

A case of pancreatic Burkitt lymphoma: radiological findings

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ABSTRACT

Burkitt lymphoma predominantly involving the pancreas is very rare. There is no specific clinical, laboratory, or radiological sign of the disease. Thus, the diagnosis, especially of focal involvement, is usually difficult. We here report ultrasonography, computed tomography and magnetic resonance imaging findings of primary pancreatic Burkitt lymphoma in a case with unusual radiological presentation.

Key words: • pancreas • Burkitt, lymphoma • ultrasonography • computed tomography • magnetic resonance imaging

Lymphoma predominantly involving the pancreas is rare; the incidence of malignant lymphoma of the pancreas in extranodal disease is estimated to be only 0.6% (1). Since the clinical symptoms and signs resemble those of pancreatic adenocarcinoma, primary diagnosis of pancreatic lymphoma is generally difficult. The prognosis of patients with advanced stage malignant lymphoma is poor; however, chemotherapy may be more effective than it is for pancreatic adenocarcinoma.

Burkitt lymphoma/Burkitt cell leukemia is a B-cell neoplasm which often presents with extranodal disease and occurs most often in children (endemic, sporadic) and immunocompromised hosts. Endemic cases, usually from Africa, involve the facial bones or jaw of children, mostly containing EBV genomes (2). It may also involve other structures, including the liver, adrenal glands, stomach, intestine, pancreas, salivary glands, thyroid, testicles and the heart. Histologically, it is a poorly differentiated lymphocytic lymphoma. Macrophages with abundant clear cytoplasm are usually scattered uniformly throughout the tumor resulting in the “starry sky” effect (3).

Radiological findings of Burkitt lymphoma in the abdomen and pelvis have been reported; however, findings of pancreatic involvement of Burkitt lymphoma by magnetic resonance imaging (MRI) have not been clearly described (4). We here report on a case with unusual radiologic imaging (ultrasonography [US], computed tomography [CT], and MRI findings) of primary pancreatic Burkitt lymphoma.

Case report

A 20-year-old man presented with weight loss, back pain, mandible numbness, night sweats, and poor exercise tolerance. In his history, no remarkable finding was noted other than tonsillitis three months previously. Physical examination findings were normal. Cranium and chest radiographs were normal. The laboratory assessment revealed mild normocytic normochromic anemia and very high lactate dehydrogenase levels. Other laboratory findings were normal.

Abdominal US did not reveal any organ or lymph node involvement other than a solitary lesion in the pancreas. High definition zoom view of the pancreatic lesion showed a 28 × 30 mm nodular lesion with low echogenicity in the body of the pancreas which was partially well rounded and had septations (Fig. 1). Thoracoabdominal CT images without and with contrast did not show any organ or lymph node involvement but the solitary lesion in pancreas. Abdominal CT confirmed the lesion as a homogeneous convexity posterior to the stomach surface that could not be distinguished from pancreatic parenchyma. On contrast-enhanced CT images, the lesion was defined as a nodular, homogeneous, and hypodense mass that showed minimal contrast enhance-

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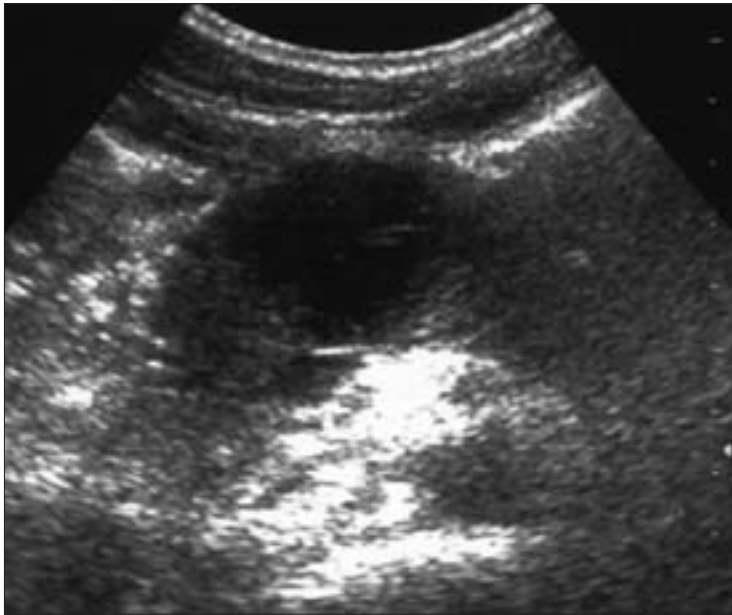


Figure 1. Transverse abdominal US image shows a nodular lesion with low echogenicity, partially well rounded and with internal septation in the body of the pancreas.



