A Rare Presentation of Childhood Burkitt`s Lymphoma with Tense Ascites, Massive Pleural Effusion and Abdominal Mass

Mehri Sirous and Azadeh Ebrahimi*

Department of Radiology, School of Medicine, Alzahra Hospital, Isfahan University of Medical Sciences, Isfahan, Iran

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Abstract- Burkitt's lymphoma is a lymphatic tumor classified as non-Hodgkin lymphoma with small non cleaved cells. The disease has an incidence of two cases per million patients in North America, and usually present with an intra-abdominal tumor, or enlarged cervical lymph nodes. Massive amounts of peritoneal and plural fluid is rarely seen in these patients. A three year-old girl presented with massive pleural effusion, ascites, mesenteric infiltration, and intraperitoneal mass is discussed in this report. Burkitt's lymphoma is a highly aggressive tumor with a very rapid growth and a variety of different presentations. It is potentially curable, if highly intensive multidrug chemotherapy is used with aggressive central nervous system prophylaxis.

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Key words: Burkitt lymphoma; pleural effusion; ascitic fluid

Introduction

Lymphoma is the third most common childhood cancer, accounting for 10-15 percent of newly diagnosed cancers in children (1, 2). Non-Hodgkin's lymphoma accounts for 60% of the pediatric lymphomas and Burkitt`s lymphoma is the most common subtype (3). The incidence of Burkitt`s lymphoma is two cases per million in North America (the sporadic form) and up to 100 cases per million children, in Africa in equatorial regions (the endemic form) (4).

Burkitt's lymphoma is a B-cell neoplasm composed of monomorphic, medium-sized cells with basophilic cytoplasm and a very high proliferation rate (5, 6). The Real classification has two categories including Burkitt lymphoma, and Burkitt-like lymphoma (a morphologic variant of Burkitt lymphoma) (5). Burkitt lymphoma is characterized by a colonal proliferation of mature B-cells, and there is a chromosomal translocation between chromosome 8 and one of three chromosomes 2, 22 and 14 that leads to expression of a protoncogene (7). Development of the disease has been confirmed to be etiologically linked to EBV especially in endemic form (8). Burkitt's lymphoma is seen in all age groups, and males are the majority of patients (3 to 4:1) (9).

Three distinct clinical features of Burkitt's lymphoma can be recognized: endemic, sporadic, and immunodeficiency-associated (3, 10). The endemic or African form presents as a jaw or facial bone tumor with invasion to extra nodal sites including mesentery, ovary, testis, kidney, breast, and especially to the bone marrow and meninges (3, 11). The sporadic or American form has an abdominal presentation frequently with massive disease and ascites, involving distal ileum, stomach, cecum and mesentery, kidney, testis, ovary, breast, bone marrow, or central nervous system (3, 9, 11, 12). Immunodeficiency-related cases mostly involve lymph nodes; this form and sporadic cases may present as acute leukemia (13-16). There is almost always elevated blood LDH and uric acid, indicating rapid cell turnover (17).

Case Presentation

Our case was a previously healthy three year-old female, presented with a two week history of persistent periumbilical abdominal pain that was not associated with meals. Diarrhea and non bilious vomiting were added to her symptoms later, as well as one episode of melena and hematemesis. Dyspnea and malaise were also associated with her symptoms. The young patient was disoriented at the time of admission, and respiratory distress and confusional state were superimposed later. She did not have any history of NSAIDs ingestion, weight loss or fever. No histories of excessive bleeding or gastrointestinal and liver disease were reported by her parents.

***Corresponding Author:** Azadeh Ebrahimi
Department of Radiology, Al-Zahra Hospital, Isfahan University of Medical Sciences, Isfahan, Iran. Postal Code: 81746
Tel.: +98 913 1097283, Fax: +98 311 6662735, E-mail: azadehebr@yahoo.com

