SAJS

Case Report

Primary pancreatic lymphoma in an HIV patient: Dilemmas in diagnosis and management

E. LOOTS, F.C.S. (S.A.)
F. ANDERSON, M.MED., F.C.S. (S.A.)
S. R. THOMSON, CH.M., F.R.C.S. (ENG. & EDIN.)

Department of General Surgery, Addington Hospital and Nelson R. Mandela School of Medicine, University of Natal, Durban

The HIV pandemic has introduced a new dimension to the diagnosis and management of pancreatic head masses, adding opportunistic infections and malignancy to the differential diagnosis. Tuberculosis[1,2] and Kaposi's sarcoma have been well described as primarily involving the pancreas and are diseases which can in theory be managed medically without surgical resection. We report another obscure malignancy that adds to this quandary in the HIV-positive patient.

Case study

A 44-year-old HIV-positive man presented with obstructive jaundice, complaining of continuous abdominal pain and anorexia. He had completed a 6-month course of treatment for pulmonary tuberculosis 2 months earlier. Recently instituted antiretroviral therapy had improved his CD4 count from 63 to 123 cells/µl.

He was well nourished, jaundiced and had no lymphadenopathy. Abdominal examination revealed a mild hepatomegaly. The bilirubin level was raised at 69 µmol/l, and the alkaline phosphatase and gamma-glutamyl transpeptidase levels were 472 U/l and 736 U/l, respectively. The albumin level was 19 g/l.

Ultrasound examination excluded cholelithiasis and confirmed the presence of dilated intra- and extrahepatic bile ducts. A computed tomography (CT) scan demonstrated the presence of a 3×2 cm hypodense lesion in the head of the pancreas (Fig. 1). The superior mesenteric artery, superior mesenteric vein and portal vein were free of invasion. The chest and mediastinum were free of metastases or mediastinal lymphadenopathy. A radial endoscopic ultrasound scan (EUS) demonstrated a 2 cm homogeneous mass in the head of the pancreas.

A provisional diagnosis of early pancreatic adenocarcinoma was made. In view of the patient's good general condition laparotomy was performed with the intention of proceeding to pancreaticoduodenectomy. The tumour was resected with all the affected lymph nodes in the hepatoduodenal ligament (Fig. 2).

Postoperative recovery was slow, with wound and drain site sepsis. This was managed conservatively with CT scan monitoring and subsided after 3 weeks. The patient was discharged for oncology follow-up after a hospital stay of 4 weeks.

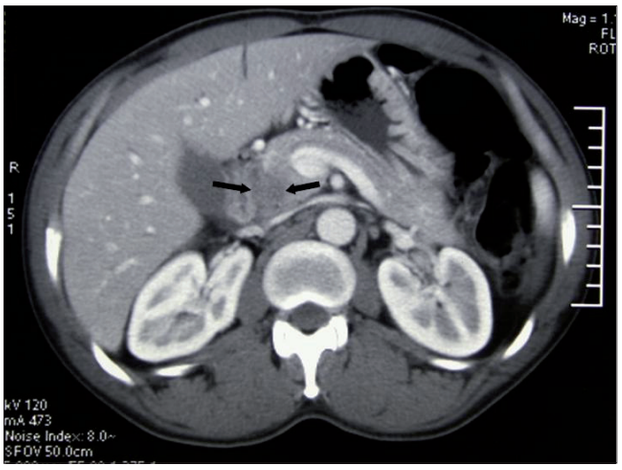

Fig. 1. CT scan image demonstrating a 3x2 cm hypodense lesion in the head of the pancreas suggesting pathology other than adenocarcinoma of the pancreas.

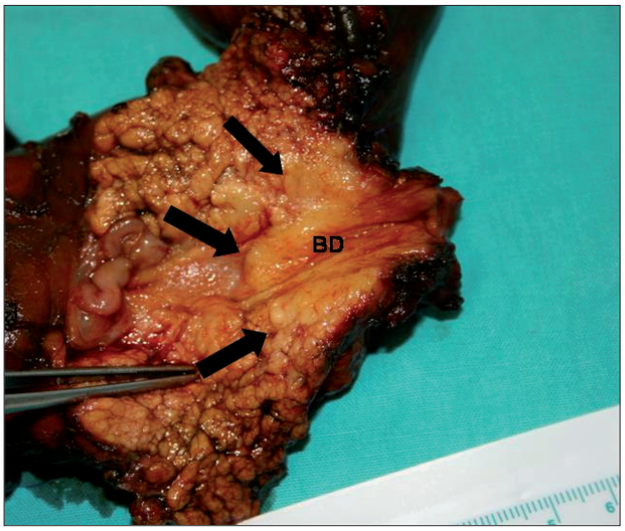

Fig. 2. The resected pancreaticoduodenectomy specimen. The arrows indicate the tumour transition zone in the head of the pancreas. The pancreatic part of the bile duct (BD) is also invaded with tumour.

