Non-Hodgkin’s lymphoma of the intestine in a child presenting as intussusception- A case report and review of literature

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Abstract:
Primary Non Hodgkin’s lymphoma of the intestine is more common in children. Intussusception is more common in infants aged 6 to 24 months and its incidence decreases in children. Here we presented a case of primary Non Hodgkin’s lymphoma of ileocaecal region in a 5 year old child presenting as acute abdomen due to intussusception and discusses the management options.

Key words: Gastrointestinal lymphoma; Intussusception; Intestinal obstruction; Non Hodgkin’s lymphoma; Primary intestinal tumour.

Introduction

Intussusception is a process which occurs when a proximal segment of a bowel (intussusceptum) becomes telescoped into a distal segment (intussuscipiens) pulling the associated mesentery along with it. This condition is encountered more commonly in neonate in an idiopathic form at around 6 to 24 months of age. It is believed that hyperplasia of the Payer’s patches in the terminal ileum may be the initiating factor secondary to weaning [1]. It is often associated with a preceding viral upper respiratory tract infection or gastroenteritis which resulted in hypertrophy of Payer’s patches due to viremia. In the older children true pathological lead point may be encountered, that may include lymphomas, meckel’s diverticulum, hamartomatous polyp, hemangiomas and even

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invaginated appendicular stump [2]. Herewith we present a case of Non Hodgkin’s lymphoma of ileocaecal region in a child presented as acute abdomen due to intussusception and discuss management of this and brief review of literature.

Case report

A 5 year old child presented to the emergency department with acute pain abdomen since 3 days. The pain was colicky in nature with relief for 15 to 20 minutes with occasional passing of bloody stool. On general examination, the child was afebrile with diffuse tender abdomen with mobile tender mass in the right lumbar region extending to epigastrium. Per rectal examination revealed bloody stool and apex of protruded bowel suspecting intussusception. There was no generalized lymphadenopathy or hepatosplenomegaly. On investigation, complete blood count, chest and abdomen x-ray were normal. Ultrasonography of the abdomen revealed bowel mass in the right lumbar region with lumen in lumen sign (target sign) suggesting acute intestinal obstruction due to ileocolic variety of intussusception. Since it was more than 3 days, emergency laparoscopy was planned. On table there was ileocolic intussusception leading to acute intestinal obstruction. There were some mesenteric lymph nodes were enlarged. Decompression of the intussuscepted bowel was under taken. There were no necrosis of the bowel but some friable mass was felt in the terminal ileum extending in to the caecum. Right hemicolecotomy was planned. Post operative recovery was uneventful. Histopathology of the lesion revealed fleshy mass predominantly in the terminal ileum with involvement of caecum suggestive of diffuse large B-cell Non Hodgkin’s lymphoma. Mesenteric lymph nodes were reactive hyperplasia. Resected margins were clear. Staging work up showed normal marrow cells and CT thorax. The disease was staged as primary ileocaecal Non Hodgkin’s lymphoma stage 1 according to Ann- Arbor classification and patient was referred to cancer centre for postoperative chemotherapy. There was no sign of recurrence even after 3 years of follow up.

Discussion

Primary lymphoma of the small bowel is rare, accounting for less than 2% of the gastrointestinal malignancies and 10-20% of small bowel malignancies [3]. They arise from the lymphoid tissue in the small bowel, and are more common in the ileum owing to the presence of lymphoid Payer’s patches. This primary Non Hodgkin’s lymphoma of the gastrointestinal tract is the most common site of extra nodal lymphoma [4]. Dawson’s criteria explain the small bowel lymphoma to be described as primary [5]. It includes no mediastinal or peripheral lymphadenopathy, normal white blood cell and differential counts, predominant intestinal involvement and no spleen and hepatic involvement. Our case report meets all these criteria, hence described as primary one. Among primary Non Hodgkin’s lymphoma of the intestine mucosal associated lymphoid tissue (MALT) lymphoma is most common, next is the diffuse large B-cell lymphoma. But in western population 60-80% of intestinal lymphomas are diffuse large B-cell [6].

In review of the literature, the out come of primary Non Hodgkin’s lymphoma presenting as intussusception in children, two authors opine in deferent ways. In Ein SH et al. study only 3 out of 10 children were long term survivors [7]. On the contrary Pure P. et al. reported only one death in entire series of 292 children with primary lymphoma of the intestine presenting as intussusception [8]. For localized disease surgery is the main modality of treatment, followed by adjuvant radiotherapy or chemotherapy [3]. Complete resection of the involved segment along with enlarged mesenteric lymph node is important in long term survival. Complete resection of the bowel has the main advantage of reducing complication like bowel perforation, bleeding or the tumour lysis syndrome during chemotherapy phase [9]. Debulking of the tumour has got no role in the disseminated lymphoma of intestine because it leads to more complication and delay in recovery of the patient which in turn lead to delay in initiation of chemotherapy.

Conclusion

Lymphoma is the most common malignant tumour of the intestine in children. Intussusception is more common in infants aged 6 to 24 months and its incidence decreases in children. But whenever a child above this age group presents with acute abdomen due to intussusception high index of suspicion should be there for the presence of primary lymphoma of the intestine. Complete resection of the localized disease along with involved mesenteric lymph nodes followed by adjuvant systemic chemotherapy is the main management of primary Non Hodgkin’s lymphoma of the intestine.

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FIGURE

References