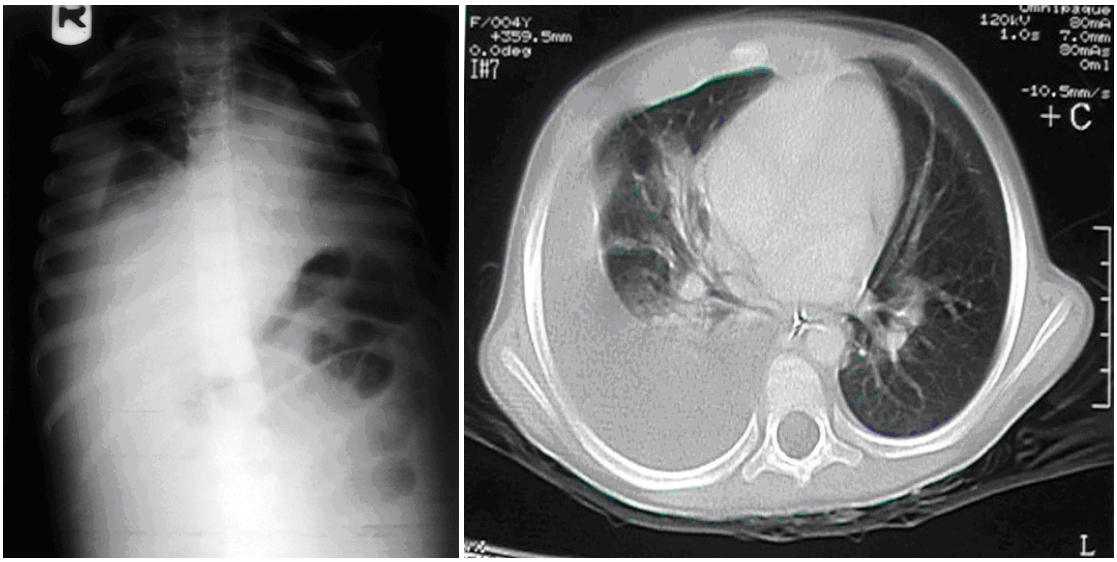

Figure 1. CXR and Chest CT scan revealed right side pleural effusion and right lung collapse

Upon physical examination, the patient was pale, afebrile and hemodynamically stable. Her abdomen was soft with no rebound tenderness or guarding. It was distended and tense ascites was present. There was no lymphoadenopathy or hepatosplenomegaly, but an epigastric mass and a rectal mass (found during rectal exam) approximately 4cm above the anus were palpable.

