenous spleen with numerous hypoechoic patches of variable sizes likely to be splenic abscesses. Based on the US finding splenic tuberculosis was suspected and computerized tomography (CT) scan of abdomen was requested. After 24-h of conservative treatment, he remained pyrexial despite improvement in his general condition and abdominal signs (abdomen became soft and non tender). CT scan of the abdomen showed multiloculated splenic abscesses with suspected tears at the medial and lower side of the spleen involving the mesenteric fat and adherent to the left colon (Fig. 1). Peritoneal effusion with sporadic mesenteric lymphadeopathies was also noted. Blood culture grew Escherichia coli and therefore antibiotics were changed to intravenous piperacillin/tazobactam (Tazocin) 4.5 g twice daily. As his clinical condition markedly improved (the fever settled with marked improvement in his general condition and abdominal signs), colonoscopy was performed on the 4th day after admission. A mass occupying more than 50% of the colon circumference at 75 cm just proximal to splenic flexure with the possible site of perforation was noted (Fig. 2). Multiple biopsies were taken to exclude malignancy but this revealed marked active chronic inflammation suggesting diverticulitis rather than Crohn’s disease.

A week after his admission, repeat CT scan revealed an increase in the size of the splenic abscesses with spillage of colonic contrast into the abscess cavity was also demonstrated (Fig. 1). US-guided drainage was therefore performed; the affluent pus at first but later became faeculent. As clinical condition was improving, conservative treatment was continued. However, after 10 days of admission, he became pyrexial again with increasing abdominal pains. He was taken for an emergency exploratory laparotomy. The findings were enlarged spleen with complete liquifaction of its inferrior pole into the abscess cavity. There was also thickened splenic flexure with a well sealed abscess cavity, but rest of the abdominal cavity was clean. The site of perforation and fistulization into the spleen which was presumed to be a diverticular opening was identified and the abscess cavity contained faecoloma and pus. There were many hugely enlarged mesenteric lymph nodes, but the liver was entirely normal. Frozen section of one excised fleshy mesenteric lymph node was negative for malignancy. The cavity was
There is no doubt that early diagnosis may prevent development of perforation, but the diagnosis is often delayed due to vague symptoms and unusual presentations. Furthermore, as in this case perforation may be the first manifestation of the disease. Some authors even advocate performing hemicolectomy to prevent future perforation which is as high as 45%. However, this is only valid with caution for patients who are diagnosed after repeated endoscopic colonic biopsies (i.e. without surgery) and are expected to receive chemotherapy.

Radiological studies in colonic lymphoma show mucosal nodularities, areas of bowel wall thickening or submucosal masses. However, these findings are non-diagnostic and often repeated colonoascopic biopsies may be needed to establish the tissue diagnosis. But even with these measures one may fail to distinguish lymphoma from carcinoma, metastases, or chronic inflammation. Hence, in most cases, the final diagnosis may only be made with confidence after laparotomy and colonic resection.

The differentiation between the primary colonic and secondary colonic involvement is of great therapeutic and prognostic importance. Moreover, because differences in prognosis and treatment plan, primary colonic lymphoma must be differentiated from other colonic neoplasms and from nodal lymphoma. Dawson criteria (established in 1961) for diagnosing primary gastrointestinal lymphoma depends on absence of palpable superficial lymphadenopathy, absence of mediastinal lymphadenopathy on chest X-ray examination, confinement of disease to gastrointestinal tract which is confirmed by radiology imaging or laparotomy with or without regional lymphadenopathy, absence of hepatic or splenic involvement except through direct spread and finally normal bone marrow aspirate and peripheral blood smear (normal total and differential white blood cell counts). Our patient had mild leucocytosis which may be due to abscess formation, and had no palpable superficial lymphadenopathy or detectable mediastinal lymphadenopathy on radiological investigations. The splenic involvement was due to direct invasion, fistulaization and perforation of the primary colonic lesion rather than due to the primary involvement of the spleen. Moreover, bone marrow aspirate was negative. Hence, based on Dawson’s criteria our case fits well with primary colonic lymphoma.

The treatment for localized lymphoma at diagnosis is combination chemotherapy which leads to prolonged survival and possibly cure. Surgery followed by adjuvant chemotherapy is the standard treatment and using this approach the 5-years survival ranges from 27% to 55%. However, disseminated lymphoma at diagnosis carries poor prognosis. Our case may be considered as disseminated colonic lymphoma as a result of perforation and infiltration into the spleen. However, the patient is alive and free of disease recurrence at 5 years.

The disease is usually aggressive and although the 5-year survival is similar to that in gastric lymphomas (75%), it is significantly higher than in patients with small intestine lymphoma.

4. Conclusion

We report this case to highlight a rare and rather an unusual presentation of NHL of the colon (colo-splenic fistula) in an immunocompetent patient. It also highlights the difficulty with which the diagnosis of colonic NHL is made leading to delay in initiating appropriate medical management. The diagnosis in such cases is only made after laparotomy and resection of the involved colon.

Conflict of interest

Nothing to declare.
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Ethical approval

Patient consent was obtained and is available for review upon editor's request.

Author contributions

Dr. Ali Al-Zahir wrote the case summary and looked up the literature and participated in initiating the first draft. Dr. A-W Meshikhes initiated the idea and wrote the final draft.

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