

## CASE REPORT

# A Rare Case of Primary Pancreatic Burkitt Lymphoma in a Young Indian Male. Case Report and Review of the Literature

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### ABSTRACT

**Context** Lymphomas of the gastrointestinal system are usually of a non-Hodgkin's type. Primary lymphomas of the pancreas are uncommon and Burkitt lymphoma involving the pancreas is very rare. It is important to recognize this entity because it can mimic adenocarcinoma but its management is entirely different. **Case report** We present the case of a young Indian male who presented with rapidly progressing obstructive jaundice, gastric outlet obstruction and severe weight loss. **Conclusion** Early diagnosis of this aggressive tumor and prompt induction of chemotherapy dramatically improved the patient's condition and avoided unnecessary surgical intervention.

### INTRODUCTION

In the middle of the last century, Denis Burkitt, working in central Africa, noted children with grossly distorted faces, and lesions involving the face, and upper and lower jaws. This malignancy, initially thought to be a sarcoma and later established to be a lymphoma, was given the name Burkitt lymphoma [1]. Burkitt lymphoma is a high-grade B-cell neoplasm of a non-Hodgkin lymphoma type. Primary pancreatic lymphomas, usually non-Hodgkin lymphomas, made up fewer than 2% of extranodal malignant lymphomas and 0.5% of all pancreatic masses [2]. The frequency of Burkitt lymphoma among primary pancreatic lymphoma is not known. A PubMed search for primary pancreatic Burkitt lymphoma and pancreatic Burkitt lymphoma revealed 17 case reports/series, six of them in the pediatric age group.

This tumor is very rarely seen primarily involving the pancreas. Burkitt lymphoma is one of the fastest growing malignancies in humans. Because of its rapid proliferation, early diagnosis and treatment offers the best chance of survival.

### CASE REPORT

We report the case of a 21-year-old male who presented with jaundice of a 25-day duration followed by increasing swelling in the epigastric region. This was associated with severe abdominal pain, abdominal distension, vomiting and fever. He also had significant weight loss. There was no history of any major illness in the past. Examination revealed a cachectic, febrile patient with deep scleral icterus, without peripheral lymphadenopathy. An abdominal examination revealed a tender mass 7x7 cm in size in the epigastric region with an enlarged liver, which was felt separately from the mass. The spleen was not palpable. Free fluid was present in the abdomen. The results of the clinical tests carried out are as follows: hemoglobin: 10.8 g/dL (reference range: 13.0-16.0 g/dL); MCV: 86 fl (reference range: 78-100 fl); total leucocyte count: 12.8 x10<sup>9</sup>/L (reference range: 4.5-11.0 x10<sup>9</sup>/L) with 80% neutrophils (reference range: 40-70%); erythrocyte sedimentation rate: 32 mm/1<sup>st</sup> h (reference range: 0-17 mm/1<sup>st</sup> h); platelet count: 750 x10<sup>9</sup>/L (reference range: 150-350 x10<sup>9</sup>/L); total bilirubin: 646 µmol/L (reference range: 5.1-17.1 µmol/L); direct bilirubin: 374 µmol/L (reference range: 1.70-5.13 µmol/L); SGOT: 136 U/L (reference range: 5-40 U/L); SGPT: 309 U/L (reference range: 5-40 U/L); serum alkaline phosphatase: 2,220 U/L (reference range: 80-270 U/L); serum amylase: 325 U/L (reference range: 0-120 U/L); LDH: 480 U/L (reference range: 70-240 U/L); prothrombin time: 14 s (control: 11 s), random blood sugar: 61 mg/dL (reference range: 70-100 mg/dL); CA 19-9: 656 U/mL (reference range: 0-37 U/mL); calcium: 8.5 mg/dL (reference range: 9-11 mg/dL); uric acid: 3.5 mg/dL

Received August 13<sup>th</sup>, 2009 - Accepted September 18<sup>th</sup>, 2009

**Key words** Burkitt Lymphoma; CVAD protocol; Jaundice, Obstructive; Pancreas

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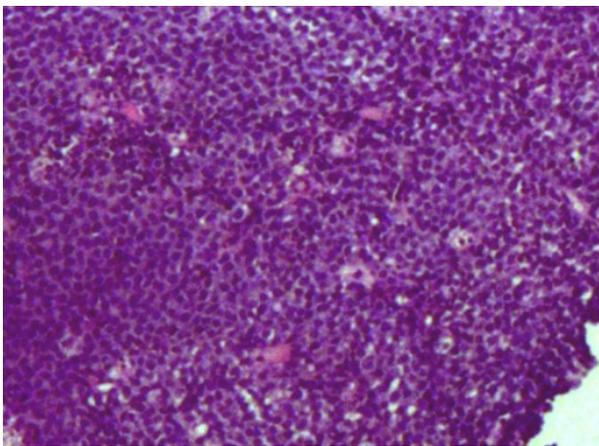
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**Document URL** <http://www.joplink.net/prev/200911/14.html>

