Burkitt Lymphoma Presenting as Intussusception in an Adult: a Rare Entity

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ABSTRACT

Intussusception is uncommon among adults. Burkitt lymphoma of intestine is a rare entity. It is more common in lymphoid organs, tonsils and in midline (1). We report the case of a 28 year old man who presented as ileocolic intussusception and resection specimen was suggestive of Burkitt lymphoma. Adjuvant treatment in the form of chemotherapy regimen R-EPOCH comprising of rituximab, vincristine, cyclophosphamide, doxorubicin and oral prednisolone was given.

Keywords: Non-Hodgkin's Lymphoma, Burkitt lymphoma (BL), Intussusception, ileocaecal

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Introduction
Primary gastrointestinal lymphoma is rare, constituting 5% of all lymphomas and 1 to 4% of all malignancies of the alimentary tract. Only 1 to 2% of all primary gastrointestinal malignancies arise in the small intestine; of these, fewer than 30% are lymphomas. Common extra-nodal sites of involvement include the central nervous system, bone marrow, bowel, skin, lung, and liver (2).
Lymphoma is a rare cause of adult intussusception. It is more common in children. We present a case of intussusception in an adult where histopathology proved Burkitts type of Non-Hodgkin’s lymphoma. Although BL is considered a nodal lymphoma, the extranodal involvement is present in more than 80% of the cases (3). The extra nodal Burkitt lymphoma especially occurs in gastrointestinal tract, and head and neck areas, bone marrow, genito-urinary tract, bones, central nervous system and liver (4).

Case Report
A 28-year-old man presented with abdominal pain of 3 months duration. Pain was gradual in onset, colicky and intermittent without any associated aggravating or relieving factors. Patient had history of intermittent loose stools, no associated vomiting or fever. Clinical Examination showed an ill-defined mass in the right iliac fossa. Blood investigations were within normal limits. X-ray abdomen was normal. Ultrasound of abdomen showed pseudo kidney sign involving caecum and ascending colon suggestive of infective or infiltrative pathology. A computed tomographic (CT) scan showed whirlpool sign suggestive of ileocolic Intussusception in the right iliac fossa (Figure 1). Intraoperatively, ileocolic intussusception was seen in right iliac fossa associated with multiple lymph nodes (figure2). Right hemicolectomy with ileocolic anastomosis was done. Patient tolerated surgery well and was discharged on sixth post-operative day. Histopathological examination showed a neoplasm composed of sheets of large lymphoid cells which was suggestive of non-Hodgkin’s lymphoma. Immunohistochemistry was suggestive of burkitts lymphoma. A staging work up by PET scan was done. Report was suggestive of mild grade metabolically active adjacent mesenteric lymph nodes suspicious for lymphomatous involvement. Metabolically active omental nodule along the greater omentum in the pelvis on right side suggested lymphomatous involvement.

Discussion
Intussusception of the bowel is defined as the telescoping of a proximal segment of the gastrointestinal tract within the lumen of the adjacent segment (Figure 3). This condition is more common in children than in adults. The condition is distinct from intussusception in children, where it is usually primary and benign, and pneumatic or hydrostatic
reduction is sufficient in 80% of the cases. Most common cause of adult intussusception is polyp. But in our case, it turned out to be a Burkitt lymphoma. The B-cell-derived highly-malignant Burkitt lymphoma was first described in 1958 by Dennis Burkitt in the jaw of Ugandan children. The gamma-herpes virus Epstein-Barr (EBV) was 6 years later isolated from cells cultured from Burkitt lymphoma (5).

Primary lymphoma of the gastrointestinal tract accounts for 30–40% of all extra nodal lymphomas and constitute 10–15% of all non-Hodgkin lymphomas which are primarily in midline. More than 90% of cases of adult intussusception are due to colonic adenocarcinoma or polyps. Although rare, intussusception is a recognized presenting feature of lymphoma and the most common recognised site is the ileocolic region (5,6).

It presents with a variety of vague symptoms, thus making its preoperative clinical diagnosis difficult. The clinical presentations of adult intussusception are nonspecific such as nausea, vomiting, gastrointestinal bleeding, change in bowel habits, constipation or abdominal distension and have been reported as chronic, consistent with partial obstruction. The classic paediatric presentation of acute intussusception (a triad of cramping abdominal pain, bloody diarrhoea and a palpable tender mass) is rare in adults.

The mechanism of bowel intussusception is unknown in primary or idiopathic cases. In contrast, secondary intussusception initiates from a pathologic lesion of the bowel wall or irritant within the lumen that alters normal peristaltic activity and serves as a lead point. Variability in clinical presentation and imaging often makes diagnosis a surgical challenge. Plain abdominal films may show features of intestinal obstruction. Ultrasonography can show the classical imaging features such as the “target” or “doughnut” sign (7). Abdominal CT is considered the most sensitive imaging modality with a reported diagnostic accuracy of 58%-100% and can distinguish between intussusception with and without a lead point. The characteristic features of CT scan include an inhomogeneous “target” or “sausage”-shaped soft-tissue mass with a layering effect, mesenteric vessels within the bowel lumen are also typical (7).

Non Hodgkin’s lymphoma (Burkitt variety) occurs mainly in midline. It is important to keep a differential of malignancy, so that during surgery adequate margins and lymph node clearance can be obtained. Although malignancy is not uncommon in intussusception, Burkitt variety is uncommon and carries poor prognosis.

FIGURE 2: intraoperative picture showing intussusception.
Conclusion

Intussusception in an adult cannot be managed conservatively as in a child. Intussusception of bowel in adults is rare and is usually associated with an underlying cause.

Burkitt is a variety of non Hodgkin lymphoma, its presentation as an intussusception is a rare entity.

Although treatment for irreducible ileocolic intussusception is right hemicolectomy, keeping in mind malignancy, adequate margins and lymph node clearance should be done.

References