International Journal of Biomedical And Advance Research

ISSN: 2229-3809 (Online)
Journal DOI:10.7439/ijbar
CODEN:IJBABN

Review Article

Ileocaecal intussusception secondary to Burkitt's lymphoma

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Abstract

A 20 year old male presented with features of intestinal obstruction. Contrast enhanced computer tomography revealed a target lesion at cecal region consistent with ileoceacal intussusception. Emergency laprotomy was done with hemicolectomy and ileo-colic anastomosis. Histopathology of the specimen showed features of non Hodgkin lymphoma. Immunohistochemistry showed tumor cells positive staining for CD20, CD79a and weakly positive for CD10, Bcl-2, Bcl-6 and negative for CD3 consistent with Burkitt's lymphoma.

Keywords: NHL; Burkitt lymphoma;intussuception

1. Introduction

Gastrointestinal site is the most common extra nodal localization of lymphomas. Stomach is most frequently affected by lymphoma of gastrointestinal tract. Primary intestinal Non Hodgkin Lymphoma is the third most common small intestinal neoplasm after adenocarcinoma and carcinoid tumors. Presentations of intestinal lymphoma are of nonspecific abdominal pain, weight loss, fever, constipation. Rarely they can present as acute abdomen with obstruction perforation. We present a rare case of ileo-caecal intussusception secondary to Burkitt's lymphoma.

2. Case Report

A 20 year old male presented to emergency department with pain abdomen of 2 days duration with vomiting and constipation. Patient was hemodynamically stable and on examination there was an ill-defined mass in the right iliac fossa. Contrast enhanced computer tomography showed a target lesion in ileocaecal region with multiple dilated loops proximally. Features were consistent with ileo-cecal intussusception.(Fig 1) All blood investigations were within normal limit. Patient was taken up for emergency laprotomy which showed ileo-caecal intussusception with an exophytic growth.(Fig 2,3) With high suspicion of malignancy no attempts were made to reduce the intussusception and right hemicolectomy with ileo-colic anastomosis was done. Post operative period was uneventful.

Histopathological examination of the specimen revealed a firm grey white area in the middle of both resected surgical ends.Microscopy showed adipose infiltrating small round cell lesion, reaching upto the serosa. The cells were in sheets,having large vesicular nuclei with minimal cytoplasm. Prominent nucleoli with scattered mitotic figures.(Fig 4) Lymph node shows reactive lymphoid follicles. Features suggestive of a Non Hodgkins lymphoma of Small intestine. Immuno histochemistry showed tumor cells positive for CD20,CD79a weakly positive for CD10 focally weakly for Bcl-2, Bcl-6 negative for CD3 consistent with Burkit's Lymphoma.

Patient underwent whole body computer tomography for staging which showed few lymph nodes in Right upper paratracheal, lower pretracheal aortopulmonary window and bilateral axilla and few small nodes in right iliac fossa. Serum LDH-174U/L and all oter blood parameters were within normal limits. Bone marrow biopsy was normo to hypocelluar with reactive lumphocytosis. CSF analysis was within normal limits.

Due to financial constraints patient was started on CHOP regimen and is on regular followup. At 1year patient showed no signs of recurrence or any abdominal symptoms.

Fig 1: Computer tomography picture showing 'target sign' ileo-caecal intussusception.



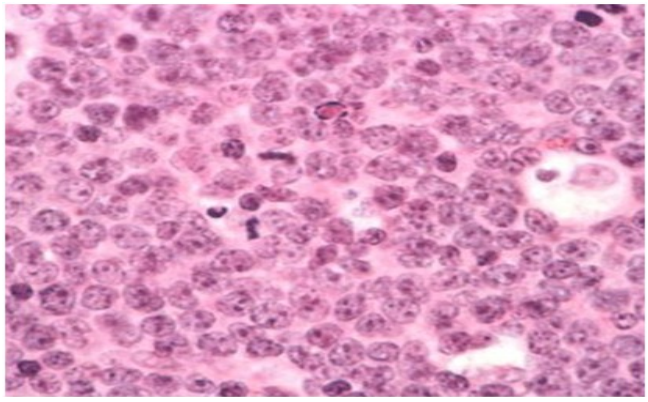
Fig 2: Intra operative picture showing ileo-caecal intussusception



Fig 3: Intra operative picture showing intussusception with exophytic growth.



Fig 4: Histopathology image showing cells with large nuclei and scanty cytoplasm and scattered mitotic figures. Features suggestive of Burkitt's lymphoma.



