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Abstract

Burkitt’s Lymphoma (BL) is an uncommon type of Non-Hodgkin’s Lymphoma. It is mainly seen in children in African and Algerian region. In India it is a rare disease. It mainly affects B lymphocytes. The commonly involved sites are either jaw or abdomen in Indian scenario. BL affecting the ileo-cecal junction is again observed uncommonly. We would like to present a rare case where a young female child has suffered from acute abdomen with features of intussusception and underwent resection of the Ileo caecal Junction mass along with ileo-ascending anastomosis, the histopathology of which showed BL.

Key words: Burkitt’s lymphoma, ileo-cecal intussusception.

Case History

Five years old Muslim female child presented in paediatric OPD with chief complaints of on and off abdominal pain since one month & abdominal distension since 2 days. On enquiry, there was history of (H/O) mild peri-umbilical colicky pain, loss of appetite and loss of weight since one month. There was no H/O similar complaints in the past. There was no H/O Tuberculosis or contact with tuberculosis. Her immunization was complete till the age. There were no bowel or bladder disturbances.

On physical examination, she was afebrile, with pulse rate of 100/min, respiratory rate was 32/min and blood pressure was 120/60 mm of Hg. Her abdomen was distended with hyperperistaltic bowel sounds and there was tenderness all over the abdomen. As the abdomen was distented and tender and the child was non cooperative, no mass could be appreciated on palpation. Per rectal examination was within normal limits.

On investigating, her complete hemogram showed Hb-11.7 gm%, White Blood Cell count -16700/cu mm with 78% neutrophil counts & Platelet Counts-156000/cu mm.

Her X-ray abdomen was taken in erect position which showed multiple air fluid levels.

On Ultrasonography of abdomen and pelvis it showed mainly mesenteric lymphadenopathy and a suspicious mass in right iliac fossa.

Patient was initially treated with conservatively with Ryle’s tube aspirations, IV antibiotics, analgesics, IV fluids with vital parameters, abdominal girth, intake-output monitoring.

Since patients symptoms were not relieved, Computed Tomogram (CT) scan of abdomen was done.

CT report showed features suggestive of ileo-cecal intussusception with bowel within bowel appearance. As there was clinical dilemma for large ileal mass, no hydrostatic reduction was attempted and a decision to perform an exploratory laparotomy was taken.

On exploratory laparotomy, after reduction of intussusception there was evidence of IC mass; multiple enlarged mesenteric lymph nodes. The resection of the affected area of Ileo-caecal Junction along with the mass was done with ilio-ascending End to End anastomosis which was done in 2 layers with absorbable suture material. Also, mesenteric lymph node biopsies were taken.
On gross and cut opened specimen showed a large polypoidal mass in caecum with dilated proximal ileum and narrow ascending colon. The mass was hard and black coloured with areas of haemorrhage and necrosis.

Her histo-pathology report showed High grade Non Hodgkin’s Lymphoma.

An Immunohistochemistry (IHC) at Tata memorial centre (TMC) confirmed the diagnosis of Burkitt’s lymphoma. As the IHC was done at different centre, we could not get the IHC image as per their protocol.

In post operative period, patient recovered well. Her Ryle’s tube was removed on 4th post operative day and sips started. Abdominal drain was removed on 8th postoperative day.

With HPR and IHC reports, a paediatric oncologist’s opinion was taken where she was started on IV hydration, Tab. Allopurinol and Tab. Prednisolone for a period of 15 days.

She was treated with pre-phase COP regime followed by MCP842 regimen.

Ki67, a marker of cellular proliferation, was 97% thus distinguishing it from diffuse large B cell.

MCP 842 protocol includes alternating cycle A and cycle B. The drugs are adriamycin , cyclophosphamide, cytarabine and ifosphamide with intrathecal IT Methotrexate.

As St . Jude (Murphy staging) it was stage III. Complete staging workup includes Bone marrow and Cerebro-Spinal Fluid [CSF] study was done before starting the chemotherapy.

Patient tolerated the chemotherapy drugs well. Now, at 2 years follow up patient is doing well.
Discussion

Burkitt's lymphoma is an uncommon type of Non-Hodgkin Lymphoma (NHL). Burkitt's lymphoma commonly affects children. Dennis Burkitt first described this entity in 1956 in equatorial Africa. It is a highly aggressive type of B-cell lymphoma that often starts and involves body parts other than lymph nodes. In spite of its fast-growing nature, Burkitt's lymphoma is often curable with modern intensive therapies.

In an Indian series of solid malignant tumours in children, Pramanik et al., in 1997, studied 263 cases over a 10-year period and found only two cases (0.76%) of BL [1,2]. Many etiologic theories have been postulated. The role of Epstein Barr Virus [EBV] in BL is not well understood. The virus may be a prime etiologic agent or a co-carcinogen. This virus preferentially infects B cells via the C3d complement receptor, CD 21. Other co-factors may include chromosomal abnormalities, immune defects, and protein energy deficits.

