CASE REPORT

Diagnostic challenge of Burkitt's lymphoma at early age

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SUMMARY

A four-year-old Ibanese boy presented with subacute abdominal distension for two months duration. Ultrasound and computed tomography (CT) scan showed solid liver masses as well as bowel and intraperitoneal lesions. Initial diagnosis of intraperitoneal inflammatory process as in tuberculosis with non-liquefied liver abscess with differential diagnosis of neoplastic process was made. Liver biopsy and peritoneal fluid analysis revealed Burkitt's lymphoma (BL). We aim to highlight the diagnostic challenge of BL in this young age group emphasizing on the ultrasound and CT features of intraabdominal BL. We would also want to stress the importance of early diagnosis of BL as it is known to be the most aggressive tumour within 24 hours yet to have good survival if early diagnosis was made.

KEY WORDS:
Paediatric, intraabdominal lesion, Burkitt's Lymphoma

INTRODUCTION

Burkitt's lymphoma (BL) is one of the subtypes of Non-Hodgkin's lymphoma and is one of the most aggressive type of lymphomas in current practice as reports have shown it can increases in size within 24 hours.[1,3] It is commonly presented among children between five to seven years old and less common in children below five years old.[1]

CASE REPORT

A four-year-old Ibanese boy, presented with history of abdominal distension for five days and had been unwell for two months prior to presentation. On examination, he was afebrile, with multiple cervical lymphadenopathies. His abdomen was distended and tender on palpation. There was no hepatosplenomegaly and other systemic examinations were unremarkable.

Blood investigation showed normal total white cell and differential count, mild anaemia and thrombocytosis. Full blood picture revealed normochromic normocytic anaemia and mild thrombocytosis with no blast cell. Abdominal radiograph was grossly normal and no evidence to suggest intestinal obstruction. C-reactive protein level was slightly raised (0.8mg/dl; normal: <0.5mg/dl). Procalcitonin markers was high at 0.27ng/ml (normal: <0.05ng/ml), indicating local infection. The ESR, TB PCR, Mantoux test and tumour markers (Alpha fetoprotein and Ca19-9) were normal. Liver function test was slightly raised but no evidence to suggest obstructive biliary system.

Ultrasound abdomen (Figure 1) and computed tomography (CT) scan abdomen (Figure 2) revealed complex hypoechoic solid lesions in the left lobe of liver, peritoneal space with complex ascetic fluid. The right sided colon was eccentrically thickened with no aneurysmal dilatation of the bowel lumen seen.

An urgent peritoneal fluid aspiration and liver biopsy were performed using ultrasound guided under general anaesthesia. The peritoneal fluid aspirate revealed turbid fluid with high LDH level: 3859U/L. Histopathological evaluation of the biopsied liver tissue showed malignant lymphoid cells consistent with Burkitt's lymphoma. He had been diagnosed with Stage 3 disease based on bone marrow aspirates analysis.

Patient developed respiratory distress which required paediatric HDU care due to bilateral pleural effusion. Subsequently he developed neutropenic sepsis with mucositis and dental abscess. He responded well to respective treatments and chemotherapy commenced afterwards.

Follow up ultrasound two weeks after initial presentation and starting chemotherapy showed significant reduction of the size of liver lesion. Subsequent ultrasound after two month following chemotherapy revealed complete resolution of the liver mass and no residual bowel wall thickening in right iliac fossa.

