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Imaging of Burkitt lymphoma in pediatric patients

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Abstract. The imaging procedures utilized at presentation in the diagnostic work-up of 19 children with Burkitt lymphoma were reviewed. The distribution of disease was compared to other tumors of childhood so that the most valuable modalities could be identified. Burkitt lymphoma is a rapidly growing tumor in the child, making it essential to suggest the diagnosis as quickly as possible so that biopsy and treatment can be instigated. The primary area of involvement was abdominal (15 of 19), gastrointestinal, intraperitoneal adenopathy, hepatic and pancreatic without retroperitoneal adenopathy. Pleural effusions were common without hilar and mediastinal adenopathy. This is in contrast to other tumors of childhood where mediastinal and hilar disease in the chest and retroperitoneal node involvement in the abdomen are common. Thus sonography is an excellent imaging modality, easily identifying the extent of the disease and so suggesting the diagnosis.

Burkitt lymphoma is one of the fastest growing tumors with a potential tumor-cell doubling time of less than 24 h [1]. It is therefore extremely important to suggest the diagnosis early in the presentation, so that treatment can be initiated as soon as possible. Clinically, Burkitt lymphoma can masquerade as many disease complexes, making the diagnosis difficult. A variety of radiologic procedures for diagnosis and staging have been described in the literature, depending on the mode of presentation of the disease [1–8]. Much of this literature has compared American Burkitt lymphoma with African Burkitt lymphoma. However, in the North American child presenting with a solid tumor, the differential diagnosis includes other lymphomas, neuroblastoma, and the soft tissue sarcomas. An analysis of the presentation in Burkitt lymphoma has been contrasted to other lymphomas, neuroblastoma, and soft tissue sarcomas. The purpose was to identify which modalities were most useful in achieving a rapid diagnosis and in staging of the disease.

Materials and methods

During the 10-year period from 1972 until 1982, 19 children with biopsy proven Burkitt lymphoma were admitted to The University of Michigan Medical Center. There were 17 boys and two girls, all were Caucasian. The age at presentation ranged from four to 20 years with an average of 10.7 years and a mean of 11.7 years. The radiographic studies at presentation were reviewed and correlated with the clinical constellation of findings.

Results

Clinical findings

The most common presenting symptoms was abdominal mass followed by abdominal pain, weight loss and malaise, vomiting or diarrhea. Tumor distribution at onset of the disease most frequently involved the abdomen (15 of 19 patients) followed by chest (6 of 19 patients) and then osseous involvement (5 of 19 patients). In several patients, more than one area was involved. Nasopharynx, neck, and ovarian involvement were found at onset of the disease in one patient each.

Radiographic findings

Gastrointestinal. Six of the nine patients with barium examinations had abnormalities including: nodular submucosal infiltration of the gastric wall (two patients), a paracecal mass with involvement of the terminal ileum (one patient) (Fig. 1a), encasement of the bowel (three patients) (Fig. 1b) and perforation of small bowel with intra-abdominal abscess formation (one patient).

Nine of the ten patients with sonographic examination of their abdomens, had findings related to the gastrointestinal tract including: multiple sonolucent masses in the liver (three patients)