Figure 2. a, b. Noncontrast axial abdominal CT image (a) shows the lesion in the pancreatic body is indistinguishable from parenchyma with homogenous convexity outbulging towards posterior gastric surface (arrows). Post-contrast axial abdominal CT image (b) shows the pancreatic lesion displaying mild and homogeneous contrast enhancement (arrows).

ment (50 HU vs. 63 HU before and after IV contrast injection) (Fig. 2). T1-weighted turbo spin echo MRI showed a nodular, partially well-circumscribed, homogeneous, hypointense lesion in the body of the pancreas (Fig. 3a). T2-weighted turbo spin echo MRI showed a homogeneous, hyperintense pancreatic lesion (Fig. 3b). On post-gadolinium T1-weighted MR images (Fig. 3c), areas of linear and punctate contrast enhancement were observed along the septations inside the lesion. US guided fine needle aspiration cytology of the mass showed diffuse B cell high grade (Burkitt) lymphoma infiltration (Fig. 4).

After the diagnosis, doxorubicin based combination chemotherapy was induced. However, the patient's condition worsened rapidly, and he died from sepsis during the second month of chemotherapy.

Discussion

Multiorgan involvement including pancreas is relatively common in Burkitt lymphoma, occurring in 82% of cases in one autopsy series (5). Pancreatic involvement alone, without other organ or lymph node involvement is the definition of primary pancreatic lymphoma. Burkitt lymphoma with isolated involvement of pancreas is quite rare. Patients present with rapidly growing masses and a very high lactate dehydrogenase, but are potentially curable with intensive doxorubicin-based combination chemotherapy. In contrast to African Burkitt, undifferentiated American Burkitt lymphoma has a lower incidence of jaw involvement and a higher incidence of abdominal tumors, pleural effusion, and retroperitoneal adenopathy (5, 6).

Involvement of the pancreas may be either focal or diffuse with multiple discrete nodules or diffuse involvement. Sometimes the pancreas is embedded in massive peripancreatic lymphadenopathy, in which case it is not possible to know if the pancreas is involved or merely compressed (7).

Differential diagnosis mainly involves adenocarcinoma and pancreatitis. In solitary primary pancreatic lymphoma, the mass is often larger than a typical adenocarcinoma.

Absence of prominent pancreatic canal dilatation regardless of mass size and lymphadenopathy located below the level of left renal vein favor the diag-