DISCUSSION

Non Hodgkin Lymphoma (NHL) is the third most common malignancy in childhood after leukaemia and central nervous system (CNS) tumours contributing approximately 15% of childhood malignancy.[1] Burkitt's lymphoma (BL) is classified as one of the subtype of NHL and being the most frequent subtype of NHL during childhood.[2] Generally, the overall incidence of BL in less than 5 year old age group is small that was reported as 0.8% from all types of childhood cancer.[1] The mean age group of this type of cancer is approximately 8 years old and male is more common to be affected than female.[1,3] As in other types of lymphoma, BL can involves variety of locations in the body. Head and neck involvement is common in endemic variant that frequently seen in Africa that has been associated with endemic Epstein-Barr virus (EBV) whereas abdominal involvement is more common in sporadic form.[2,3]

The diagnosis of childhood BL requires histological tissue biopsy. Imaging guided tissue biopsy is often practiced when

This article was accepted: 13 June 2017
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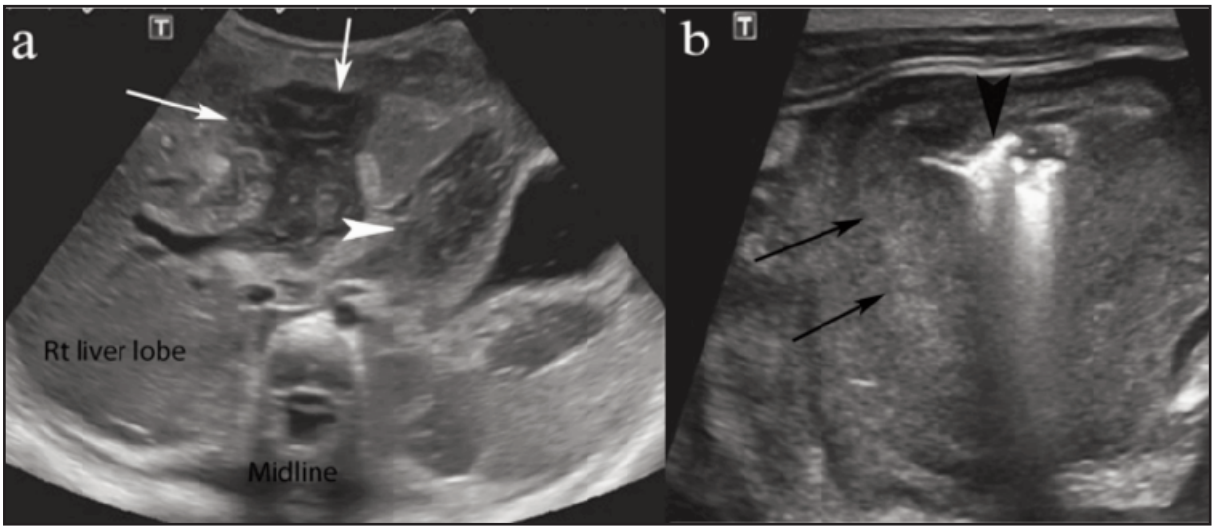

Fig. 1: Ultrasound of abdomen at time of presentation. (a) There is hypoechoic lesion in left liver lobe (white arrows) with another focal lesion in the peritoneal space at the gastrohepatic ligament (white arrowhead). (b) In the right iliac fossa region, the cecum is eccenterically thickened with complex ascitic fluid surrounding it (black arrow). The gas filled cecal lumen is shown by the black arrowhead.

