

Case Report

Burkitt's lymphoma: Causing obstructive jaundice in an adult

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Obstructive jaundice secondary to Burkitt's lymphoma (BL) is very rare. It is important to recognise this condition as it can mimic adenocarcinoma of the pancreas, but the management of the two conditions is very different. In this study, we present the case of a seventy-nine year old male with right upper quadrant pain, jaundice, and pruritus. It is crucial to consider BL as a differential diagnosis of obstructive jaundice as both the prognosis and treatment are markedly different.

Key words: Burkitt's, Jaundice, lymphoma, adult.

INTRODUCTION

Obstructive jaundice secondary to gallstones or malignancy is not an uncommon presentation in surgical practice (Collier and Morris, 2006). However, obstructive jaundice due to Non-Hodgkin's lymphoma (NHL) is quite rare (Fidias et al., 1995; Kessel et al., 2005). In only 1% of the patients with NHL, obstructive jaundice is the presenting sign of the disease (Joo et al., 2004). The mechanism of obstructive jaundice due to NHL is through enlarged lymph nodes compressing the biliary tree. The most common sites of obstruction tend to be the liver hilum and distal common bile duct (CBD) (Ravindra et al., 2003).

Most NHL cases that develop obstructive jaundice are histologically diffuse B-cell lymphomas. Burkitt's lymphoma (BL) is a solid sub-type of B-cell lymphoma and occurs commonly in children (Joo et al., 2004). This tumour is very rarely seen involving the pancreas (Nistala et al., 2009). This study presents a case of an adult patient with BL whose presenting symptoms were jaundice, abdominal pain, and pruritus.

CASE REPORT

A 79 year old Vietnamese male presented to the emergency department with a one week history of right upper quadrant pain and jaundice. The pain was described as a dull

ache without any radiation. He also described obstructive symptoms, such as dark coloured urine and pruritus, but denied any change in colour of his stools. He denied any fevers or sweats at home and he had no nausea, vomiting or any change in bowel habit. He did report some decreased appetite, but no significant weight loss. He denied any excessive alcohol consumption and he had not travelled overseas recently. The patient also denied any recent tattoos or intravenous drug use. There was no previous history of gallstones or jaundice and he had not been on any antibiotics recently. The only past history included that of hypertension. On examination, he was a man of thin build and clearly jaundiced with icteric sclerae. All his vital signs were within normal limits. Abdominal examination revealed a vague fullness in the epigastrium region, but an otherwise soft abdomen. There was no palpable lymphadenopathy. Laboratory assessment revealed a normal full blood count with normal white cell count. The liver function tests were markedly deranged with alkaline phosphatase (ALP) of 915 IU/L, gamma-glutamyl transpeptidase (GGT) of 1124 IU/L, alanine transaminase (ALT) of 493 IU/L, aspartate transaminase (AST) of 362 IU/L, and total bilirubin of 196 µmol/L. Tumour markers demonstrated a carbohydrate antigen (CA) 19.9 level of 169 U/ml and carcinoembryonic antigen (CEA) of 0.7 µg/L. He had a lipase level of 22 IU/L. Sputum and blood cultures for TB were negative. The hepatitis screen showed that the patient had cleared a past hepatitis B infection (Hep B core antibody positive, Hep B surface antigen positive and a moderately positive surface antibody). He was negative for hepatitis C virus (HCV) and human immunodeficiency virus (HIV).

An abdominal computed tomography (CT) scan was performed which demonstrated an enlarged heterogeneous pancreatic head mass causing biliary tree dilatation with dilated intra and extra-hepatic ducts (Figures 1A and B). There was also diffuse thickening and soft tissue density involving the greater omentum, caecum, and terminal ileum.

Following these results, the patient underwent a laparoscopy

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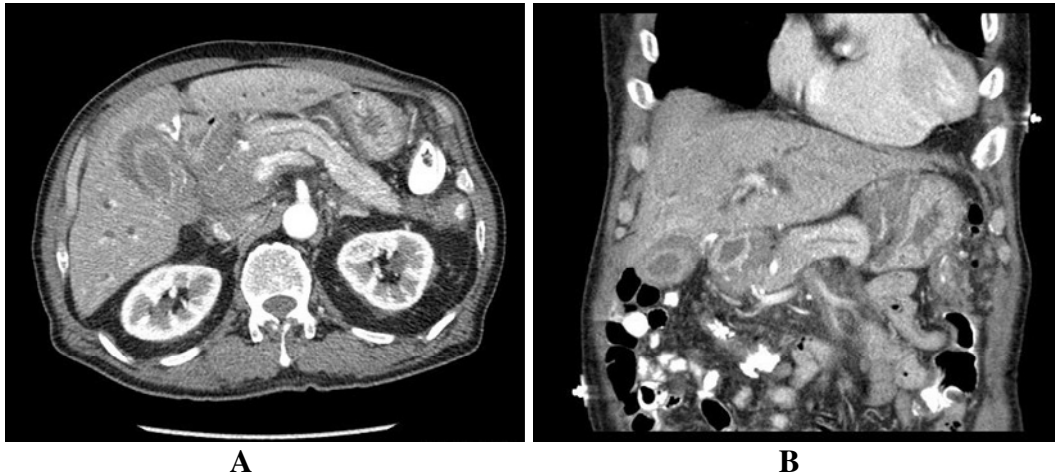


Figure 1. Axial (A) and coronal (B) slices of abdominal CT highlighting the pancreatic head mass causing intrahepatic dilatation.

