

Case Report

Diffuse Large B-Cell Lymphoma of the Caecum in A Non-HIV Patient - A Rare Occurrence: Case Report

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Abstract: Primary gastrointestinal (GI) lymphomas constitute less than 8 % per cent of total gut neoplasms. Most of them are Non-Hodgkin's type of Lymphomas. Among the Extra-nodal tissues Gastro-intestinal tract is the most common site followed by Head and Neck region, Central nervous system, Thorax, Spleen, Adrenal glands, Uro-genital tract, skin and Bone. The stomach is the most common location of GI lymphomas; followed by the small intestine and large intestine. Although gastrointestinal tract is the most common extra-nodal site yet caecum involvement is rarest. It has low incidence but favorable outcome. Histologically, B-cell lymphomas are by far the most frequent type found in this location, gastrointestinal lymphomas are a diverse group of neoplasms, many of which are characterized by distinctive clinico- pathological settings. Diffuse large B-cell lymphoma and marginal-zone lymphoma of mucosa-associated lymphoid tissue are commonly encountered, but other less common entities can pose diagnostic challenges, mimicking reactive, benign and malignant conditions. Ultrasound (US) is often the first imaging modality used in patients with vague abdominal symptoms and can detect several patterns of involvement in cases of lymphoma. Computerized tomography (CT) is valuable for the staging of the disease. We report a rare case of a 35 year old gentleman who presented with colicky abdominal pain, vomiting, intussusception and bleeding per rectum was diagnosed as a case of caecal lymphoma on histopathology.

Keywords: Intussusception , Caecal Lymphoma, B-Cell , Gastro-Intestinal Tract, Non-Hodgkin's.

INTRODUCTION

The involvement of extra nodal sites is a common feature of non-Hodgkin lymphomas (NHL's). Moreover, some NHLs are considered to originate at sites other than the lymph nodes or spleen and are referred to as primary extra nodal lymphomas (PE-NHL) [1-2]. The origin of most PE -NHLs can be ascribed to one given organ system or site. However, there exists a heterogeneous collection of NHLs that may involve multiple sites throughout the body at presentation. Although most of these cases are localized to the gastrointestinal (GI) tract, they also involve a variety of extra nodal sites outside the GI tract. The term MALT lymphoma was first described by Isaacson and Wright in 1983 [3]. Primary gastrointestinal lymphoma is very rare, constituting only about 1-4% of all gastrointestinal malignancies [4]. Lymphomas involve the gastrointestinal tract either as primary neoplasms or as part of disseminated disease. Primary lymphoma of the caecum is rare among the gastro intestinal malignancies. Non-Hodgkin lymphoma accounts for almost all Gastro-intestinal lymphomas[5-7]. The signs and symptoms of colonic lymphoma are non-specific. The lack of specific symptoms can be the

reason for delayed diagnosis. The most common presentation is abdominal pain, with weight loss and changing bowel habits in 60-90% of patients. A palpable abdominal mass can sometimes be noted on the initial physical examination. Adult intussusception is uncommon and accounts for 5% of all intussusception and 1% of all bowel obstruction[8]. Mucosa associated lymphoid tissue (MALT) and diffuse large B cell lymphomas (DLBCL) are the two histological subtypes most commonly observed[9]. Ultrasound (US) is often the first imaging modality used in patients with vague abdominal symptoms and can detect several patterns of involvement in cases of lymphoma. Computerized tomography (CT) is valuable for the staging of the disease and is an essential complementary study to barium examination. It establishes the extent and shape of a lymphoma, demonstrates nodal involvement and possible infiltration of the liver or spleen[10].

CASE REPORT

A 35 year old young gentleman presented to casualty with abdominal pain since 15 days on and off, colicky in nature, sometimes associated with vomiting.

- History of bleeding per rectum one episode 15 days back
- History of fever since 4 days
- History of burning micturition since 2 days
- Patient is a Non-Diabetic, Non-Hypertensive, Non-HIV, HbsAg – Negative.