Burkitt's lymphoma cells contain a reciprocal chromosomal translocation, the most frequent of which is an 8q24; 14q32 translocation. Three subtypes of BL have been identified [4]:

1. Endemic Burkitt's lymphoma (EBL): It is observed in Africa. It has a peak incidence between 3-8 years of age and the male to female ratio is 2:1. The commonest site of disease presentation in EBL is the face with multiple facial bone involvement. It usually involves the maxilla and mandible [3].

2. Sporadic Burkitt's lymphoma (SBL): It is observed in western countries like America. The sporadic form affects older individuals, with a mean age of 11 years and has no gender predilection. It is more likely to have leukemic or bone marrow involvement and less likely to have jaw involvement. In SBL, the most common site of presentation is the abdomen.

3. Burkitt's lymphoma in immunocompromised patients with HIV: Burkitt's lymphoma is also known to be associated with HIV infection. Most patients are adults with marked immunosuppression. There is tumour presentation both in lymph nodes and at extra nodal sites particularly in CNS, bone marrow and gastrointestinal tract.

Levine et al (1982) classified the cases of the American BL as follows:

Stage I: single tumour mass (extra-abdominal 1A or abdominal 2A).

Stage II: two separate tumour masses on the same side of the diaphragm.

Stage III: involvement of more than two separate masses or disease on both the sides of the diaphragm.

Stage IV: pleural effusion, ascitis or involvement of the central nervous system (malignant cells in the cerebrospinal fluid) or bone marrow.

Burkitt's lymphoma can be distinguished histologically and cytologically from other forms of malignant lymphomas [5,6]. Histologic sections show an undifferentiated type of B-cell lymphoma. Immature cells 10-25 mm in diameter proliferate and have several prominent nucleoli within rounded nuclei. The sheets of tumor cells are interspersed with large pale macrophages, providing the "starry-sky" appearance, which is typical of but not unique to BL.

Immunohistochemical stains Ki-67, CD-19, CD-20, CD-22, CD-79a protein may be useful in diagnosis.

Several laboratory findings are consistently abnormal in BL patients. Serum lactate dehydrogenase (LDH) is elevated to a level corresponding to the extent of the tumour dissemination. Increased activity of alanine aminotransferase, serum alkaline phosphatase, and immunoglobulins has been reported. Anemia and leukocytosis are common. In addition, the ESR and blood urea nitrogen (BUN) may be elevated.

The treatment strategy consists of surgery followed by chemotherapy [7]. The result of chemotherapy and immunotherapy are excellent. Surgical debulking of large localized jaw or abdominal tumours is beneficial prior to chemotherapy. However there are no clear cut guidelines regarding the operation strategy and the minimum resection margins regarding our case scenario [8]. More over when situation complicates with features of intussusception again no specific surgery study is available like right hemi-colectomy versus ileo-ascending anastomosis as we performed in our case. Das DK et al emphasize the importance of FNAC for intestinal lymphoma diagnosis [9].Important is margins of resection should be clear from tumour involvement which can be confirmed on frozen section study. Cyclophosphamide 40 mg/kg in a single intravenous administration and repeated about 2 weeks later has given good results. Vincristine and methotrexate are also successful in some cases. A recent report suggests that a combination of
cyclophosphamide, vincristine, and methotrexate give better results than any drug used alone [10].

Radiotherapy produces excellent results but because of the adverse effects & complexities of immune system, it is largely not used. Good prognostic factors are younger age at diagnosis, minimal tumor burden and rapid initiation of chemotherapy, emphasizing the importance of early diagnosis and prompt treatment. Remissions occur in more than 90% of patients with high-dose alkylating agent therapy as seen in this case. Relapse occurs in two thirds of cases, most often in patients with advanced stages. With combination chemotherapy, the overall 2-year survival rate is 55% with a range of 80% for low-stage disease and 40% for the advanced stage disease.

Our case showed a complex feature of intussusception which is not observed otherwise elsewhere in patients with Burkitt lymphoma. Considering a large mass situated in ileo-caecal junction associated with intussusception we decided to go ahead with exploration rather than attempting a hydrostatic reduction for it or any other conservative measures. It was a right decision in end as the tumour mass would have never reduced to conservative reductions methods. Entire abdominal cavity was also explored and findings were confirmed.

We have given our child chemotherapy after checking her Complete Haemogram regularly to which she has responded quite well. We would like to report such a rare case of Burkitt lymphoma presenting with ileo-caecal intussusception in a small female child at our institute for which excision of tumour mass along with resection and end to end ileo-ascending anastomosis was done followed by chemotherapy because of which now the child is disease free.

References