SMALL INTESTINAL BURKITT'S LYMPHOMA PRESENTING AS AN ACUTE ABDOMEN SECONDARY TO ILEO-COLIC INTUSSUSCEPTION IN A 28 YEARS OLD MALE: A CASE REPORT

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Abstract

SMALL INTESTINAL BURKITT'S LYMPHOMA PRESENTING AS AN ACUTE ABDOMEN SECONDARY TO ILEO-COLIC INTUSSUSCEPTION IN A 28 YEARS OLD MALE: A CASE REPORT

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Significance: Burkitt's lymphoma is a highly aggressive and rapidly growing B-cell neoplasm. Symptoms are nonspecific and renders its diagnosis to be difficult. Sporadic Burkitt's lymphoma accounts only for 1%-2% of lymphoma in adults. Intestinal intussusception secondary to Burkitt's lymphoma in adults is considered to be uncommon.

Clinical Presentation: This is a case of a 28 years old male presenting with right lower quadrant pain. His physical examination presented with an acute abdomen appearing with hypoactive bowel sounds, with direct and rebound tenderness on the right lower quadrant with noted guarding. The rest of the organ systems were unremarkable.

Management: CT Scan done revealed a rounded soft tissue mass in the ileocecal junction and is seen as a lead point of intussusception. Patient underwent exploratory laparotomy with right hemicolectomy and ileo-transverse colostomy with end to side anastomosis and biopsy taken from the ileal mass was consistent with Burkitt's lymphoma which was positive for CD 20 immunostaining. Patient was then started on 6 cycles of chemotherapy thereafter.

Recommendation: The importance of considering intussusception in adults as a possible cause of an acute abdomen cannot be overemphasized. This may contribute to possible future case series or studies for allowing earlier detection and management of this rare condition.

Keywords: Burkitt's lymphoma, intussusception, acute abdomen

Introduction:

Primary gastrointestinal lymphoma represents only 1– 4% of all gastrointestinal malignancies with Burkitt' s lymphoma accounting for 0.3– 1.3% of all non-Hodgkin' s lymphomas [1]. Burkitt's Lymphoma of the small Intestine can affect any portion of the small bowel, but typically involves the ileum and is a rare type of gastrointestinal lymphoma that is more commonly seen in children and adolescents [2].

Intussusception is the telescoping of one segment of the gastrointestinal tract into its adjacent segment. It is uncommon in adults, with only two to three cases occurring in a population of 1, 000,000 annually [3]. Adult intestinal intussusception is infrequent which accounts for 1 to 5 percent of mechanical bowel obstructions that is often caused by intraluminal neoplasms such as the following carcinoid tumors, small bowel carcinomas, and lymphomas [4]. Intussusception can be categorized by etiology such as benign or malignant lesions or may be idiopathic. It can also be classified by its involvement as to entero-enteric, ileo-colic or colo-colic.

Intermittent abdominal pain is the most common presentation. The diagnosis is often made clinically with abdominal CT scan findings of a target sign on the axial view and appearance of a sausage-shaped mass on coronal view accompanied by tissue biopsy through surgical resection [4].

This reports an uncommon case of ileo-colic intussusception secondary to a small intestinal Burkitt's lymphoma presenting in a young adult male Filipino.

Objectives:

General Objective:

To present a case of a 28 years old Filipino male diagnosed with ileo-colic intussusception secondary to small intestinal Burkitt's lymphoma.

Specific Objectives:

1.) To review on the epidemiology, etiology, and diagnosis of small intestinal Burkitt's lymphoma presenting as an ileo-colic intussusception.

2.) To review on the management and prognosis of small intestinal Burkitt's lymphoma presenting as an ileo-colic intussusception.