On Examination abdomen was soft with Epigastric tenderness. Patient condition was diagnosed as Acid Peptic Disease and referred to surgery department. Patient's blood pressure -110/80 mm of Hg, Cardio-vascular system and respiratory systems were normal. Per Abdomen – Soft, Tender in the right lumbar region, Mass palpable in Right lumbar region. Mass was not mobile, bimanually palpable, ballotable and resonant. Patient was treated symptomatically with anti-spasmodics, antacids and proton pump inhibitors. Patient was advised to undergo Major surgical profile, X-ray Chest postero-anterior view and USG abdomen. All laboratory Investigations were normal. Ultrasound findings – Target sign in right upper quadrant of abdomen with minimal ascites. Impression: Intussusception.

Patient was further advised to undergo CECT Abdomen which revealed mass in the caecum (FIGURE 1). Patient was stable, posted for Explorative laparotomy for reduction of Intussusception (FIGURE 2). On laparotomy a mass was palpable in the cecum (FIGURE 3). Caecal mass along with the Ileo-caecal junction and part of the intestine was excised and sent for Histopathological examination (HPE).

HISTOPATHOLOGY

Grossly, we received specimen of distal Ileum, Ileo-caecal junction and proximal ascending colon and mesenteric lymph node separately. Ileo-caecal junction, caecum cut opened and everted showing a polypoidal growth measuring 5 x 5 x 2.5 cm (FIGURE 4). Resected small intestine measuring 8 cm in length. Caecum and ascending colon measuring 10 x 7 x 2 cm. Appendix measuring 5 cm in length. Received single lymph node measuring 1.5 x 1 cm.

Microscopy revealed submucosal tumor tissue arranged diffusely and showing monotonous population of cells with individual cells are large, oval to round with mild pleomorphism, hyperchromatic nuclei and scant cytoplasm (FIGURE 5,6 &7). Tumor tissue was invading up to the serosa without involving the adjacent structures. Resected margins are free of tumor cells. Appendix shows lymphoid hyperplasia. Mesenteric lymph node shows reactive hyperplasia and sinus histiocytosis. A diagnosis of primary diffuse large B-cell Lymphoma was given. TNM Staging –T3 N0 M0.

Immuno-Histochemistry (IHC) with antibody CD-20 was done to confirm the histology of the cell type which came to be positive which confirmed that the cell of origin is B-Cell (FIGURE 8).



Fig. 1: CT Image showing intussusceptions and caecal mass.



Fig. 2: Pre-operative image of the patient with area of the swelling marked with blue Ink.

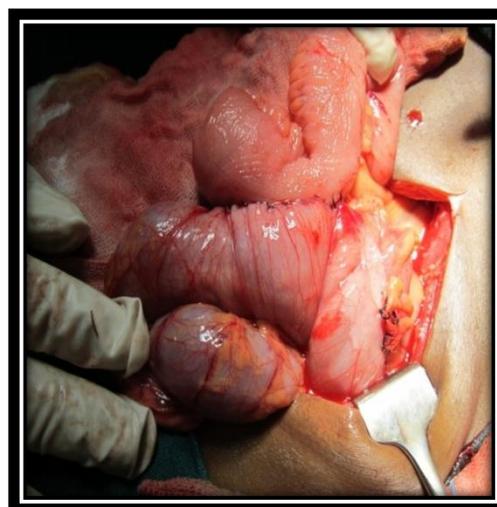


Fig. 3: Intra-operative image with caecal mass and intussusception.

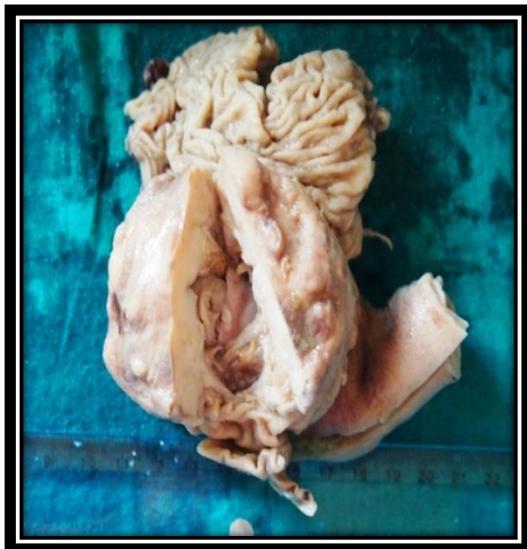


Fig. 4: Gross specimen of ileo-caecal junction with everted polypoidal caecal mass.