3. Discussion

Intussusception occurs when a proximal segment of bowel (intussusceptum) telescopes into the lumen of an adjacent distal segment (intussuscipiens) and can occur anywhere within the gastrointestinal tract. Although fairly common in children, adult intussusception is relatively rare representing only 1% of patients with bowel obstructions.[1,2].

Intussusceptions are classified according to location, with the most common classification system divides intussusception into four categories: enteric, ileocolic, ileocecal, and colonic[3-6]. Enteric and colonic intussusceptions are those that are confined to the small intestine and large intestine, respectively. Ileocolic intussusceptions are defined as those with prolapse of the ileum through the ileocaecal valve into the colon and these constitute 15% of all intussusceptions. The ileocaecal valve and the appendix preserve their normal anatomic position and the organic lesion is usually in the ileum[7]. These organic lesions are mostly benign although malignant lesions, can also be seen[8-11].

Symptoms of intussusception in adults are non-specific and diagnosis is generally made during laparotomy. In 25% of patients, intestinal obstruction, rectal bleeding and a mass in the abdomen is seen. Barium enema, CT and colonoscopy are helpful in diagnosis[12-14], with barium enema being the most important diagnostic tool. It may show the characteristic "coiled spring" or "spiral sheath" appearance, or a mushroom or crescentic shaped appearance at the level of obstruction . Pseudokidney and spiral images of intestinal loops are specific CT signs. Endoscopy can be used for definitive diagnosis of intestinal intussusceptions .

In adult intussusceptions, the treatment of choice is surgical, with resection of all intussusceptions without intraoperative reduction being advocated. The type of surgical intervention is based on the patient's medical history and intraoperative findings[3].

Primary intestinal NHL is the third most common small intestinal neoplasm after adenocarcinoma and carcinoid tumors.The gastrointestinal tract (GIT) is the most frequent primary extranodal localization for an average of 12% of all NHL and 30%-40% of all extranodal sites [15]. With primary intestinal NHL to account for 21%-54% of all GIT sites[16].

The incidence of the disease is rising particularly among immunocompromised patients. While little is known about the pathogenesis of this disease, patients with celiac disease have a 200-fold increased risk of developing intestinal lymphoma, the so called enteropathy-associated T-cell lymphoma.[17].

There are at least two definitions of primary GI NHL in use. The one by Dawson et al.(18) is restricted to localized disease (stages IE, IIE), whereas that by Lewin et al[19]. Requires that patients exhibit GI symptoms or predominant lesions in the GI tract.

Four main sites of origin for GI NHL can be distinguished at diagnosis. The most frequent location is the stomach (74.5%). The second largest group is lymphoma originating in the small bowel (8.2%), followed by those of the ileocecal region (7.2%). Isolated involvement of the duodenum and colon are rare.Pain is the main diagnostic symptom in most cases (76%), followed by loss of appetite in (46%) , loss of weight(35%). Constipation and ileus are more frequently encountered in intestinal lymphoma and considered as diagnostic symptoms for the intestine when combined as signs of occlusion. Occult loss of blood or macroscopic bleeding is most frequent when the stomach or the ileocecal region are involved. Perforation and obstruction are rare initial manifestations. B symptoms (fever, night sweats and loss of weight) occur in about25% of cases. Median time from onset of symptoms to diagnosis is the shortest for ileocecal lymphoma (75 days) and longest for multiple GI involvement (135 days)[20].

Pathological diagnoses are made according to the Revised European-American Lymphoma (REAL) classification or the World Health Organization (WHO) classification. Ann Arbor classification[21] incorporating Musshoff's sub-division of stage II(22) and the one by Radaszkiewicz et Al(23) for stage I. The suffix "E" should be restricted to describe invading growth, whereas the suffix "X" was proposed for extranodal origin, followed by the information of the actual stage (eg,X[stomach]II). In case of simultaneous multiple primary GI involvement, the organs should be enumerated after the suffix "X" (eg, X[stomach, colon]II)[24].