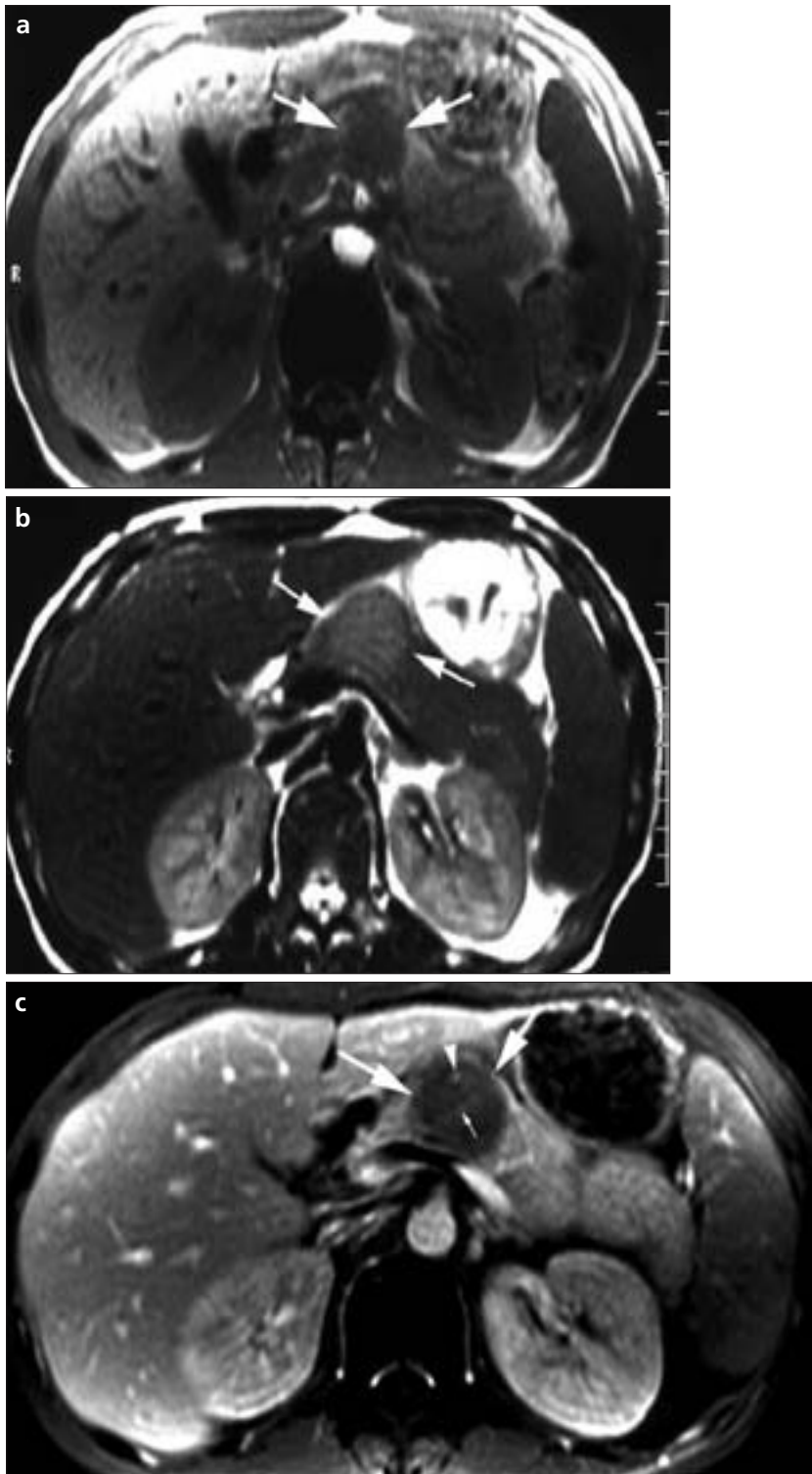


Figure 3. a–c. Axial T1-weighted MRI (TR, 120; TE, 5.3) (a) demonstrates a nodular, partially well rounded, homogeneous, hypointense lesion in the body of the pancreas (arrows). Axial T2-weighted MRI (TR, 5500; TE, 120) (b) demonstrates homogeneous, hyperintense lesion (arrows). Axial contrast-enhanced T1-weighted MRI (TR, 151; TE, 4.1) (c) shows that only the septae in the lesion (large white arrows) display contrast enhancement in a punctate (white arrowhead) and linear (small white arrow) fashion.

nosis of primary pancreatic lymphoma. Pancreatic lymphomas, unlike pancreatic adenocarcinoma, rarely cause vas-

cular stenosis or occlusion. Jaundice is also infrequent. Burkitt lymphoma can present as diffuse enlargement of the

entire pancreatic gland, caused by tumor-induced pancreatitis or pancreatitis secondary to tumor lysis following chemotherapy (8, 9).