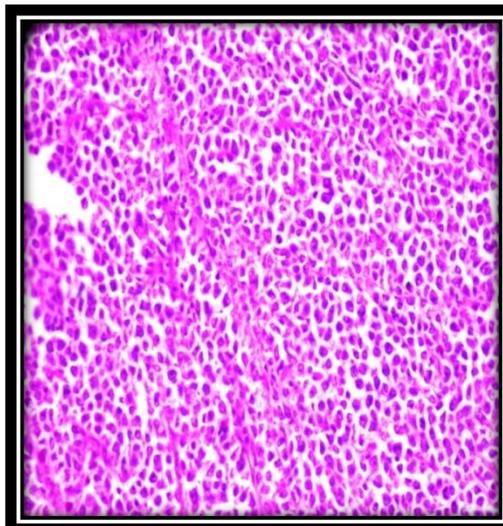


Fig. 7: High-power view of tumor tissue showing monotonous population of cells with mild to moderate pleomorphism and scant cytoplasm.

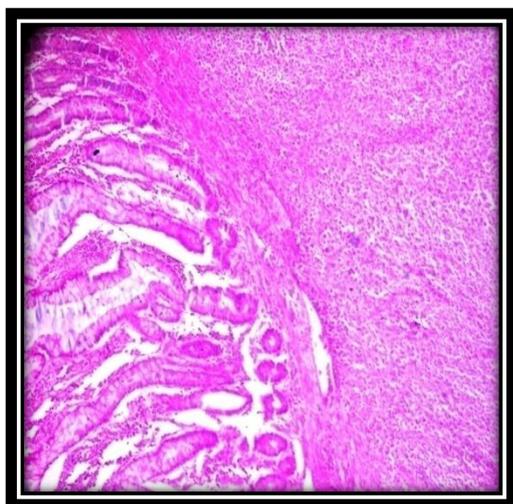


Fig. 5: Low-power view of mucosa with submucosal tumor tissue.

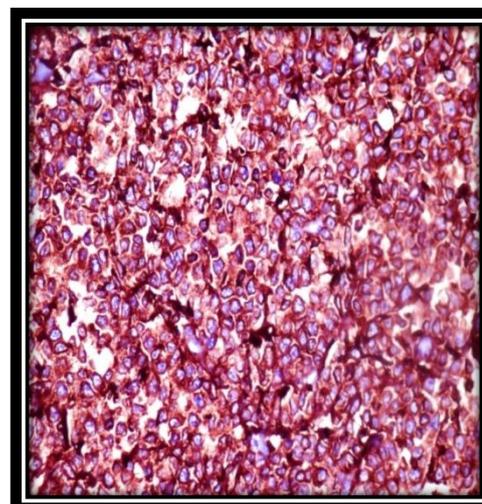


Fig. 8: Immuno Histochemistry image showing strong cytoplasmic positivity for marker CD-20.

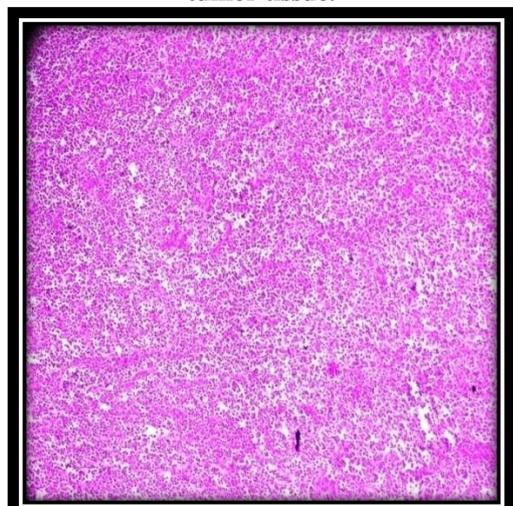


Fig. 6: Low-power view of the tumor tissue arranged in sheets.