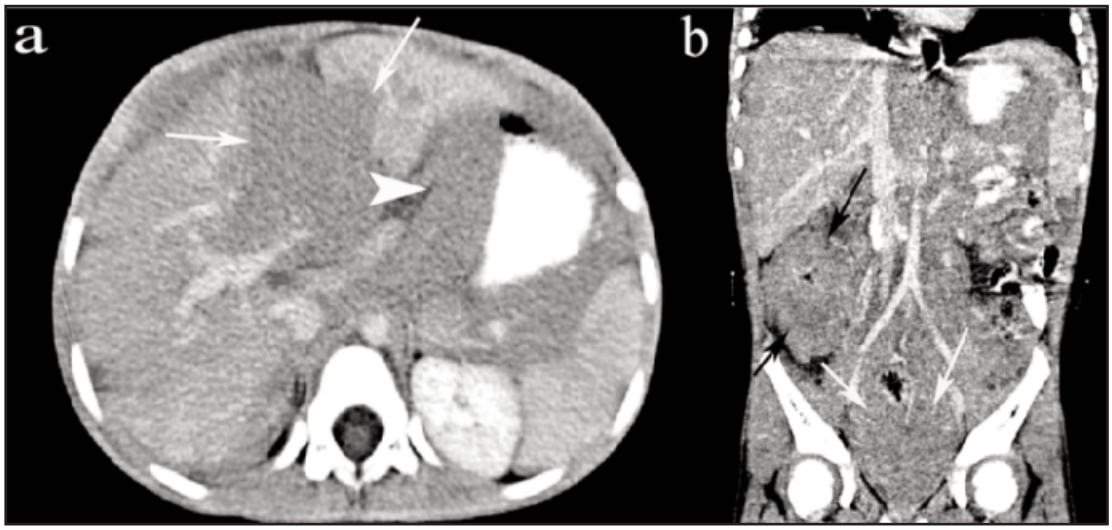

Fig. 2: Contrast enhanced Computed Tomography (CT Scan) abdomen in axial (a) and coronal (b) plane. (a) There is hypodense solid lesion in left liver lobe (white arrows) and in gastrohepatic peritoneal space (white arrowhead) as seen on ultrasound. (b) Coronal view shows the thickened cecum (black arrows) with solid soft tissue lesion in the rectovesical space (white arrows).

BL involves intra-abdominal structures. Radiological imaging mostly required to assess the site, extent of intraabdominal involvement and vascularity assessment of the lesion.[3] On ultrasound, abdominal BL imaging findings include solid hypoechoic mass lesion, asymmetric bowel wall thickening, ascites or lymphadenopathies which were also present in this patient. CT scan will define the bowel and retroperitoneal involvement in cross sectional imaging and sometimes may lead to upstaging of the disease.[2,3] Bowel, intraperitoneal/mesentery and retroperitoneal involvement of BL are infiltrative in nature in which this patient have multifoci soft tissue masses in the upper peritoneal cavity as well as in the pelvis. Secondary involvement of the intra-abdominal solid organs such as liver, spleen or kidney is more common than primary involvement.[3] Therefore, the liver lesion in this patient is likely secondary to adjacent peritoneal disease that has infiltrated the left liver lobe.

Other common possible diagnosis in younger age group is subacute bacterial peritonitis which includes intra-abdominal tuberculosis (TB).[4] Ileocecal involvement accounts for 80-90% of patient with abdominal tuberculosis with more than 50% of patients may show circumferential thickening of the caecum and terminal ileum as in our patient.[4] Lymphoma was a less favourable diagnosis at the time of presentation in our patient based on early age at presentation and clinical suspicion of infection, but this should be something that need to be thought about when

dealing with infiltrative intra-abdominal solid mass lesion. Liver involvement in tuberculosis usually manifest in nodular lesions of miliary (micronodular) or tuberculoma (macronodular).[4] Ileocecal and liver involvement in this case mimicking tuberculosis, however, infiltrative liver lesions on imaging and the diagnosis from culture of peritoneal fluid of this patient were negative for tuberculosis.

With prompt diagnosis, five-year survival rate for those younger than 20-year-old is quite high, reaching up to 72%.[1] The main stay of treatment would be chemotherapy as what has been commenced in this patient. Role of surgery in BL usually when patient presented with surgical emergencies such as acute intestinal obstruction, per-rectal bleeding or intussuception.[5]

CONCLUSION

Diagnosis of Burkitt's lymphoma in young age group is rather challenging as the presentation is commonly non-specific and imaging features may mimic inflammatory changes as in abdominal tuberculosis. Since urgent and invasive tissue diagnosis could only be arranged until malignant lesion was suspected, this disease need to be considered whenever there is ill-defined solid infiltrative hypodense lesion in the abdomen or pelvis regardless of age group presentation. Early diagnosis of Burkitt's lymphoma is crucial as this disease has excellent survival rates because it is chemo responsive even though it is known to be aggressive on imaging.

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