Histological examination revealed a high-grade B-cell lymphoma with involved margins. The diagnosis was in keeping with the criteria for a primary pancreatic lymphoma (PPL). The patient has received 4 cycles on the CHOP regimen (cyclophasphamide, doxorubicin, vincristine and prednisolone) and is progressing well.

Discussion

Pancreatic masses in HIV-positive patients pose diagnostic and management dilemmas. This is typified by the resection of isolated tuberculous disease in the pancreatic head because of diagnostic doubt.[2] The recognition of isolated Kaposi's sarcoma[3] and now PPL has added to this conundrum, as all these diseases have the potential to be treated medically.

PPL is exceedingly rare, accounting for less than 1% of pancreatic masses. The presence of PPL in a patient with HIV/AIDS has been described once in the literature.[4] The clinical symptoms are similar to those of pancreatic adenocarcinoma. Pain and weight loss are the most common symptoms. However, jaundice, with which our patient presented, is present in only 37% of patients with PPL. Systemic signs of fever, chills and night sweats are uncommon (2%) in comparison with the frequency noted in disseminated lymphoma.[5]

The CT scan may provide clues to the diagnosis of PPL. Absence of the pathognomonic double-duct sign, size greater than 6 cm, an invasive growth pattern with retroperitoneal invasion, absence of calcification or necrosis[6] and lymph nodes below the level of the renal veins are findings that suggest an alternative diagnosis to adenocarcinoma of the head of the pancreas. None of these was evident in our case.

The above nonspecific radiological features mean that if the condition can potentially be managed non-operatively, there is a need for preoperative pathological diagnosis. The safety of preoperative transperitoneal biopsy has been questioned on the grounds that it may result in trans-coelomic disease or needle tract seeding. The very low risk of dissemination must be weighed against the benefits of a histological diagnosis that will alter management.[7]

EUS adds value in the diagnosis of smaller lesions with a diameter of less than 1 cm. We had access only to a radial scanner. A sector scanner would have allowed fine-needle aspiration biopsy (FNAB) for cytology without the risk of needle tract seeding of a potentially resectable tumour into the peritoneal cavity. This type of tissue sampling has been shown to be highly accurate in the context of adenocarcinoma before institution of neoadjuvant therapy.[7] However, the sensitivity and specificity of FNAB for rare pathologies is not well defined. Its inability to diagnose the only case of PPL in a series of 101 pancreatic head masses[12] suggests that its credibility as a single diagnostic modality requires further evaluation.

The management of PPL is based on that of systemic B-cell lymphomas. Patients with these chemosensitive tumours have a life expectancy of approximately 6 months without therapy. The CHOP regimen is the least hepatotoxic and hence the most attractive choice in the jaundiced patient. In the single HIV-positive individual in whom PPL has been reported, chemotherapy and biliary stricture managed by endotherapy, resulted in 18 months of disease-free survival.[4]

The role of surgery in PPL is unclear. There are 3 reports of more than 10 patients, constituting a total of 42 cases.[5,8,9] Of these 42 patients, 7 had resections, 8 had a bypass operation and 24 had chemotherapy in combination with surgery. It is evident that chemotherapy has generally been the mainstay of therapy, but the reports show that resective surgery alone or in combination with chemotherapy had the most favourable results in terms of disease-free survival.[5,8]

The risk of performing major surgery in HIV-positive patients is also poorly quantified. The presence of HIV infection or a low CD4 count has not been shown to predict poorer survival after surgery in a heterogeneous cohort. Wildi *et al*. described successful pancreaticoduodenectomy for adenocarcinoma in an AIDS patient with an uneventful postoperative course.[10] However, there is a paucity of data indicating whether HIV patients who are antiretroviral treatment-naive or on antiretroviral therapy will withstand or benefit from major surgery. Until more robust data are available we should assess and optimise all co-morbidities before making an overall decision about resective surgery.[11] The use of image-guided biopsies in HIV-positive patients has the potential to distinguish inflammatory pancreatic head lesions from malignant tumours and to diagnose rare tumours for which non-surgical therapy may be more appropriate.

In the HIV-positive patient it would be advantageous to be able to diagnose pancreatic head masses that would benefit from medical management without surgery. CT findings are nonspecific, and EUS with FNAB has a theoretical appeal that needs to be evaluated. In an HIV-positive patient who is generally otherwise healthy, resective surgery also enables a definitive diagnosis to be made, and is a reasonable approach that does not preclude postoperative chemotherapy.

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