

Case Report: Pediatric duodenal Burkitt's lymphoma

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Case Report

A seven-year-old boy was admitted with complaints of pain in the abdomen, vomiting, low grade fever, and jaundice for 10 days. There was no diarrhea, melena, hematemesis, or abdominal distension. The hemoglobin was 7.4 gm%. Total WBC count and the differential count were within normal limits. The erythrocyte sedimentation rate was 60. Total bilirubin was 5 mg%, with a direct bilirubin of 3 mg% and an indirect bilirubin of 2 mg%. SGPT was 22 IU/ml, SGOT was 170 IU/ml, and the serum alkaline phosphatase was 543 u/l (normal range: 150–250 u/l). The patient was HIV nonreactive. The chest radiograph was normal.

USG showed a hypoechoic lesion surrounding the second part of the duodenum, causing narrowing of the lumen. There was thickening of the duodenal wall: it was 2 cm thick anteriorly and 3 cm thick posteriorly [Figure 1]. The lesion was vascular [Figure 2]. No adjacent periduodenal

or peripancreatic lymphadenopathy was seen. The barium study [Figure 3] showed loss of mucosal folds in the duodenum, with preservation of the lumen.



Figure 2: Color Doppler image shows marked vascularity within the mass (arrows)

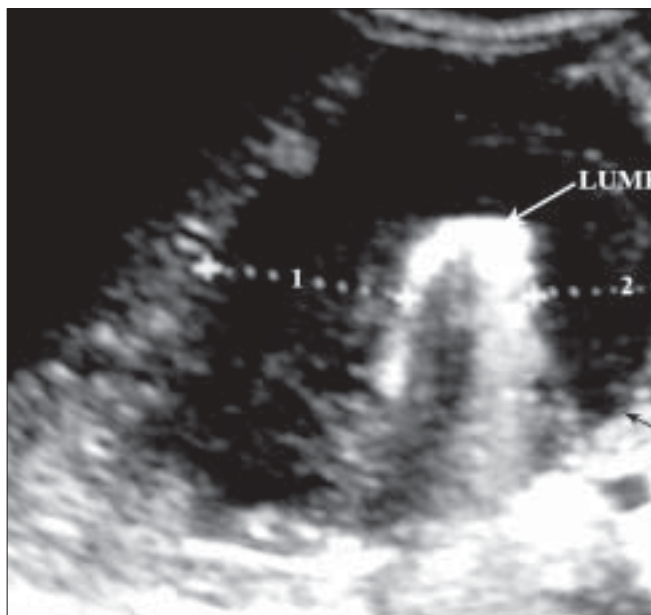


Figure 1: Transverse USG image shows a hypoechoic lesion (arrows) surrounding the second part of the duodenum, with preservation of the lumen

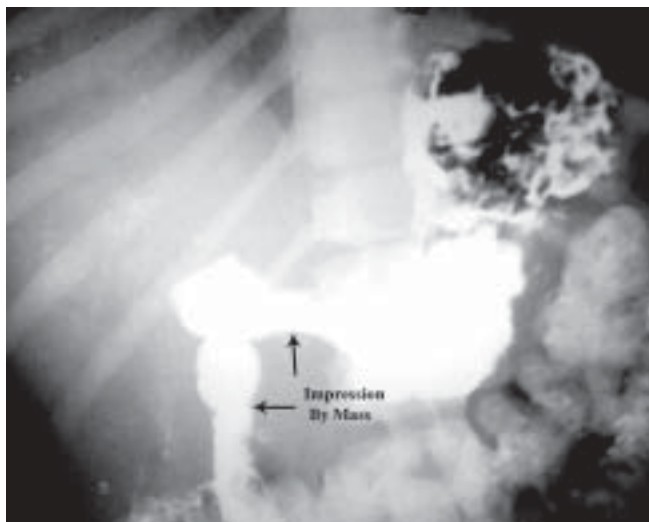


Figure 3: A barium meal image shows loss of the normal mucosal folds, with preservation of the lumen

On CT scan, there was a large, well-defined, markedly enhancing soft tissue mass involving the entire duodenum [Figures 4 and 5]; there was marked thickening of the duodenal wall, with preservation of the lumen. The CBD was obstructed by the mass, with dilatation of the intrahepatic biliary radicles. A biopsy was performed, which showed the findings of non-Hodgkin's lymphoma, specifically Burkitt's lymphoma (BL). However, the Epstein-Barr (EB) virus could not be visualized.