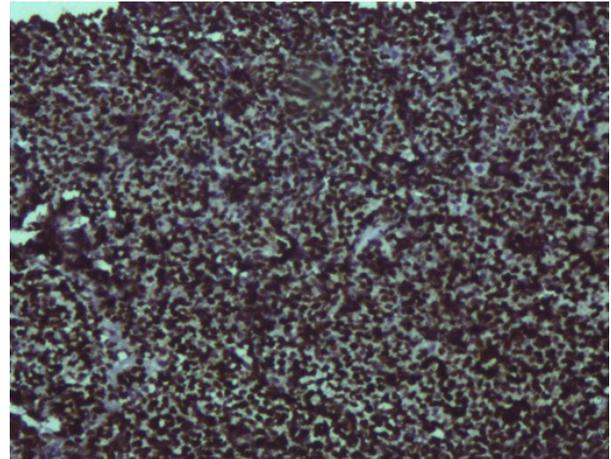


**Figure 1.** CT scan of the abdomen before chemotherapy showing arterial involvement by the mass.

(reference range: 2.0-6.5 mg/dL); creatinine: 1.2 mg/dL (reference range: 0.8-1.5 mg/dL). A CT scan of the abdomen (Figure 1) showed a 12x11x10 cm solid mass in the head of the pancreas, encasing the duodenum and the cystic duct. It was compressing the common bile duct and causing intrahepatic biliary duct dilation. The mass also involved the portal vein and hepatic artery. A CT scan of the thorax showed no lymphadenopathy. Upper GI endoscopy showed tumor infiltration of the first and second parts of the duodenum. Biopsies were taken; the histopathological examination was suggestive of high grade non-Hodgkin's lymphoma. Endoscopic ultrasound showed a solid mass in the region of the pancreatic head with involvement of the portal vein. EUS-guided FNA was carried out on the mass; however, no conclusive opinion was possible because the sample was insufficient. US-guided percutaneous biopsy of the mass showed a malignant round cell tumor suggestive of non-Hodgkin's lymphoma. On immunohistochemistry (Figures 2 and 3), the tumor cells expressed leukocyte common antigen CD20<sup>+</sup>, CD10<sup>+</sup>, bcl 6<sup>+</sup> and were CD5<sup>-</sup>. The Mib 1 index was 99%. These findings were consistent with Burkitt lymphoma. The ascitic



**Figure 2.** Starry sky appearance on H&E staining (4x).

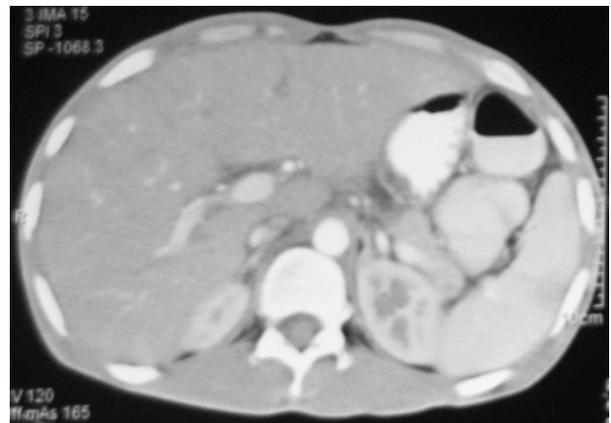


**Figure 3.** Mib1 staining showing 99% positivity (4x).

fluid field was full of tumor cells. Bone marrow and cerebrospinal fluid studies were normal. As a result of the above investigations, the patient was diagnosed as having Burkitt lymphoma of the pancreas and was treated with 2 cycles of CHOP (cyclophosphamide, adriamycin, vincristine and prednisolone) following which his general condition improved dramatically. The vomiting and pain subsided; the ascites disappeared, and total bilirubin and alkaline phosphatase levels decreased to 115.6  $\mu$ mol/L and 680 U/L, respectively, over a period of one week. A CT scan of the abdomen (Figure 4) was repeated and showed a marked reduction in the size of the tumor mass. He was started on a hyper CVAD (cyclophosphamide, vincristine, doxorubicin, dexamethasone) regimen as definitive therapy.

## DISCUSSION

Lymphomas are the only tumors, other than adenocarcinomas, which occur with some frequency in the pancreas, although they are rarely found in the pancreas. Primary pancreatic lymphomas represent less than 2% of all NHLs, although nearly one-third of non-Hodgkin's lymphoma patients will develop pancreatic



**Figure 4.** CT scan of the abdomen after chemotherapy showing a marked reduction in tumor size.

involvement at some point in their disease course. Cases have been described in both pediatric and adult population, presenting with obstructive jaundice [3, 4], pancreatitis [5] or symptoms of gastric outlet obstruction. In older individuals with elevated CA 19-9 levels, Burkitt lymphoma can be mistaken for adenocarcinoma of the pancreas and they therefore undergo major surgery [3].