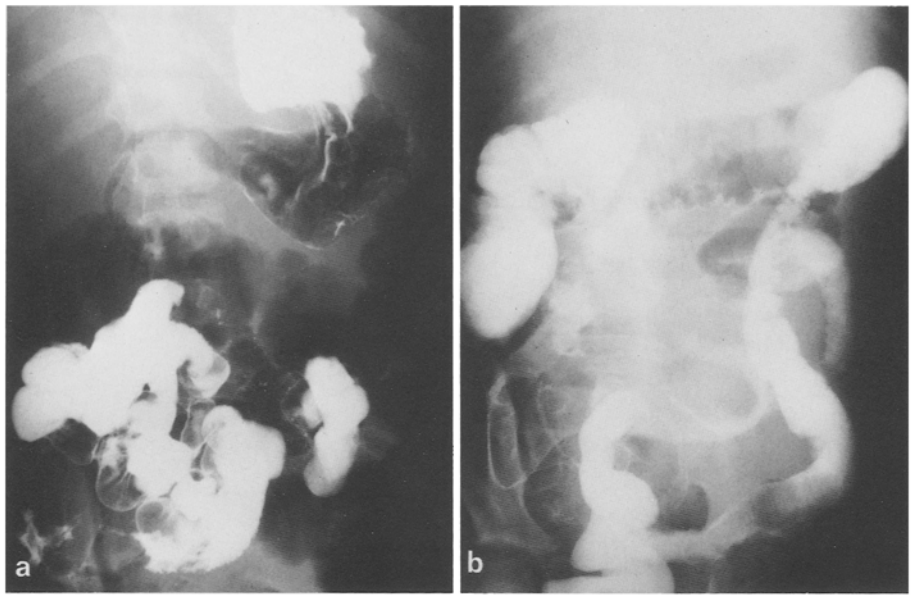

Fig. 1. **a** 11-year-old boy with Burkitt lymphoma shows nodular submucosal infiltration of stomach and a paracaecal mass with narrowed terminal ileum on upper gastrointestinal and small bowel exam. **b** 12-year-old boy presented with abdominal mass and ascites. Barium enema reveals encasement and separation of small and large bowel

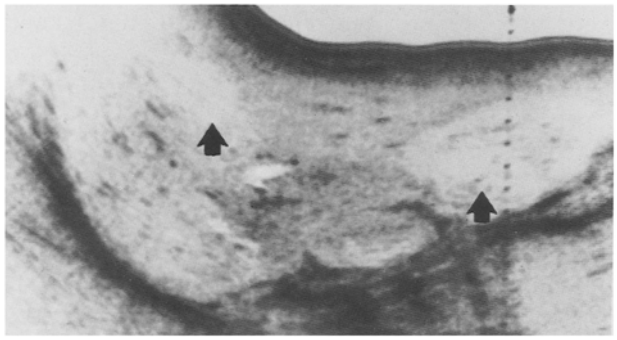

Fig. 2. The longitudinal sonogram through the liver in this 20-year-old youth reveals several sonolucent lesions in the hepatic parenchyma (arrows)

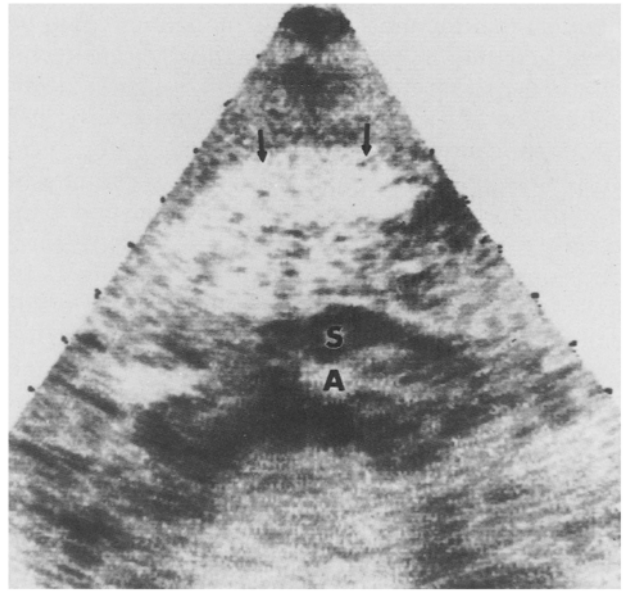

Fig. 3. This 3-year-old boy on sonography demonstrated a diffusely enlarged pancreas (arrows). S = superior mesenteric artery. A = aorta

(Fig. 2), dilatation of the biliary tree secondary to masses within the porta hepatis (two patients), diffuse homogeneous enlargement of the pancreas (two patients) (Fig. 3), sonolucent masses in the spleen (one patient), and mesenteric masses (seven patients) (Fig. 4). Two additional patients had evidence of nodular encasement of bowel on sonography (Fig. 5) and three patients had ascites. Computed axial tomography (CT) in three of these nine patients with abnormal ultrasounds revealed similar findings except the ascites was missed on CT in one case. Both CT and the barium studies were limited in delineation of the extent of the disease in some cases from inadequate contrast preparation, because the oral contrast was vomited or a level of obstruction was demonstrated beyond which no barium passed. Thus, distal additional areas of disease were not detected on these studies which were identified on sonography.

Genitourinary. Intravenous pyelograms were abnormal in eight of the 16 patients studied. The most common abnormalities were obstructive uropathy due to abdominopelvic tumors (four patients) and renomegaly (three patients). Bilateral renomegaly with focal areas of decreased density in the renal parenchyma were shown in one patient on CT (Fig. 6). Sonography also identified renomegaly (one patient) and hydronephrosis (two patients). The renomegaly was a diffuse increase in size of the kidneys without general or focal abnormalities in echogenicity.