The case:

A 28 years old male Filipino sought consultation due to right lower quadrant pain. 4 days prior to admission, patient had onset of crampy right lower quadrant pain 6/10 with associated nausea and bloatedness. He had no fever, no flank pain nor vomiting, no anorexia, and no episode of loose stools, with no recent change in bowel habits and claimed to have daily bowel movement and flatulence. Patient tolerated condition until on the day of admission, increasing intensity of the right lower quadrant pain to a scale of 10/10 thus prompted admission. Patient has no known comorbidities nor prior surgeries. He is a non-smoker and non-alcoholic beverage drinker. He has no known food allergies, denied any illicit drug use and has no history of blood transfusions. Patient has no heredofamilial diseases such as hypertension, diabetes, bronchial asthma, hepatobiliary diseases, blood dyscrasias or cancer.

At the emergency room, vital signs were stable with a blood pressure of 120/70mmHg, temperature of 37^0 C, respiratory rate of 20 cycles per minute and heart rate of 76 beat per minute. Abdominal examination revealed consistent with an acute abdomen that appeared slightly distended with hypoactive bowel sounds, tympanitic on percussion with direct and rebound tenderness on the right lower quadrant with noted guarding. The rest of the organ systems was unremarkable. The initial impression at this time was acute appendicitis.

Ancillary tests were taken which revealed the following: CBC revealed leukocytosis with WBC of 14.95 x 10^3/uL, microcytic, hypochromic pattern of anemia with a hemoglobin of 13.7 g/dl, and with a predominance of neutrophils of 78%. Electrolytes and creatinine were unremarkable. A CT scan of the whole abdomen with contrast was done which revealed a rounded soft tissue mass approximately measuring 3.5x4.2x3.6cm in the ileocecal junction and is seen as a lead point of intussusception that is consistent with an ileocolic type of intussusception.