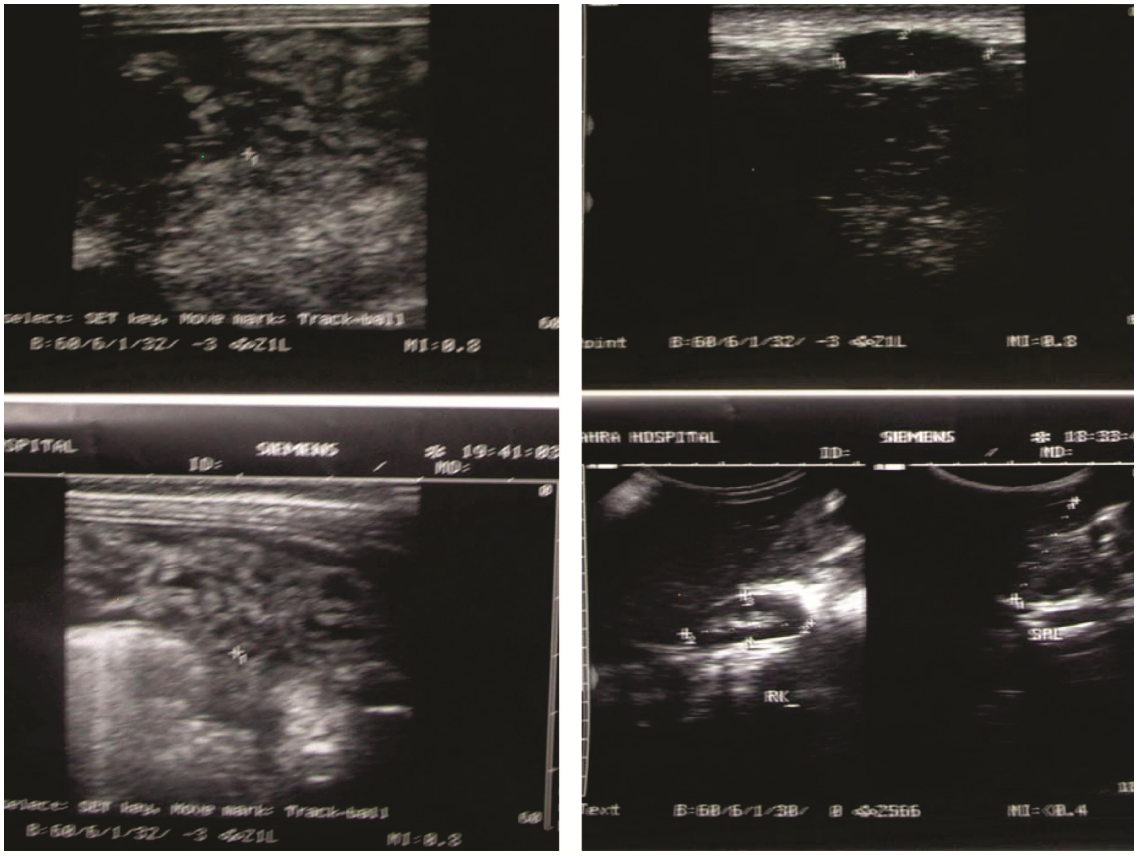

Figure 2. Sonogrphy; A mass with mixed echogenicity was seen that extended from lower border of the liver to lower portion of spleen. Mesenteric and small bowel involvement was also noted. Rectal wall was thickened and rectum was filled with fluid

A complete blood count confirmed leukocytosis with 12690 cc/dl, hemoglobin concentration of 9.5 g/dl, normal platelet count and normal cell differentiation. In addition, serum blood urea nitrogen (BUN), creatinine (Cr) and lactate dehydrogenase (LDH) concentration were markedly increased. Radiologic evaluation by chest x-ray revealed bilateral pleural effusion, predominantly on the right side (Figure 1). Ultrasonography revealed a markedly enlarged mass with mixed echogenicity extending from the lower border of liver to the lower portion of spleen as well as massive abdominal ascites (Figure 2). Spiral abdominal-pelvic CT scan with oral and intravenous contrast revealed an intraperitoneal mass that was 6 × 8 × 7 cm in size and a necrotic center. Mesenteric and small bowel involvements were also evident. The rectal wall was thickened and the rectum was filled with fluid (Figure 3).

In cytolopathologic study of pleural and peritoneal fluid, infiltration of immature lymphoid cells with fine chromatin was observed in a bloody background. The cells contained round nuclei and basophilic cytoplasm. Karyorrhexis, Cytoplasmic lipid vacuoles and cellular debris were also evident. Immunohistochemical staining of the biopsied rectal lesion was suggestive of Burkitt's lymphoma including diffuse infiltration of atypical lymphoid cells with numerous mitoses and medium-sized, round, and uniform nuclei.

Serum peritoneal dialysis was performed for the patient due to increased BUN and Cr and acute renal failure. During dialysis her blood pressure decreased and cardiopulmonary arrest occurred. Resuscitation was unsuccessfully performed, and the patient expired.