Discussion

Dennis Burkitt first described the small noncleaved-cell lymphomas, Burkitt's type, as a distinct clinical entity in 1958 during studies in equatorial Africa, where the tumor is endemic.^[1] It is a malignant proliferation of undifferentiated B lymphocytes that most often affects children.^[2] There is

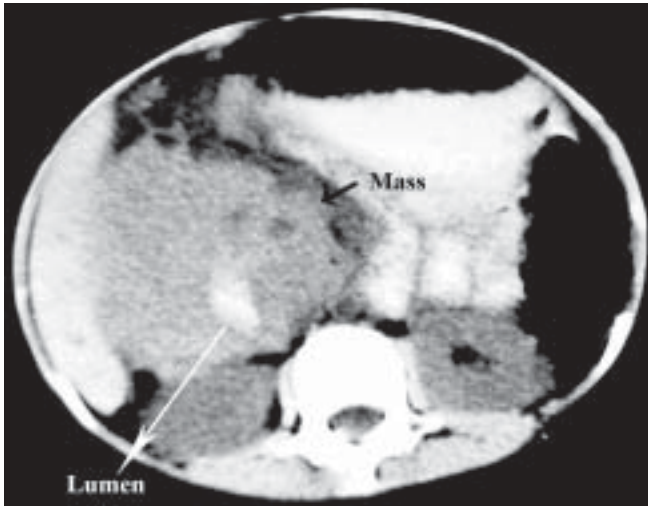


Figure 4: Plain CT of the abdomen with oral contrast shows a large, well-defined, soft tissue mass (arrow), with preservation of lumen (long arrow)

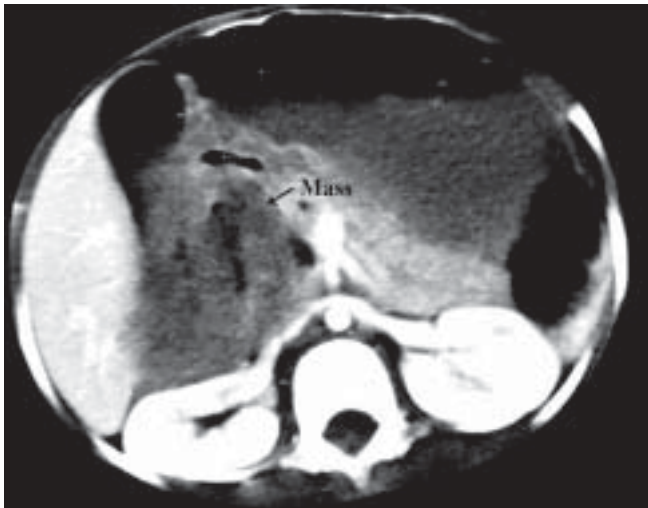


Figure 5: Contrast-enhanced CT of the abdomen shows an enhancing soft tissue mass (arrow) with preservation of the lumen

strong evidence linking BL with the EB virus.^[3] The three distinct clinical forms of Burkitt's lymphoma include the endemic, sporadic, and immunodeficiency-associated types.^[4] Though it is endemic in Africa, it also occurs throughout the world, accounting for 40 to 50% of childhood non-Hodgkin's lymphomas in nonendemic areas.^[1] Cases occurring outside Africa are histopathologically indistinguishable from those occurring in endemic areas.^[1] Cases in endemic areas have a high propensity for involvement of the bones of the face, particularly the jaw, maxilla, and orbit, especially in young children. Involvement of these sites is unusual in nonendemic areas.^[1]

BL is uncommon in the gastrointestinal tract.^[5] The sites of involvement include the duodenum, cecum, ascending colon and jejunum; in the pelvis, the ovaries may also be involved. Because of the paucity of lymphoid tissue in the duodenum, primary duodenal lymphoma is a rare entity, accounting for less than 5% of all small bowel lymphomas.^[6]

BL is a tumor of B cells that express surface IgM and Pan-B-cell markers such as CD-19 as well as CD-10.^[7] It can be diagnosed morphologically with a high degree of accuracy. The cells are homogenous in size and shape. Demonstration of a very high proliferative fraction and presence of t(8;14) or one of its variants, t(2;8) (c-myc and the gamma light chain gene) or t(8;22) (c-myc and the k light chain gene) can be confirmatory.^[4]

Patients present with peripheral lymphadenopathy or intraabdominal masses. The disease is typically rapidly progressive, with a propensity for central nervous system (CNS) metastases. The initial evaluation should, therefore, always include CSF examination to rule out metastases.^[4] Typical clinical features include colicky or persistent abdominal pain, weight loss, and fever. In about 1/5th of the patients, there is a mass. Some patients have associated protein loss, which may lead to generalized edema but not to a malabsorption syndrome.^[8] Sometimes, the symptoms are nonspecific and may have been present for many years before a diagnosis is established.^[9] Other symptoms include perforation or fistula formation, obstructive symptoms, GI bleeding and, very rarely, jaundice due to obstruction of the CBD.^[6,8,10]