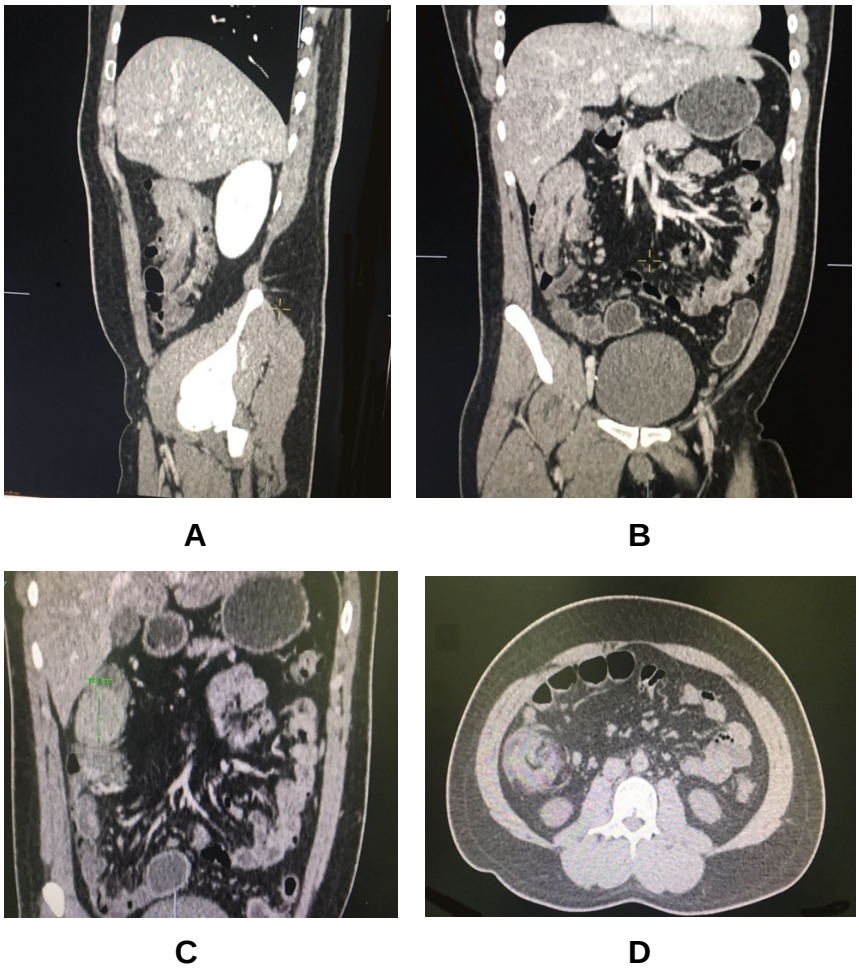
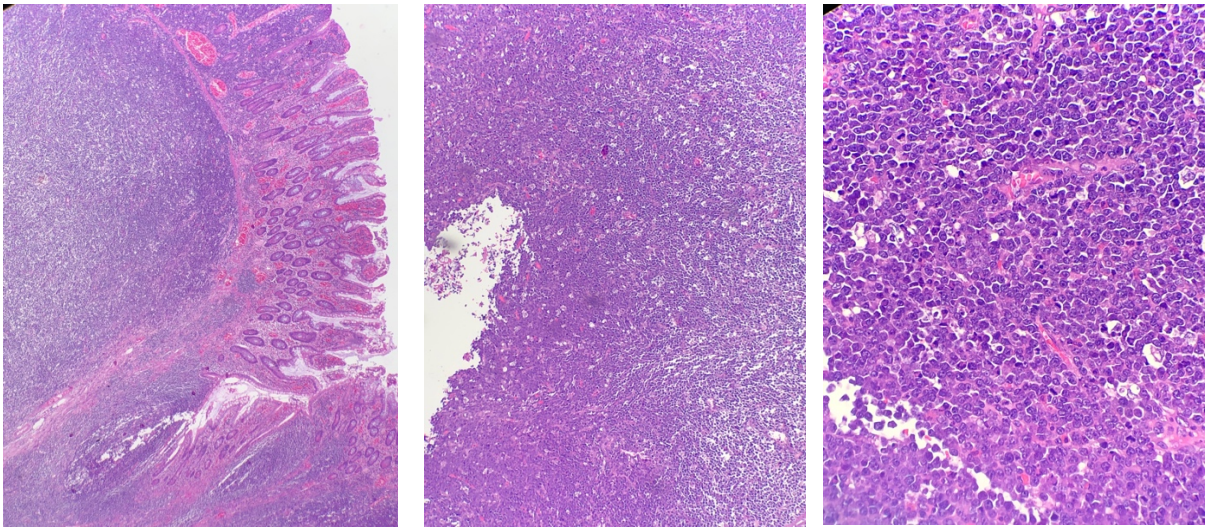


Figure A. demonstrates a sagittal view with noted telescoping of the ileal segment into the ascending colon. Figure B and C both shows both a coronal view with noted telescoping of the ileal segment into the ascending colon. For figure C, a sausage shaped mass is noted indicative of intussusception. Figure D shows the target sign which shows alternating hypo/hyperdense layers suggestive of intussusception.

Patient was then started on iv ampicillin-sulbactam and subsequently referred to surgical service and scheduled for exploratory laparotomy with right hemicolectomy and ileo-transverse colostomy with end to side anastomosis. Intraoperative findings included the small bowel ileum which intussuscepted into the cecum with a length of 5cm with a lead point at the ileocecal valve measuring 4x4x3cm with an adjacent tumor of 1.5cm. There was also note of a 4x2cm confluence of hard lymph nodes in ileocecal mesentery. A positive nodal involvement was observed out of the 2/15 pericolic lymph nodes examined. There was no peritoneal seeding. The liver and the rest of bowels appeared grossly normal. Biopsy was taken from the ileal mass and sent for histopathological studies which stained positive for CD 20, CD 3 was negative, BCL2 was negative, Ki 67 was a 100%

proliferative index and was revealed to be an atypical lymphoid proliferation favoring high grade non-hodgkin's lymphoma consistent with Burkitt's lymphoma.



