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Primary Colonic Lymphoma Masquerading as Inflammatory Bowel Disease

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Abstract

Background: Extra-nodal lymphoma with the colon as the primary site, in the absence of predisposing conditions, is an extremely rare clinical scenario. *Case Report*: We present one such case where a primary colonic lymphoma masqueraded as inflammatory bowel disease (IBD) and presented with perforation peritonitis warranting emergency resection. *Conclusion*: Extra-nodal lymphoma of the colon can masquerade as inflammatory bowel disease.

Keywords: Colon, Inflammatory Bowel Disease, Intestinal Perforation, Non-Hodgkin Lymphoma, Peritonitis.

Introduction

Extra-nodal lymphoma primarily involving the colon is very rare and account for less than 1% of colonic malignant tumors [1,2]. Gastrointestinal tract accounts for nearly 35% of extra-nodal lymphoma [2-4]. Stomach is the most common site of primary gastrointestinal lymphoma, followed by small intestine. The clinical presentation of colonic lymphoma is variable and non-specific resulting in diagnostic dilemma. The case presented here was interesting because this patient with lower gastrointestinal symptoms was erroneously being treated as inflammatory bowel disease, until he developed colonic perforation and peritonitis which not only warranted surgery, but also achieved the histological diagnosis. The relation between colonic lymphoma and inflammatory bowel disease is an enigma. This includes an increased incidence of non-Hodgkin lymphoma (NHL) in inflammatory bowel disease (IBD) patients on immunosuppressants [5]. B cell lymphoma complicating Crohns disease is a diagnostic challenge [6]. Very rarely flare up events and non-response to medications in Crohn's disease patients may be due to an underlying second pathology like NHL [7]. Extra-nodal lymphoma of the colon can also masquerade as inflammatory bowel disease [8].

Case Report

A 45 years old male presented with features of peritonitis and sepsis. He was suffering from abdominal pain and loose stools for three months for which he was evaluated elsewhere with colonoscopy. The other significant positive history included weight loss, intermittent fever and vomiting. Blood and biochemical investigations were essentially normal. Colonoscopy was done twice, which showed "ulcerated nodular" lesions distributed as skip lesions, from the rectum to cecum. Histopathological examination was inconclusive, demonstrating only inflammatory cells and atypical cells. However, the patient was started on 5-amino salicylic acid. Patient's symptoms persisted and presented to us with acute abdomen.

After resuscitation patient underwent cross sectional imaging which showed,

pneumoperitoneum with gross free fluid. Cecum and ascending colon were thickened with significant inflammatory stranding. His clinical condition warranted an emergency exploration. At laparotomy, there was a perforation in the cecum with fecal peritonitis. As the patient had skip lesions (colonoscopic finding) involving the entire colon, a subtotal colectomy [**Fig.1]** with ileostomy was done. Patient recovered from surgery without any complications. The HPE of the resected colon showed features of NHL, large B cell type **[Fig.2-5]**. A post-operative PET-CT confirmed the diagnosis of primary colonic lymphoma as there were no lymphadenopathy or other visceral involvement. Subsequently patient received full course of chemotherapy (CHOP regimen) and ileostomy was reversed after 18 months. At present patient is in remission.