Skeletal. Skeletal surveys were carried out in 13 patients and three had abnormalities demonstrated on the plain films. A permeative pattern in the diaphysis of the long bones with associated periosteal reaction was identified in two patients. A very well-circumscribed radiolucent defect in the mandible was seen in one patient. Two additional patients had positive radionuclide scans. All these patients had clinical evidence of osseous disease.

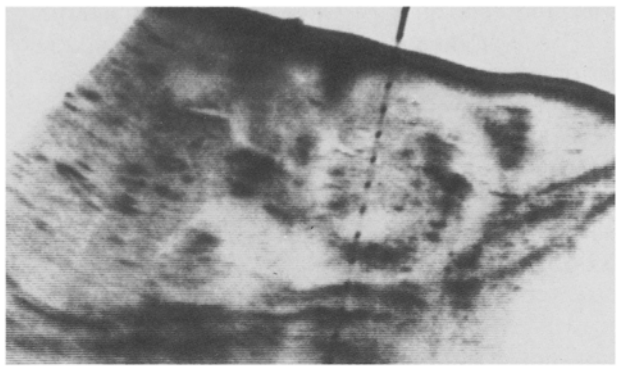

Fig. 4. Fixed mesenteric masses are demonstrated anterior to the right kidney on the longitudinal sonogram in this 13-year-old boy

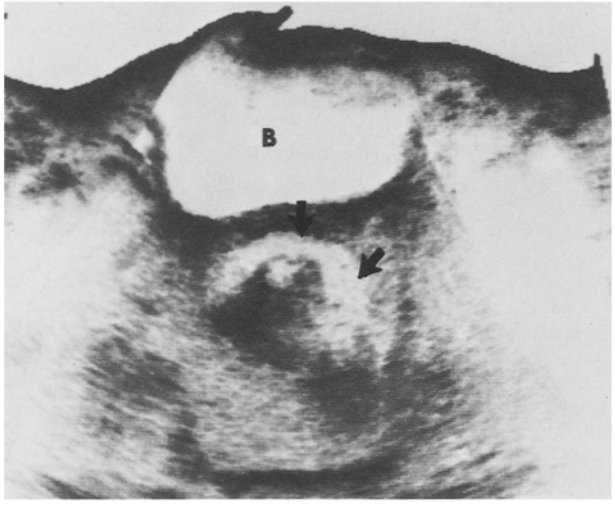

Fig. 5. A fixed pelvic mass (arrows) is identified on this transverse pelvic sonogram encasing a loop of bowel that did not change with time or water enema. B = bladder

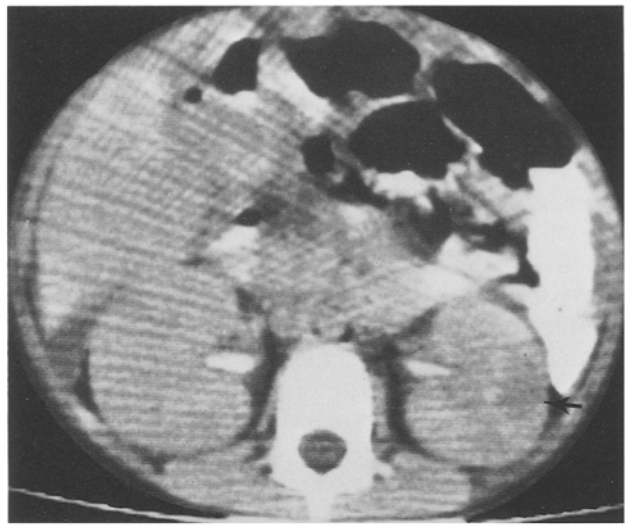

Fig. 6. A single area of lucency (arrow) within the left kidney in the CT scan in this 6-year-old with Burkitt lymphoma, thought to be Burkitt lymphoma infiltration

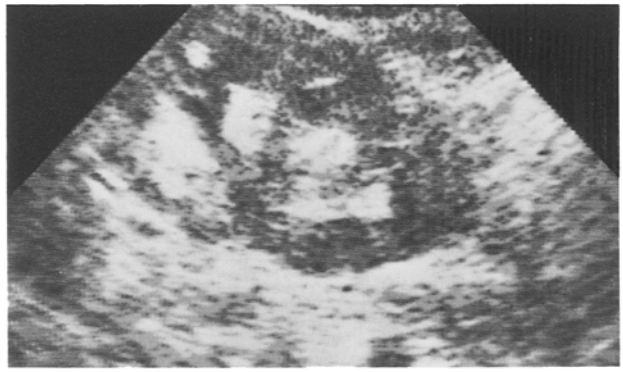

Fig. 7. The longitudinal real-time renal sonogram in this 6-year-old reveals hydronephrosis which was secondary to a large pelvic mass

Chest. Six of the 19 patients had abnormal chest radiographs at presentation including pleural effusion (five patients) and an infiltrative parenchymal pulmonary opacity associated with a left paraspinal mass (one patient). No patient in this series had hilar or mediastinal adenopathy.