A B C

Figure A. This demonstrates the normal ileal mucosa separated from that of the lymphoid proliferative cells. Figure B. This demonstrates round to slightly irregular nuclei displaying clumped chromatin pattern with several peripherally located nucleoli with noted scanty basophilic cytoplasm and mitotic figures (starry sky appearance). Figure C. This demonstrates a closer view of the neoplastic lymphoid infiltrate with intermediate to large sized cells that have round to slightly irregular nuclei displaying clumped chromatin pattern.

CEA taken was normal. Patient was then referred to an oncologist and was subsequently requested for bone marrow aspiration which revealed negative for a lymphoma cell population. Patient had no noted perioperative and post operative complications and was noted to be improving throughout the hospital course and was then subsequently discharged improved with plans for chemotherapy. Patient was eventually started on the following chemotherapeutic regimen that involved using rituximab, etoposide, doxorubicin, vincristine, prednisone, cyclophosphamide and filgrastin for a total of 6 cycles. Patient had repeated CT scan of the abdomen done every 3 months for follow up surveillance with no noted new mass lesions. A colonoscopy was done 1 year after surgery which was unremarkable that revealed normal looking colonic mucosa.

Discussion:

Burkitt's lymphoma is a highly aggressive and malignant growing B-cell neoplasm characterized by the translocation and deregulation of the *MYC* gene on chromosome 8. There are three clinical forms of recognized: endemic (African), sporadic (non-endemic), and immunodeficiency-associated. The endemic variant is found in equatorial Africa and New Guinea while the Sporadic variant is more common among Caucasians than in African or Asian Americans. In Europe, the incidence is approximately 2.2 cases per million persons per year. It represents 40%-50% of all non-hodgkin's lyphoma cases in childhood, and is more commonly seen in males than females [8]. It only constitutes 1% of all adult lymphomas. For small intestinal lymphomas, the predisposing factors includes prior malabsorption syndromes, inflammatory bowel disease, and an immunosuppressed state. It has been shown that intussusception caused by Burkitt's lymphoma presenting as an acute abdomen is rare and symptoms are nonspecific making the diagnosis to be a challenge. Some patients present with symptoms of bowel obstruction secondary to ileocecal intussusception caused by tumor growth, obstruction or bleeding, simulating acute appendicitis. Laparotomy remains the gold standard in both diagnosis and treatment with excision of the entire tumor including its appropriate margins [1].

Intussusception in adults is considered a rare condition that represents 5% of all its cases and accounts for only 1%-5% of intestinal obstructions in adults. In adults this occurs secondary to a pathologic condition that serves as a lead point that includes carcinomas, polyps, meckel's diverticulum, colonic diverticulum, strictures or benign neoplasms that are often discovered intraoperatively. Adults have lesions within the intussusception in 70% to 90% of cases [6]. Intussusception can be categorized as enteroenteric (small bowel only), colocolic (large bowel only), ileocolic (terminal ileum prolapses within the ascending colon), or ileocecal (ileocecal valve is the lead point) [7]. The clinical presentation of adult intussusception varies considerably and the classic triad of cramping abdominal pain, bloody diarrhea and a palpable tender mass is rare in adults. Signs and symptoms such as nausea, vomiting, gastrointestinal bleeding, change in bowel habits, constipation or abdominal distension may be present [5]. Abdominal pain is present in 80% of the cases of intussusception with Burkitt's lymphoma, along with nausea, vomiting, constipation, diarrhea, fatigue or malaise [8]. In the case of this patient, a 28 year old male Filipino who was apparently well with no history of recurrent diarrhea presented with acute onset of right lower quadrant pain mimicking that of acute appendicitis and on further work up was noted to have an ileocolic intussusception.