In the World Health Organization (WHO) classification, three clinical variants of Burkitt lymphoma are described: endemic, sporadic, and immunodeficiency-associated [6].

Endemic Burkitt lymphoma (eBL) refers to those cases occurring in African children (usually 4-7 years of age), with a male:female ratio of 2:1, involving the bones of the jaw and other facial bones, as well as the kidneys, gastrointestinal tract, ovaries, breast and other extra-nodal sites. Epstein-Barr virus (EBV) positivity is found in nearly all cases.

The sporadic or American variety (sBL) occurs worldwide. It includes those cases occurring with no specific geographic or climatic association. It accounts for 1-2% of lymphomas in adults and up to 40% of lymphomas in children in the U.S. and Western Europe. The abdomen, especially the ileocecal area, is the most common site of involvement. The ovaries, kidneys, omentum and Waldeyer's ring may also be involved. Lymph node involvement is more common in adults than in children. Neoplastic cells are EBV positive in 15-30% of cases, or less.

Immunodeficiency-associated Burkitt lymphoma (iBL) occurs mainly in patients infected with HIV and it is frequently seen in patients with CD4<sup>+</sup> counts greater than 200 mm<sup>-3</sup> unlike other HIV-related lymphomas [7]. They are also seen in allograft recipients and individuals with congenital immunodeficiency. Our patient was negative for HIV serology. Morphologically, classic Burkitt lymphoma consists of medium-sized cells with abundant, basophilic cytoplasm, often containing lipid vacuoles, round nuclei with clumped chromatin and multiple nucleoli with a diffuse, monotonous pattern of infiltration. A "starry sky" appearance has been described in this type of non-Hodgkin's lymphoma because of its abundant proliferative rate, frequent apoptoses and numerous macrophages containing ingested apoptotic tumor cells. Eighty percent of Burkitt lymphoma cases harbor t(8;14) translocation [6], resulting in the juxtaposition of the *c-myc* gene on chromosome 8 with *IgH* enhancer elements on chromosome 14. In the remaining 20% of cases, t(2;8) or t(8;22) are observed placing the *c-myc* gene adjacent to either the kappa or lambda light chain (IgL), respectively [6]. Patients with any of the three clinical variants are at risk for diffusion to the central nervous system (CNS) and bone marrow. The bone marrow is positive in 30-38% and the CNS is involved in 13-17% of adult cases [7].

The Ann Arbor and the St. Jude/Murphy staging systems are the commonly used staging methods. Our

patient falls in the III B class of the St. Judes/Murphy staging [7].

Features which have been associated with an adverse outcome in adults and children include older age, advanced stage, poor performance status, bulky disease, high LDH, and CNS or bone marrow involvement [6].

The cases reported from India [8, 9] are in the pediatric age group, involving the stomach and the duodenum with no direct pancreatic involvement. The important clue to diagnosis in this case was the rapidity of symptom progression and the elevated LDH levels. Studies in pediatric populations have shown that a CT scan is an important mode of investigation in characterizing the lesion [10]. Two morphological patterns were described on CT scan: a localized well-circumscribed tumor and a diffuse enlargement infiltrating the whole gland. A localized, well-circumscribed form without obstructive jaundice is described in the literature [11]. A CT scan of the abdomen showed involvement of the head of the pancreas in our case. In this instance, EUS FNA did not yield a sufficient sample for a conclusive diagnosis. This patient had low blood sugar levels. This was different from other cases where temporary diabetes mellitus requiring insulin was described [12]. This could be due to the high tumor burden and poor nutrition.

Finally, differentiation between diffuse large B cell lymphoma (DLBCL) and Burkitt lymphoma is of crucial importance since both prognosis and treatment differ. The WHO criteria for Burkitt lymphoma include classical/atypical histology, CD20<sup>+</sup>, bcl 6<sup>+</sup>, CD10<sup>+</sup>, Bcl 2<sup>-</sup>, CD5<sup>-</sup>, a Ki 67 score greater than 95% and IG-*myc*<sup>+</sup> [13]. Our patient met the above criteria to support the diagnosis of Burkitt lymphoma.

## CONCLUSION

Burkitt lymphoma of the pancreas is a rare, rapidly progressing malignant neoplasia and can present with varied clinical features. Its infrequent occurrence among adults in India should not deter the clinician from including it as a part of the differential diagnosis in a patient with rapidly developing jaundice, an abdominal mass and ascites. The importance of good clinical acumen in the early diagnosis of the disease cannot be overemphasized. The present case is a fitting example of the importance of a timely diagnosis and prompt treatment which will result in a favorable clinical outcome.

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**Conflict of interest** The authors have no potential conflicts of interest

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