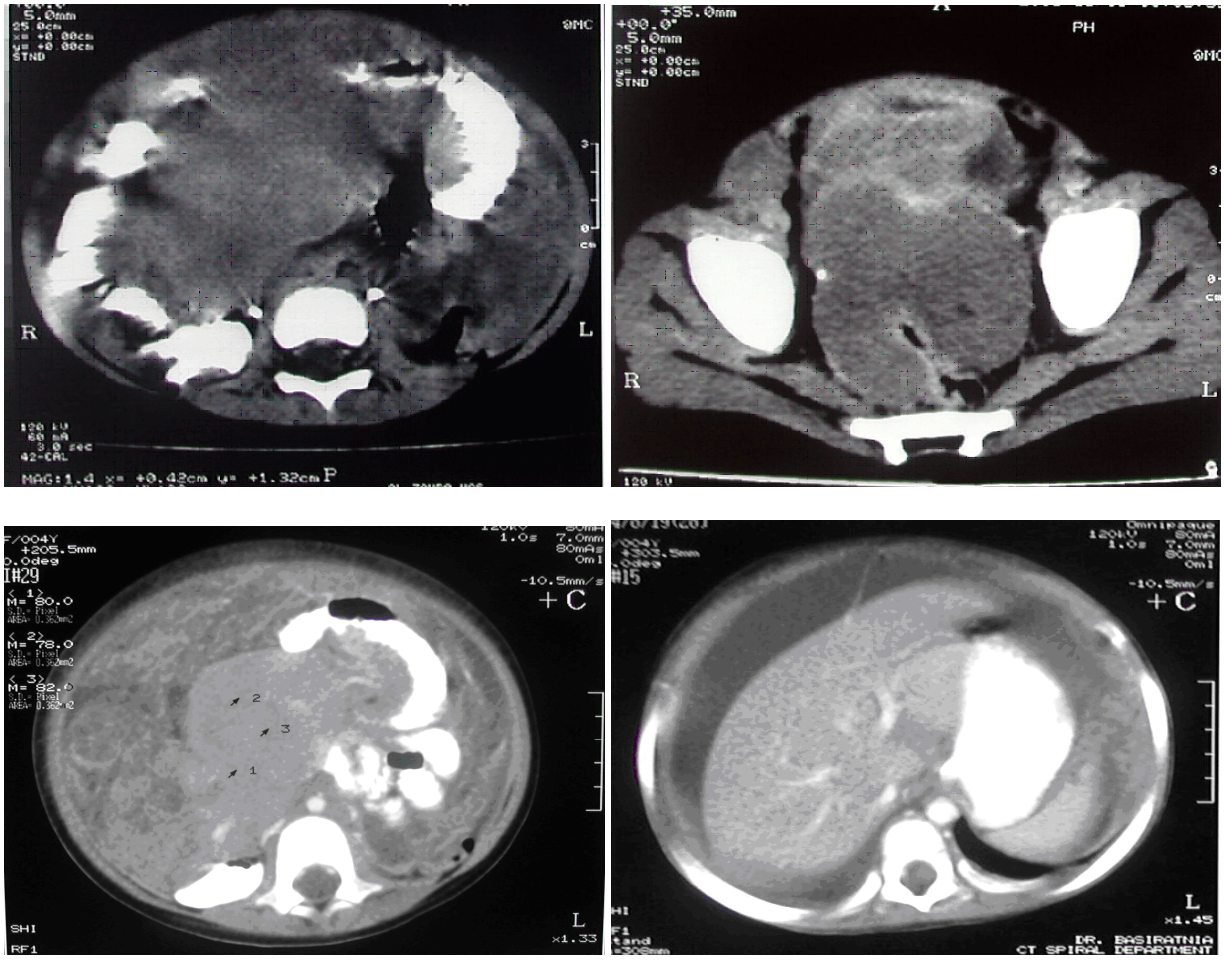

Figure 3. Spiral abdominopelvic CT scan with oral and IV contrast revealed an intraperitoneal mass approximately **6** x **8** x **7**cm in size with necrotic center, causing displacement of bowel loops. Large amount of ascites and pleural effusion was noted. Involvement of small bowel and mesenteric infiltration and rectal wall thickening were observed too

Discussion

Sporadic Burkitt's lymphoma occurs worldwide including cases with no specific geographic or climatic association. Our case was from the Middle East, which is not geographically associated with the endemic form of the disease. The most common site of involvement is abdomen, especially the ileocecal area in this variety.[4] The kidneys, ovaries, Omentum and Waldeyer's ring, are other sites of involvement (3, 9, 11, 12).

In our case, an abdominal mass, which is a typical presentation, was apparent. The omentom, small bowel and rectum were also involved. Lymph node involvement is more common among adults than children (9), and in our three year-old patient there was no lymphoadenopathy or hepatosplenomegaly evident. Patients may also have malignant pleural effusions or ascites (9, 11). Massive amounts of peritoneal and plural fluid are rarely seen in a patient, but were presenting symptoms in our case (9, 11). Rarely, patients present a disease that is primarily leukemic (classified as acute lymphoblastic leukemia). Neoplastic cells are EBV positive in 15%–30% of cases, or fewer in some series (12). Bilateral breast involvements may occur in puberty or with lactation (11).

Burkitt's lymphoma cells are generally monomorphic, medium-sized cells with round nuclei, and a basophilic cytoplasm (18). Cytologically, the cells are the small non-cleaved cells within normal germinal centers of the secondary lymphoid follicle; These cells differ from lymphoblastic lymphoma cells which have intermediate sized non-convoluted nuclei with coarse chromatin, and the cells have more abundant cytoplasm (18). The cytologic study of our case exhibited the same findings.

Serum BUN and Cr was increased in our patient due to Tumor Lysis Syndrome , a result of high rate of spontaneous cell death in the disease which can lead to potentially fatal electrolyte imbalances like hyperkalemia, hyperuricemia, hyperphosphatemia and hypocalcemia. Oliguric renal failure can occur as a result of uric acid nephropathy (4).

Melena and anemia which were evident in our case were previously reported as a rare presentation due to ulcerated lymphomatic masses in gastrointestinal tract (4). In conclusion, although Burkitt's lymphoma is a highly aggressive tumor with a very rapid growth a variety of different presentations, it is potentially curable, if highly intensive multidrug chemotherapy is used with aggressive central nervous system prophylaxis (3,19). Vigilant precautions against tumor lysis during aggressive therapy always should be taken into account.

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