Pancreatic lesions in Burkitt lymphoma are usually diagnosed by US, CT, and MRI. There is no specific radiological appearance of diffuse or focal pancreatic involvement of the disease. Diffuse or focal hypoechoic pancreatic enlargement is the major finding in US (10). Peripancreatic lymph node enlargement may not be distinguished by US from the pancreatic tumor. Hypodense, homogeneous, solid lesions on CT were initially reported as the radiological findings of pancreatic involvement of Burkitt lymphoma (11). However, small heterogeneous areas within a tumor mass can also be seen (8). CT findings include two morphological patterns: localized well-circumscribed tumor and diffuse enlargement infiltrating the whole gland. In our case, abdominal US showed a focal, partially well rounded and hypoechoic mass in the pancreatic body. We also observed several linear echoes on US within the hypoechoic solid mass resembling internal septae (Fig. 1). In line with the previous reports, we defined the lesion as a nodular, homogeneous, and hypodense mass on CT. Contrast enhancement was also poor but still homogeneous, though heterogeneous enhancement has been described in occasional cases (12). Although this appearance can be confused with ductal adenocarcinoma, and several radiological signs that assist in differential diagnosis have been reported, early needle biopsy is indicated in each case. There was mild homogenous contrast enhancement in our case, as determined by density measurements.

MRI findings in pancreatic lymphoma have not been thoroughly characterized. As with CT findings, two morphological patterns have been described on MRI (8). In the first pattern (corresponding to focal mass on CT) a well circumscribed low-signal intensity homogeneous mass with contrast enhancement on T1-weighted images and heterogeneous low-to-intermediate signal intensity higher than that of the rest of the gland on T2-weighted images can be seen. In the second pattern, diffuse involvement of the gland with low signal intensity on unenhanced T1- and T2-weighted images and mild-to-moderate contrast enhancement is

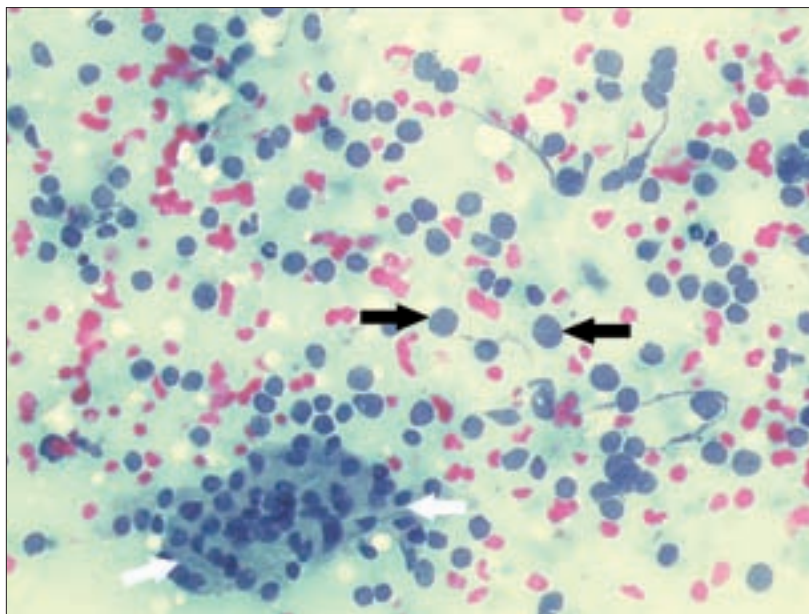


Figure 4. Fine-needle aspiration cytology demonstrates clusters of normal pancreatic acinar cells (*white arrows*) and atypical lymphoid cells with small-to-moderate sized and noncleaved round nuclei (*black arrows*) (Papanicolaou stain, x200).

observed. In our case, on T2-weighted images, the pancreatic mass was not heterogeneous but homogeneous, and it showed no enhancement after gadolinium injection. Only the septae in the lesion displayed linear and punctate contrast enhancement. These findings point out individual differences in MRI findings in patients with pancreatic lymphoma.

Lymphomas generally require larger samples, at least a core needle biopsy, for a confident pathological diagnosis (13). However, our hospital protocol is to use the least invasive intervention first. Hence, core biopsy was not required since the US-guided fine

needle aspiration yielded diagnostic cytology results. Hence, the interdisciplinary team did not ask for further invasive procedures and started the treatment based on fine-needle biopsy findings.

In conclusion, non-Hodgkin lymphoma should always be considered in the differential diagnosis of pancreatic nodular or diffuse lesions. At present, MRI should not be regarded as a superior noninvasive technique to CT in distinguishing pancreatic lymphoma from other types of masses. A rapid needle biopsy, as in our case, seems to be an accurate measure of diagnosis.

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