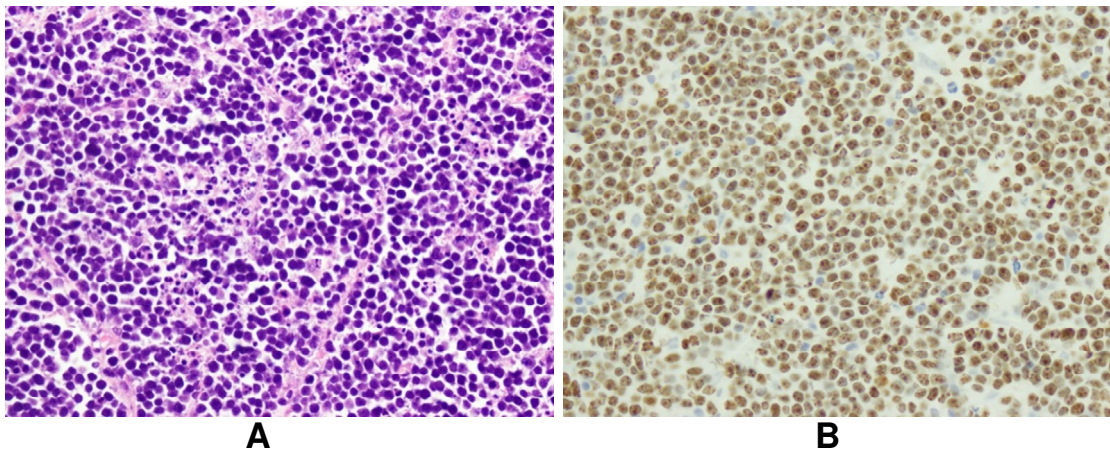


Figure 2. Burkitt's lymphoma: (A) H&E stain showing atypical lymphoid cells with 'starry sky' pattern of apoptosis. (B) Ki67 showing > 99% staining typical of Burkitt's lymphoma.

and biopsy of the omental mass. The patient also underwent an endoscopic retrograde cholangiopancreatography (ERCP) which highlighted dilated intra and extra-hepatic ducts and a long stricture in the bile duct from the porta-hepatis to the ampulla. A 7-french, 11 cm plastic stent was inserted in the CBD to relieve the obstructive jaundice. The histopathology from the biopsy of the omental mass showed small to medium sized atypical lymphoid cells with "squaring off" of nuclei and starry sky pattern of apoptosis in the background (Figure 2A). The immunostains yielded the following immunophenotypes: CD3 -ve, CD5 -ve, CD10 +ve, CD20 diffuse moderate to strong staining, CD23 -ve, CD30 -ve, CD43 -ve, CD79a +ve, TdT -ve, bcl-2 -ve, bcl-6 +ve, MUM1 +/- (focal, weak), EBER-CISH -ve, Ki67 index >95% (Figure 2B) which confirmed the diagnosis of Burkitt's lymphoma.

The patient was referred to the oncology/haematology team and post initial investigations and discussion with patient and family, commenced on a chemotherapy regime which included cyclophosphamide, doxorubicin, vincristine, prednisolone (CHOP), and intrathecal methotrexate. He was given further 5 cycles of etoposide, prednisolone, vincristine, doxorubicin, and

cyclophosphamide (EPOCH). The patient had a good early response with decreasing tumour size on positron emission tomography (PET) scan, but unfortunately relapsed post 5th cycle of EPOCH, 6 months post commencing chemotherapy.

FINDINGS AND DISCUSSION

Burkitt's lymphoma was first described by Dr. Denis Burkitt, an Irish Surgeon, who, while working in Uganda, noted a high incidence of rapidly growing tumours affecting the jaws of children (Burkitt, 1958). BL is a subtype of B-cell non-Hodgkin's lymphoma. Unlike other forms of NHL, BL is present as an extranodal disease (Joo et al., 2004). It accounts for over half of all childhood cancers in endemic areas (Equatorial Africa and Papua New Guinea), and 40 to 50% of childhood NHLs in non-endemic areas (America and Western Europe) (Murphy

et al., 1989). BL is a rare lymphoma in adults, except in HIV-positive patients (Diebold et al., 2001). It comprises less than 1% of adult NHL (Morton et al., 2006). It is usually diagnosed through a biopsy. Chemotherapy is the mainstay of treatment and with aggressive treatment, cure rates of up to 90% are possible (Levine, 2002).

The presentation of obstructive jaundice due to BL is very rare (Kessel et al., 2005; Nistala et al., 2009). Obstructive jaundice is a presenting sign in only 1% of patients with NHL (Joo et al., 2004). On the other hand, biliary obstruction secondary to primary carcinomas of the pancreas are more frequently encountered in the clinical setting. Pancreatic cancer is an aggressive cancer with five year survival rate of less than 5%. Surgical resection is the only potentially curative treatment (Lim et al., 2003).

The case presented highlights the fact that it is important to consider lymphoma as a differential for malignant biliary obstruction. From admission, the patient was investigated for differentials of biliary obstruction. A CT scan was performed due to the undifferentiated fullness in the epigastrium palpated on examination. It was the findings on the CT scan that prompted a suspicion for other causes apart from primary pancreatic carcinoma. Hence, the patient underwent a laparoscopic biopsy of the omental mass that eventually led to the diagnosis.

In this case, the patient commenced on the appropriate chemotherapeutic agents for BL once a diagnosis was made and had a good early response.

Conclusion

Obstructive jaundice secondary to BL is a rare presentation. The presentation, however, can be very similar to adenocarcinoma of the pancreas. Hence, it is important to consider BL as a differential diagnosis for obstructive jaundice as a presentation, since the treatment for both conditions is markedly different.

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