DISCUSSION

Non-Hodgkin's Lymphoma (NHL) constitutes a group of disorders originating from the malignant transformation of lymphocytes and involving either the lymph nodes or extra nodal sites. Extra nodal lymphomas may comprise 25%–49% of NHL cases, and there appears to be an increasing incidence of these lymphomas during the past decade [1]. Extra nodal lymphomas may occur in any organ. They present most frequently in the Gastro Intestinal tract, followed by Head and Neck, Thorax, Uro-genital tract, Waldeyer's ring, when tonsils are regarded as an extra nodal site. Other common sites are skin and bone. Although there are reports on Primary Extra nodal Non-Hodgkin's Lymphoma (PE-NHL) of various sites, especially GI-NHL, there remain many questions concerning the clinic-pathological features and treatment outcome of these patients. According to the literature reviewed and many authors, Diffuse Large B-Cell Lymphoma

(DLBCL) was the most frequent histological subtype, comprising 62% of cases. Next in frequency were low-grade MALT lymphomas (27%). Common age of presentation is middle age and older adults with both sexes being equally affected. Most common etiological factors include recurrent infections with Epstein –Barr Virus (EBV), inherited mutations involving tumor suppressor gene p53, patients with auto-immune diseases and organ transplantation cases who are on immuno modulator and immuno suppressor drugs. Some of the less common etiological factors include infections with various viruses and bacteria, celiac disease, inflammatory bowel disease and immunodeficiency syndromes.

Reports of colonic intussusception in adults are less common in literature and if occurs they are confined to both small and large bowel. Colonic intussusception presents in a subacute manner with prolonged abdominal pain and constipation[11]. Primary intestinal lymphoma most commonly involves the Ileo-caecal region, probably due to the high proportion of lymphoid tissue [12]. Our case was primary B-Cell lymphoma of the Ileo-caecal region which presented as intussusceptions, abdominal pain and had no regional lymph node involvement. Regional lymph nodes, if involved, are found to have a bad prognosis. The patient underwent surgery followed by chemotherapy with uneventful course during the three months follow-up period.

Immunohistochemical analysis helps in confirming the cell of origin of the lymphomas by detecting positivity for immuno-globulins and pan B antigens – CD19, CD20, CD79 without expression of CD5, CD10, CD23 and cyclin D1[13]. In our case, the B-cell marker CD-20 showed strong cytoplasmic positivity.

Treatment

Combined modality of approach that includes surgical excision and systemic chemotherapy is the preferred treatment [14]. Different therapeutic approaches were used in two subsets: Radical tumor resection (hemicolectomy) plus multi-agent chemotherapy (polychemotherapy) in early stage patients, biopsy plus multidrug chemotherapy in advanced stage patients. Polychemotherapy includes CHOP (cyclophosphamide, doxorubicin, vincristin, and prednisolone) or CHOP-like combination chemotherapy or MACOP-B-like regimens. Surgery alone can be considered as an adequate treatment for patients with low-grade NHL disease that has not infiltrated beyond the submucosa[14]. Those with limited stage disease may enjoy prolonged survival when treated with aggressive chemotherapy. Radiotherapy is beneficial for incomplete resection or non-resectable disease.

CONCLUSION

Primary caecal lymphoma is a rare occurrence; early diagnosis may prevent intestinal perforation; however, the diagnosis is often delayed in most cases due to varied clinical presentations. Surgical resection is the mainstay of treatment for localized primary lymphomas, followed by postoperative chemotherapy. Surgery alone can be considered as an adequate treatment for patients with low-grade Non-Hodgkin's lymphoma disease that does not infiltrate beyond the serosa. The overall prognosis of the caecal lymphoma when diagnosed in the early stages is good. In our case, the prognosis was good due to early stage of disease and early diagnosis and treatment.

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