A RARE PRESENTATION OF CHILDHOOD ACUTE LYMPHOBLASTIC LEUKEMIA WITH OBSTRUCTIVE JAUNDICE DUE TO PANCREATIC INVOLVEMENT - CASE REPORT

KHALID N. ABDURRAHMAN, MBChB, DCH, FIBMS*

Submitted 9 Dec 2010; accepted 28 Apr 2011

SUMMARY

Obstructive jaundice due to pancreatic mass is a rare manifestation of acute lymphoblastic leukemia in children with only a few reported cases. We report the clinical, hematological and radiological findings of a 20-month-old boy with pre-B acute lymphoblastic leukemia and pancreatic involvement. Liver function tests showed an obstructive picture and a computed tomography scan of his abdomen demonstrated bulky pancreas with a hypodense texture. Three weeks after induction chemotherapy his jaundice resolved, the pancreatic mass reduced in size and he is now in complete hematological remission. Acute lymphoblastic leukemia may mimic other causes of a pancreatic mass and should be considered as part of the differential diagnosis when atypical features are present.

Duhok Med J 2011;5(1): 78-83.

Key words: Acute leukemia, Obstructive jaundice, Pancreatic involvement, ALL

Acute Lymphoblastic Leukemia (ALL) is the most common malignancy reported among Iraqi children.[1] Common features at presentation include pallor, fever, fatigue, bleeding tendency, lymphadenopathy, splenomegaly and hepatomegaly. Other features are rather infrequent at presentation including central nervous System (CNS) and testicular involvement.[2] Cholestatic jaundice is quite an unusual presentation of ALL, it is even rarer to be caused by involvement of the pancreas resulting in obstructive jaundice. We report on a 20 month old male child with ALL presenting as a pancreatic mass and obstructive jaundice.

CASE PRESENTATION

A 20-months-old boy presented with a 7-day history of jaundice. He had no significant past medical history. Physical examination showed jaundice, anemia, hepatomegaly (liver span 9 cm), splenomegaly (4 cm BCM) and painless firm right testicular swelling. Liver function tests showed an obstructive picture [serum total bilirubin 14.3 mg/dl with direct of 11.4 mg/dl, alanine transaminase of 42 IU/L (reference range: 10-45 IU/L) and serum alkaline phosphatase of 565 IU/L (reference range: 71-142 IU/L)]. Complete blood picture revealed a hemoglobin concentration of 7 g/dl, total white cell count of 10.9 x 10^9/L with 44% blasts, and platelet count of 160 x 10^9/L. Bone marrow aspirate showed infiltration by a heterogeneous population of blasts constituting 71% of all nucleated elements and confirmed a diagnosis of acute leukemia. Immunophenotyping demonstrated a pre- B-cell Acute Lymphoblastic Leukemia phenotype.

Contrast-enhanced computed tomography of the abdomen showed a diffusely enlarged pancreas with distension of gall bladder, free of gall stones (Figure 1).

Examination of CSF indicated no evidence of CNS involvement. Testicular biopsy showed infiltration with lymphoblasts.

He was commenced on chemotherapy according to the MRC UKALL 2003

* *Assistant professor of Pediatrics, Department of Pediatrics, School of Medicine, Faculty of Medical Sciences University of Duhok, Heevi Pediatric Teaching Hospital. Email: drkhnawaf@yahoo.com*

protocol.

A significant drop in bilirubin and alkaline phosphatase was noticed within 10 days and they both returned to normal in three weeks. After 28 days of initiation of induction chemotherapy, a repeat bone marrow assessment showed complete hematological remission (blasts constituting 4% of all nucleated cells). A repeat of the abdominal CT scan demonstrated significant reduction in the size of the pancreas (Figure 2).

Written informed consent was obtained from the patient's guardian.