BL begins submucosally, presumably in lymph follicles or Peyer's patches, and grows submucosally as a soft, diffuse, infiltrating lesion that leaves the mucosa intact in the early stages.^[8] In contrast to a carcinoma that encircles and obstructs the bowel, it tends to expand longitudinally along the bowel. Later, ulceration of the mucosa may occur. Regardless of the extent of the lesion or its appearance, the involved bowel remains strikingly pliable, so that a lesion that looks as if it ought to be obstructing the entire bowel may not particularly trouble the patient.^[8]

Common CT scan findings include a thickened bowel wall, which is sometimes nodular and either diffuse or focal, with discrete mesenteric lymph node masses. It is often possible to see a thickened but otherwise preserved pattern of valvulae conniventes at one or the other margin of the infiltrated segment. Unlike primary adenocarcinoma and metastatic disease, extensive lymphomatous encasement of the bowel may occur without significant narrowing.^[11] Rarely, calcification can be seen on CT done prior to therapy.^[12] Similar findings can be seen on MRI studies in patients with small-intestinal lymphomas. The tumor masses have homogenous, intermediate signal intensity on T1W images and heterogeneously increased signal intensity on T2W images.

USG shows small submucosal nodules, which may be easily overlooked. Many patients have large, very hypoechoic, and ulcerated bowel masses. Linear high-amplitude echoes with ring-down artifacts, indicating gas in the residual lumen or ulceration, are frequently seen. Regional lymph node enlargement may be visualized.^[13]

BL is treated with high doses of chemotherapeutic agents, and CNS prophylaxis is done with methotrexate. The cure rate is 60%.^[7]

References

1. Link MP, Donaldson SS. The lymphomas and Lymphadenopathy, chap-37, Hematology of Infancy and childhood, vol-2, Nathan and Oski, editors. 4th ed, W.B. Saunders Company: p. 1324.
2. Hupp JR, Collins FJ, Ross A, Myall RW. A review of Burkitt's lymphoma: Importance of radiographic diagnosis. *J Maxillofac Surg* 1982;10:240-5.
3. Harsh Mohan. The lymphoid system, chap-13, Textbook of Pathology, 4th ed, Jaypee: p. 419.
4. Armitage JO, Lango DL. Malignancies of lymphoid cells-chap-112, Harrison's Principles of Internal Medicine, 15th ed, Vol-1. Braunwald Fauci, Kasper, Hauser, Longo, Jameson, editors. McGraw Hill: p. 723.
5. Muchmore JH, Haddad CG, Goldwag S. Primary Non-Hodgkin's lymphoma of the Duodenum, *The American Surgeon*, vol-60, Southern Surgical Congress: Atlanta; 1994. p. 12,924-8.
6. Basavaraj Mundasad MJ. Hawe- Lymphoma presenting as a bleeding duodenal ulcer: A case report. *Int J Trop Med* 2006;3:1.
7. Burkitt's lymphoma. The haemopoietic and lymphoid systems, Chap-12. Kumar, Cotran, editors. Robbins- Basic Pathology, 6th ed. A Prism Indian Edition. p. 368.
8. Spiro HM. Other Steatorrheas-clinical gastroenterology, Chap-26, 4th ed, McGraw Hill: p. 471.
9. Smith C, Kubicka RA, Thomas CR Jr. Non-Hodgkin lymphoma of the gastrointestinal tract. *Radiographics* 1992;12:887-99.
10. Kessel B, Moissier B, Korin A, Soimu U, Ashkenazi I, Asquenasi A, et al. Burkitt's lymphoma: An unusual cause of obstructive jaundice. *Asian J Surg* 2005;28:301-4.
11. Robert E Koehler, David S Memel, Robert J Stanley. Gastrointestinal tract- computed body tomography with MRI correlation. Joseph KT, Lee, Sagel SS, Stanley RJ, Heiken JP. chap-11, vol-1, 3rd ed. Lippincott Williams and Wilkins: p. 660.
12. Petit, M Panuel, C Scheiner, P Derred. Calcifications in untreated primary duodenal lymphoma: An exceptional entity. *Pediatr Radiol* 1994;24:283-4.
13. Wilson SR. The gastrointestinal tract, chap-8, Diagnostic ultrasound. Rumack CM, Wilson SR, Charboneau WJ. vol-1, 2nd ed. Mosby.

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