Research Paper

Volume : 5 | Issue : 5 | May 2016 • ISSN No 2277 - 8179 | IF : 3.508 | IC Value : 69.48

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

Medical Science

KEYWORDS : jejunum Lymphoma, B-Cell , Gastro-Intestinal Tract, Non-Hodgkin's.

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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 jejunum 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 60 year old gentleman who presented with colicky abdominal pain, vomiting, and loss of weight was diagnosed as a case of jejunum lymphoma on histopathology.

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 jejunum is rare among the gastro intestinal malignancies. Non-Hodgkin lymphoma accounts for almost all Gastro-intestinal lymphomas[5- 7]. The signs and symptoms of small intestinal 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. 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 60 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 loss of weight 1 month
- History of fever 5 days
- Patient is a Non-Diabetic, Non –Hypertensive, Non- HIV, HbsAg – Negative.

On Examination abdomen was soft with Epigastric and left lumber tenderness. Patient condition was diagnosed as Acid Peptic Disease and referred to surgery department. Patient's blood pressure -130/80 mm of Hg, Cardio- vascular system and respiratory systems were normal. Per Abdomen – Soft, Tender in the left lumber region. No mass was palpable. Patient was treated symptomatically with anti- spasmodics, antacids and proton pump inhibitors. Patient was advised to undergo Major surgical profile, X-ray Chest posterio-anterior view and USG abdomen. All laboratory Investigations were normal. Ultrasound findings – small bowels Dailated(3-3.5 cm) with thickened and edematous walls and shows multiple echoes with strong posterior shadow in left lumber region.

Patient was further advised to undergo CECT Abdomen (FIGURE 1, 2). Patient was stable, posted for Explorative laparotomy (FIGURE 3). On laparotomy a mass was exophytic in jejunum 15 cm fron DJ junction (FIGURE 4). Jejunum mass resected and jejuno-jejunal anastomosis done and sent for Histopathological examination (HPE).

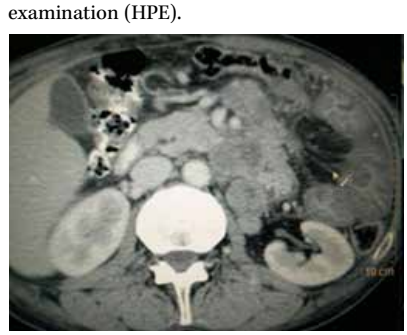

Figure1: CT scan showing mass in jejunum

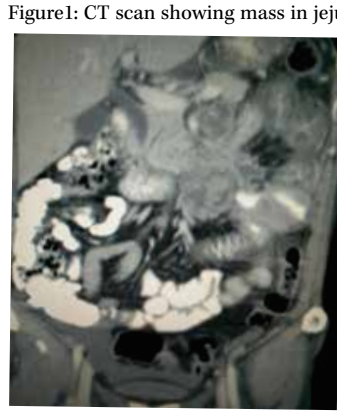

Figure2 : CT scan showing lymphnode metastasis

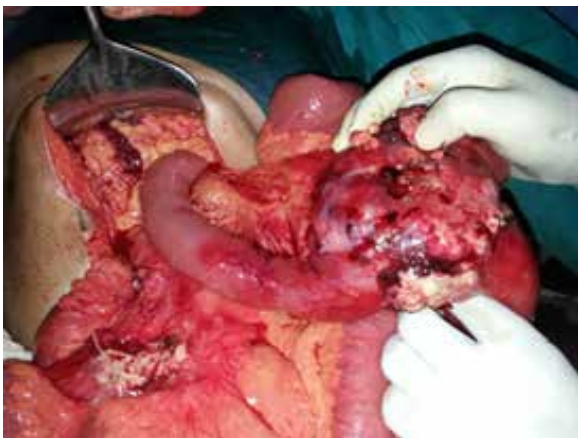

Figure 3 : Intraoperative image of jejunum mass

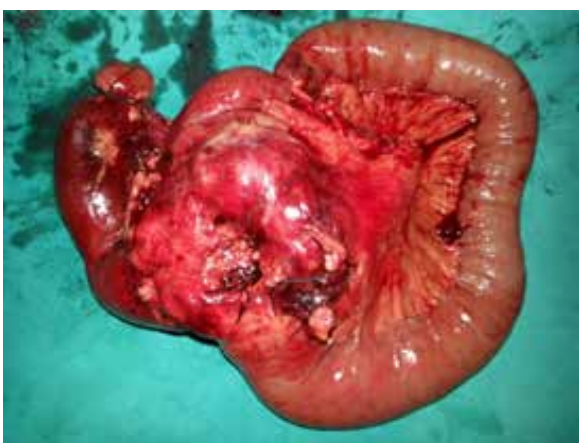

Figure 4 : Image of resected jejunum mass

HISTOPATHOLOGY :

Grossly, we received specimen jejunum with mass and mesenteric lymph node separately. Resected small intestine measuring 45 cm in length mass away from one end 8 cm and 5cm from 2nd end. 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. Tumor tissue was invading up to the serosa and mucosa. Lymphnode shows replacement of lymphnode architecture by lymphoma cells. 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. A diagnosis of primary diffuse large B- cell Lymphoma (Non-Hodghkin's type) was given.

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.

Our case was primary B-Cell lymphoma of the jejunum region which presented as abdominal pain, vomiting, loss of wieght and had 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 immunoglobulins and pan B antigens – CD19, CD20, CD79 without expression of CD5, CD10, CD23 and cyclin D1[12]. 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 [13]. Different therapeutic approaches were used in two subsets: Radical tumor resection 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[13]. 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 jejunum 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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Volume : 5 | Issue : 5 | May 2016 • ISSN No 2277 - 8179 | IF : 3.508 | IC Value : 69.48

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