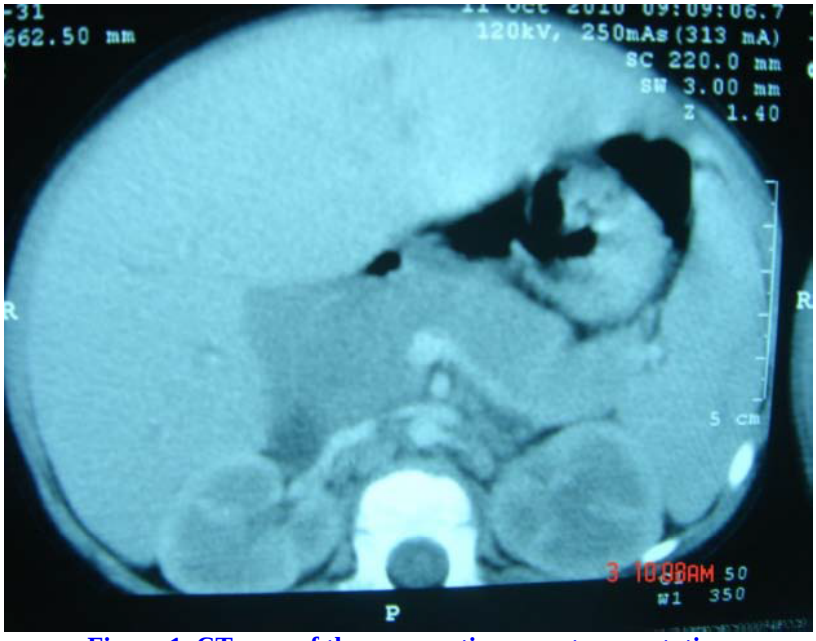

Figure 1. CT scan of the pancreatic mass at presentation

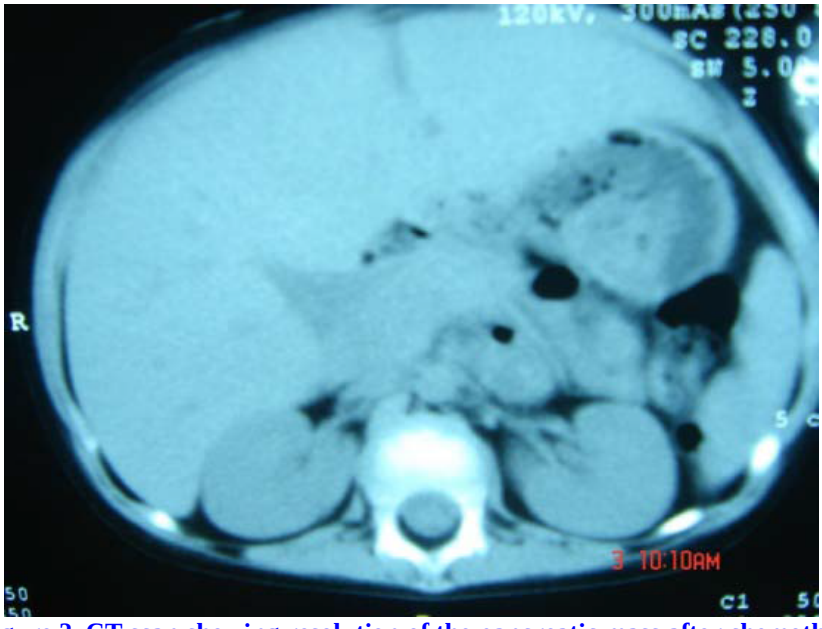

Figure 2. CT scan showing resolution of the pancreatic mass after chemotherapy

DISCUSSION

Although ALL is primarily a disease of bone marrow and peripheral blood, any organ or tissue may be infiltrated by the abnormal cells. Such infiltration may be clinically apparent by physical examination or it may be occult and detectable only by histologic sampling.[2]

Pancreatic involvement in ALL is very rare at presentation and only a few cases have been reported previously with ages ranging from 10 weeks to 39 years.[3-8] This may be asymptomatic or associated with obstructive jaundice.[3-5,8] In our case report the patient presented with cholestatic jaundice and further investigations uncovered the acute leukemic process. Pancreatic involvement has been reported in association with precursor B-cell phenotype, similar to the current case report,[4,7] and also in association with T-cell and mature B phenotypes.[6,8] Furthermore, our patient had in addition to his pancreatic involvement, involvement of another extramedullary site (testicular involvement) which is similar to some of the previous reports where additional extramedullary organ(s) involvement were noted.[7,8]

Pancreatic involvement has also been reported in other hematological malignancies, such as Non-Hodgkin's Lymphoma and granulocytic sarcoma.[8-10] In addition to pancreatic involvement, cholestatic jaundice in acute lymphoblastic leukemia, may also occur through diffuse infiltration of liver sinusoids or the common bile ducts by leukemic blasts.[11,12]

In conclusion, ALL is a rare cause of obstructive jaundice secondary to a pancreatic mass. It should be considered in the differential diagnosis of obstructive jaundice, since such extramedullary involvement by the leukemic process is highly responsive to systemic anti-leukemic therapy, which may be critical for subsequent management.

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پوختە

دەگمەنترین حالەتەکێ پەنجە شێرا خوینیێ یا زاروکا – راپورتا حالەتەکێ

زەرکا گرتیبونی ژبەر گرێگا پەنکریاسێ دەگمەنترین حالەتەکێ پەنجەشێرا خوینێ یا زاروکا، پتنێ چەند حالەتەك بێت هاتینە رابورت کرن ل هەمو جیهانێ. مە حالەتا زاروکەکی بیست مانك هەبو پەنجەشێرا خوینێ ل کەل گریکا پەنکریاسێ بێشکێشکر. تێستا میّلاکی دیار کر کو هەیە زەرکا گرتی و ومفراسا زکی دیارکر کو هەیە کریکا پەنکریاسێ. بشتی سی حەفتیا ژ چارەسەرکریا کیمیاوی زەرکا زاروکی کێم بو وگرێکا پەنکریاسێ ژی کێم بو. پەنجە شێرا خوینێ یا زاروکا جارنا وەکی گریکا پەنکریاسی یە، پێتڤیە ئەڤە پێش چاڤ بێت.

الخلاصة

حالة نادرة من سرطان الدم اللمفاوي لدى الاطفال باصابة البنكرياس مع يرقان انسدادي

اشهار حالة

اليرقان الانسدادي الناتج عن تورم البنكرياس من الحالات النادرة لسرطان الدم اللمفاوي لدى الاطفال. إذ تم تسجيل حالات قليلة منها، ونحن نشهر حالة طفل يبلغ من العمر عشرون شهراً مصابا بسرطان الدم اللمفاوي مع اصابة البنكرياس. حيث اظهرت فحوصات وظائف الكبد وجود يرقان انسدادي واظهر التصوير المقطعي للبطن (المفراس) وجود تورم في البنكرياس. بعد ثلاثة اسابيع من بدء العلاج الكيمياوي تبدد اليرقان وصغر حجم البنكرياس. سرطان الدم اللمفاوي قد يشبه اورام البنكرياس ويجب ان يؤخذ ذلك بنظر الاعتبار.