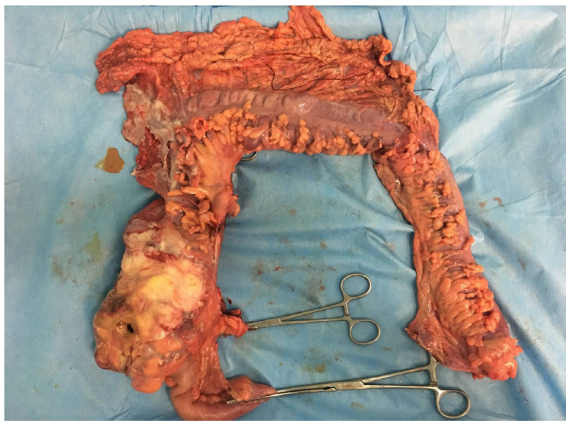

Fig.1: *Subtotal colectomy specimen with the perforation at cecum.*

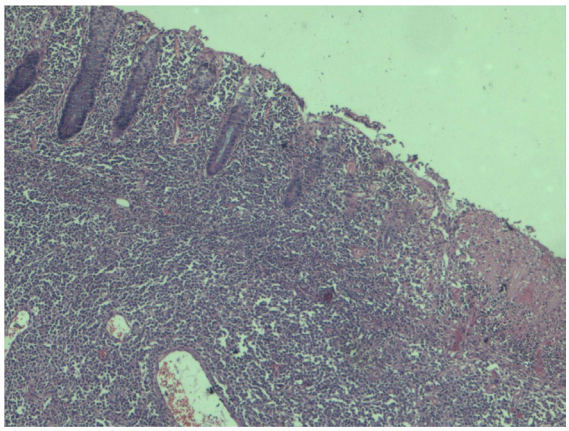

Fig.2: *Low power examination showing the large B cell lymphoma.*

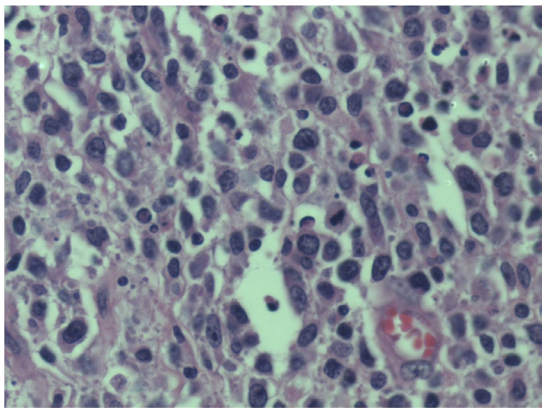

Fig.3: *High power field showing the malignancy composed of large B cells.*

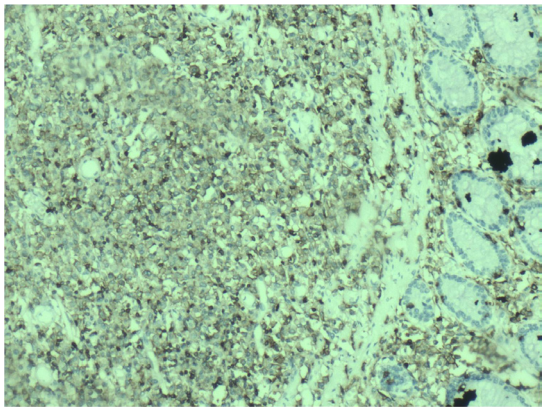

Fig.4: *IHC markers, cells positive for CD 20.*

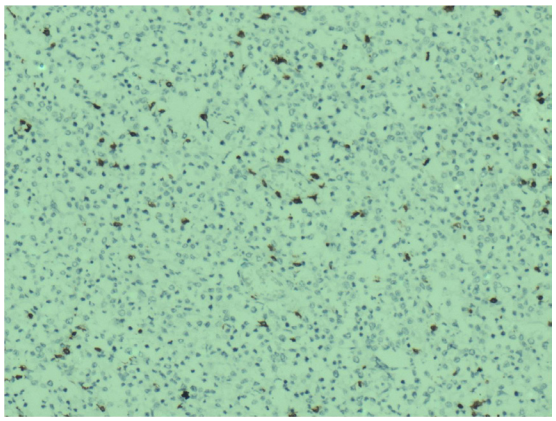

Fig.5: *IHC markers, negative for CD-3.*

Discussion

Primary colonic lymphoma is an extremely rare entity having a histological and immune-histochemical diagnosis. One needs to have a high index of suspicion. The symptoms are usually very non-specific and may mimic other pathological inflictions of the colon. Though rare, primary colonic lymphoma has been reported in the literature and most of the times it is mistaken for other common diseases involving the colon. The association between the inflammatory bowel disease (IBD) especially Crohn's disease and lymphoma is an enigma to the clinical gastroenterologist. There are reports which discusses the increased incidence of lymphoma in IBD patients on immunosuppression therapy, albeit its insignificant [5]. There are clinical reports of diagnosing Crohn's disease initially and even being treated for it with clinical response. However, symptoms recurred in follow-up and on carefully revisiting the old histology revealed it to be lymphoma [8]. Lymphoma has been implicated as a potential complication of Crohn's disease [9] and this phenomenon was exemplified in a case report of a case of Crohn's disease on treatment developed Burkitt's lymphoma after 15 years [7]. The case reported here is unusual because, the patient was an immune-competent individual with no co-morbidity presenting with lower gastro-intestinal symptoms warranting colonoscopy twice and inconclusive biopsy, but nevertheless the clinician decided to treat the patient as IBD. Since he presented to us with perforation peritonitis and sepsis, an emergency laparotomy with subtotal colectomy not only rescued the patient from acute surgical abdomen, but also arrived at a diagnosis.

The acceptable treatment of primary colonic lymphoma is systemic chemotherapy with multiple drugs (CHOP regimen). However, many a times primary colonic lymphoma present like a mass lesion, eluding histological confirmation after repeated biopsies warranting surgical resection. The other indications for surgical resection include obstruction, bleeding and perforation. Perforation is more common in colonic lymphoma than in adenocarcinoma. An untreated intestinal lymphoma can go for perforation. Interestingly perforation has also been reported after starting chemotherapy, as the tumor is chemosensitive and melts completely.

Conclusion

In summary our case emphasizes the diligence required in sampling tissues in atypical cases and the role of immunohistochemistry in the diagnosis of colonic lymphoma.

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