Gastrointestinal non-Hodgkin's lymphomas as such can be considered as a localized disease, with the exception of multiple GI involvement , Stage I is defined as disease confined to the intestine, stage II is defined as disease extending to local (II-1) or distant (II-2) nodes, stage II-E is defined as disease involving adjacent organs or tissues, and stage IV is defined as disseminated extra-nodal involvement or concomitant supradiaphragmatic lymph node involvement. The IPI risk is calculated from five parameters including age, performance status, serum LDH, number of extranodal involvement and Lugano stage[25]. Stages III and IV form about 16% of cases. A slight tendency for higher spread could be noticed in NHL of the small bowel, compared with gastric and ileocecal lymphoma, of which more than 70% are diagnosed in the very localized stages IE and II1E. stage IIE was divided by Musshoff et al.[26]. .into stage II1E (involvement of regional lymph nodes only (gastric/mesenteric) and stage II2E [involvement of distant lymph nodes (para-aortic/para-caval)]

The ileocecal region is the most common site of involvement, accounting for approximately 40% of primary sites. However, this region is mainly affected by B-cell lymphomas (95.7%). The frequent occurrence of B-cell lymphomas in the ileocecal region is associated with high proportions of DLBCL . T-cell lymphomas are extremely rare in the ileocecal region (4.3%), while involvement of the jejunum is more common in T-cell lymphomas (12.5%) than in B-cell (3.6%). A comparison of survival outcomes based on primary site of involvement revealed that involvement of the ileocecal region was associated with better survival rates than involvement of the small and large intestine.[20]

The optimal treatment strategy for intestinal lymphoma is still unclear. Although conservative treatment is preferred to surgery in localized gastric lymphomas, the same is not true for intestinal lymphomas because surgery in combination with chemotherapy has proven superior to any other treatment combination[27,28]. Regarding treatment, it has been established that the primary surgical treatment had the most favorable influence on failure-free survival in localized diseases and hence the resection may be appropriate as the primary treatment[29].On the other hand, the effectiveness of adjuvant therapy for localized NHL remains to be unclear, because some cases could be cured only by surgical resection[30,31].

The efficacy and toxicity of adjuvant therapy for localized NHL remained unclear because excess chemotherapy may provoke secondary malignancies. On the other hand, a new anti-CD20 mAb, rituximab, is effective in the treatment of

B-cell lymphoma with slight adverse effects[32].

The combination of cyclophosphamide, doxorubicin, vincristine, and prednisone, given every 3 weeks (CHOP-21) is standard chemotherapy for aggressive lymphomas. CHOEP(CHOP with Etoposide) achieves better complete remission (87.6% versus 79.4%)and 5-year event-free survival rates (69.2% versus 57.6%)than CHOP, whereas interval reduction improved overall survival .Although the CHOEP regimens induced more myelosuppression, all regimens were well tolerated. CHOEP should be the preferred chemotherapy regimen for young patients with good-prognosis (normal LDH level) aggressive lymphoma[33].

The acute reactions of different grades according to the Standard WHO toxicity criteria attributable to the chemotherapy included leukopenia (<3000 white cells per microliter) in 40% of the patients, oral ulcers in 19%, alopecia in 100%, diarrhea in 66%, nausea and vomiting in 73%. The acute reactions attributable to chemotherapy combined with radiotherapy of different grades included diarrhea in 70%, bladder irritation in 21%, fatigue in 63% and leukopenia in 45%.[34]

Patients with multiple intestinal involvements had the worst survival outcomes.There are several possible explanations for the superior survival outcomes of patients with involvement in the ileocecal region. First, T-cell lymphoma rarely occurs in the ileocecal region compared to the small and large intestine. Second, lymphomas in the ileocecal region often presented with complications, such as obstructions requiring surgical intervention. Thus, more than 50% of patients with lymphoma in the ileocecal region underwent immediate surgery[27].

Gastric and ileocecal lymphomas show a higher disease free survival (DFS) and overall survival(OS) as compared to NHL of the small bowel. The OS at 5 years for all intestinal lymphomas together is 66.2% compared to 83.3% for patients with gastric lymphoma. ECOG performance status >1, advanced stage, high grade tumors and elevated LDH significantly reduces OS. There is a significant difference in survival rates between low- and high-grade PGL in DFS and OS (at 5 years DFS and OS for low-grade lymphoma were both 98.21% compared with 60.0% and 70.91% respectively, for high-grade NHL). The DFS and OS were significantly worse for patients with high-grade with low-grade MALT-type component (73.08 and 80.77 respectively), compared to low-grade MALT-type NHL (both were 98.21%).[20]

4. Conclusion

Gastrointestinal tract is most common extranodal site for lymphoma. Intestinal lymphoma has varied presentation. Intussusception and obstruction in young adults can be secondary to intestinal lymphoma. Surgical resection with chemotherapy forms the main mode of treatment of intestinal lymphoma.

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