Computed tomography is the most sensitive diagnostic modality and can differentiate intussusceptions with or without a lead point. It has a reported diagnostic accuracy of 58%-100% and the characteristic features of CT scan includes a heterogeneous "target" or "sausage"- shaped soft- tissue mass with a layering effect, mesenteric vessels within the bowel lumen are also typical. A CT scan of the whole abdomen with contrast was requested for this patient which revealed a rounded soft tissue mass approximately measuring 3.5x4.2x3.6cm in the ileocecal junction and is seen as a lead point of intussusception.

Surgical intervention remains the definitive treatment of adult intussusceptions that involves exploratory laparotomy or laparoscopy followed by resection of lead point masses or areas of ischemia [7]. As the in case of this patient, exploratory laparotomy with right hemicolectomy and ileo-transverse colostomy with end to side anastomosis was done and biopsy taken from the ileal mass was consistent with Burkitt's lymphoma which was positive for the CD 20 immunostaining. A characteristic biopsy consistent with diffuse infiltration of layers of the intestine with a neoplastic lymphoid infiltrate consisting of intermediate to large sized cells, with round to slightly irregular nuclei displaying clumped chromatin pattern with one or several peripherally located nucleoli and noted scanty basophilic cytoplasm with mitotic figures thus giving a starry sky appearance along with a positive immunohistochemical stain for CD20 is used to confirm the Burkitt't lymphoma [10]. Primary malignant tumors of the small intestine are very rare and accounting for less than 2% of all gastrointestinal malignancies [11].

Burkitt lymphoma is very sensible to chemotherapy which includes the following drugs: cyclophosphamide, methotrexate, cytarabine, iphosphamide, etoposide, vincristine, vindesine, adriamycin, doxorubicin, dexamethasone [8]. CODOX-M plus IVAC ((cyclophosphamide, vincristine, doxorubicin, high-dose methotrexate) with IVAC (ifosfamide, cytarabine, etoposide, and intrathecal methotrexate) has traditionally been the combination chemotherapy regimen most frequently used for treatment. Among those diagnosed from 2002 to 2008, the estimated five-year survival decreased with age, being 87, 60, 48, and 33 percent for patients age ≤19 years, 20 to 39 years, 40 to 59 years, and ≥60 years, respectively. The prognosis of Burkitt's lymphoma depends on the site of involvement at the time of diagnosis. For this patient, he was eventually started on the following chemotherapeutic regimen that involved using rituximab, etoposide, doxorubicin, vincristine, prednisone, cyclophosphamide and filgrastin for a total of 6 cycles.

Conclusion:

A rare case of small intestinal Burkitt's lymphoma leading to an ileocolic intussusception present in a 28 years old Filipino male was reported. Predisposing factors for Burkitt's lymphoma include prior malabsorption syndromes, inflammatory bowel disease, and an immunosuppressed state. Signs and symptoms such as nausea, vomiting, gastrointestinal bleeding, change in bowel habits, constipation or abdominal distension may be present but are nonspecific. The diagnosis of this condition is made clinically with CT scan findings characteristic for an intussusception and through laparotomy with excision of the entire tumor including appropriate margins and biopsy of any suspicious mass lesions. A tissue biopsy done positive for CD 20 with atypical lymphoid proliferation favoring high grade Non-hodgkin's lymphoma is consistent with Burkitt's lymphoma. Management is mainly surgical but also involves initiating a chemotherapeutic regimen thereafter. The prognosis of Burkitt's lymphoma depends on the site of involvement at the time of diagnosis but at the age group of this patient is estimated to be 60% based on previous studies. The epidemiologic data regarding this condition is adults is limited and further studies are warranted.

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To the Joint Research and Research Education Committee:

The Case Report of Beatrice Delynn Go entitled Small Intestinal Burkitt's Lymphoma Presenting As An Acute Abdomen Secondary To Ileocolic Intussusception In A 28 Years Old Male: A Case Report has been reviewed and endorsed as deemed satisfactory for submission and presentation during the 2019 Joint Annual Convention.

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