Miscellaneous. Two patients had head and neck involvement manifesting in one as a nasopharyngeal mass extending into the left maxillary sinus and in the other as a peritonsillar mass which presented with swelling of the neck. The patient with primary ovarian involvement with Burkitt lymphoma did not have an ultrasound or computed axial tomography prior to surgery.

Discussion

The various imaging modalities have different limitations and contributions in suggesting the diagnosis of Burkitt lymphoma and delineating the extent of the disease. Rigorous identification of every tumor deposit is felt by some physicians to be unnecessary as systemic chemotherapy is the treatment of choice.

In our series of Burkitt lymphoma, pleural effusions were relatively common at presentation and mediastinal and hilar adenopathy were not seen. This high incidence of pleural effusions and no evidence of mediastinal and hilar adenopathy corresponds with previous reports, though one study of 40 patients found three with hilar and mediastinal adenopathy at presentation [8, 9]. The other lymphomas have a high incidence of mediastinal adenopathy, though other solid tumors of childhood do not usually present with either mediastinal or hilar adenopathy or pleural effusion.

The children with Burkitt lymphoma in this series had an overwhelming involvement of the abdomen

(15 of 19 patients). The areas of disease were primarily the gastrointestinal tract with no predilection to a particular area, and intraperitoneal adenopathy. This distribution of tumor involvement has been confirmed by others [2, 8, 9]. In contrast, the primary areas of involvement in the abdomen in other lymphomas are the retroperitoneal nodes. About 30% of all patients with other lymphomas will have positive lymphangiographic findings because of the involvement of the retroperitoneal nodes [10]. The gastrointestinal tract can be involved in other lymphomas, particularly non-Hodgkin lymphoma, the most common locations being the distal ileum and the right colon. The soft tissue sarcomas and neuroblastoma are primarily retroperitoneal tumors in the abdomen [11–13]. Ascites is not a common association with either the soft tissue sarcomas or neuroblastoma but was seen in three of our Burkitt patients. Thus the combination of intraperitoneal disease with gastrointestinal involvement with or without ascites is primarily associated with Burkitt lymphoma and not the other solid tumors of childhood.

Both liver and pancreatic Burkitt lymphoma were seen in this series of patients. Though liver involvement is seen in neuroblastoma and other soft tissue sarcomas, pancreatic disease is unusual but found in increasing incidence at autopsy in Burkitt lymphoma [14]. Careful real-time sonography is ideal in the pediatric age group to evaluate the pancreas and liver. The renal abnormalities described in this series of patients are not specific for any particular tumor. The abnormalities described on intravenous pyelography of renomegaly and obstructive uropathy can be demonstrated by ultrasound. Moreover, intravenous pyelograms often prove to be long and suboptimal studies due to poor function by uremic or obstructed kidneys.

All patients with positive skeletal surveys had clinically suspected skeletal disease since they had focal bone pain. The skeletal lesions in this series were disparate, being in one case a well-demarcated radiolucent lesion and in two others a diffuse permeative pattern in diaphysis of long bones associated with periosteal reaction. This latter pattern of bone involvement has been described in neuroblastoma and leukemia [13, 15] and in soft tissue sarcomas [11].

In our series of Burkitt patients, the combination of intraperitoneal adenopathy and gastrointestinal involvement with or without ascites but sparing of the retroperitoneal nodes should suggest the diagnosis of Burkitt lymphoma. Pleural effusion without mediastinal or hilar adenopathy are commonly associated features. Though CT, particularly the new rapid scanning units, may prove to be extremely valuable in diagnosing the extent of the disease, ultrasound appears to be an excellent imaging modality. The liver, pancreas, kidneys, adenopathy, mesenteric masses, and ovaries can be easily examined in this single study. Pleural effusion can also be detected. Careful real-time sonography avoids much of the interference of bowel gas so that a complete abdominal study can be obtained even in the very ill child. Sonography is often one of the initial screening procedures in children with suspected abdominal disease and it is extremely important in this rapidly growing tumor that the unusual distribution of disease suggest to the radiologist the diagnosis of Burkitt lymphoma.

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