Carcinoid tumours are slow growing malignant tumours, most commonly involving the appendix, ileum, rectum and other sites in the gastro-intestinal tract.1,2 Carcinoids are present in the small intestine in about 1 in 150 patients at autopsy and in the appendix in about 1 in 300 appendicectomies.3,4 Pancreatic involvement is rare. We report a case of a male Nigerian with type 2 diabetes mellitus and hypertension, presenting with features of obstructive jaundice and an abdominal mass. The mass was later diagnosed as carcinoid tumour of the head of the pancreas. The need for histologic diagnosis of all pancreatic tumours is highlighted.

Case Report

A 54-year-old male Nigerian presented to the Surgical Outpatient Department of the University of Benin Teaching Hospital, Benin City Nigeria in May 2003 with complaints of generalized pruritus, weight loss of three months duration and yellow discolouration of the eyes of one-week duration. He is a patient known to have type 2 diabetes mellitus (DM) and hypertension, both of 16 years duration, and peptic ulcer disease of 5 years duration. He claimed good compliance with diet, exercise therapy and drugs, which included glibenclamide, nifedipine and antacids as required. Pruritus was not associated with any skin lesions and there was no change in the colour of his stools and urine.

He admitted to heavy drinking and cigarette smoking for over 35 years; his daily intake, against medical advice, is approximately 60-80g ethanol and at least 10 cigarettes.

Physical examination revealed a cachectic middle-aged man with moderate pallor and jaundice. The blood pressure was 140/90 mmHg and findings on chest and neurological examination were within normal range. Examination of the abdomen revealed a soft ill-defined mass in the left lumbar region measuring 14cm x 10cm; there was no associated hepatomegaly or ascites and per rectal examination was normal. A clinical impression was made of obstructive jaundice secondary to carcinoma of the head of the pancreas in a patient with pre-existing type 2 DM, hypertension and peptic ulcer disease.

He was admitted by the surgeons and subsequently reviewed by the Endocrine Unit. He was placed on regular insulin and nifedipine in addition to a diabetic diet consisting of 2000 kcal/day. Results of investigations showed elevated total serum bilirubin of 59.8 mmol/l (5.1-17 mmol/l) and direct bilirubin of 29.1 mmol/l (1.7-5.1 mmol/l). Serum aminotransferases were within normal limits. Fasting plasma glucose was 11.8mmol/l. The abdominal CT scan showed a grossly enlarged head of the pancreas with dilatation of the pancreatic and common bile ducts, gall bladder and biliary radicles.

At exploratory laparotomy, an inoperable tumour of the pancreas was revealed and the patient had a triple by-pass surgery. Histological assessment of the pancreas confirmed the presence of a carcinoid tumour (Figure 1).

Post-operatively, chemotherapy was commenced with doxorubicin and 5-fluorouracil. Diabetes mellitus control was achieved and maintained with regular insulin; blood pressure was controlled with nifedipine. The patient was also placed on omeprazole, amoxycillin, metronidazole and antacids for treatment of peptic ulcer disease. He improved clinically and was discharged after six weeks of hospitalization. He was sent home with his routine drugs for DM and hypertension. He was subsequently seen in the Medical and Surgical Outpatient Clinics, where he remained in a good state of health until he was lost to follow-up about 15 months after surgery.

Discussion

The occurrence of obstructive jaundice secondary to carcinoid tumour of the pancreas in patients with type 2 DM is rare. Nagai et al.7 reported such an occurrence in a 66-year-old Japanese male
with a markedly dilated main pancreatic duct and DM, similar to the findings in our patient. Considering the history of weight loss, jaundice and heavy smoking for over 35 years in a middle-aged man with an abdominal mass, the CT scan findings were consistent with the presumptive diagnosis of carcinoma of the head of the pancreas. The prognosis in this case looked poor at surgery, as the median survival time for patients with surgically unresectable pancreatic carcinoma is approximately five months.8

Because of its rarity, a diagnosis of carcinoid tumour of the pancreas was not considered in our patient until the histological results were confirmatory. Carcinoid tumours of the pancreas display various non-specific imaging features on ultrasonography and CT scan, thereby causing diagnostic difficulties. Multi-slice helical tomography is useful in the diagnosis of pancreatic carcinoid and, where possible, diagnosis should be based on measurements of serotonin in serum and tumour, and/or measurement of its derivative 5-hydroxy indoleacetic acid (5HIAA).6,9,10 In hindsight, the carcinoid syndrome, as it preceded jaundice by several weeks. However, no other features of this syndrome were present in our patient.

There is no known causal relationship between DM and pancreatic carcinoid tumour. Mao et al.11 in a review of current literature on pancreatic carcinoid tumour reported that most cases did not occur in the setting of DM. They noted the rarity of pancreatic carcinoid tumours, usually diagnosed when the tumour is large, metastatic and associated with high incidence of carcinoid syndrome.11 They also noted that poor prognosis is due to delayed diagnosis and the resultant low incidence of tumour resectability.11 These features, reported in the review by Mao et al.11 are similar to the findings in our patient.

Treatment options for carcinoid tumours include medical therapy, surgical resection and selective hepatic artery embolization for hepatic metastases.13-14 Chemotherapy with drugs such as 5-fluorouracil, doxorubicin, streptozocin, dacarbazine, methotrexate and cyclophosphamide has been used with varied success rates in patients with inoperable tumours. The combination of methotrexate and cyclophosphamide is associated with objective response (i.e., >50% regression of tumour) in 55% of cases.15 Octreotide acetate is useful in tumour regression and/or stabilization.16,17 It is also useful in the management of carcinoid crisis (which may occur during surgery due to massive mediator release) and in reduction of the hormonal manifestations of the carcinoid syndrome.16,18 Our patient was placed on doxorubicin and 5-fluorouracil and was apparently stable until he was lost to follow-up. Octreotide acetate therapy could not be initiated due to financial constraints and non-availability of the drug.

The definitive diagnosis in this case was made on the basis of histological findings. Since the overall prognosis and survival rates for pancreatic carcinoid tumours are generally more favourable than those for carcinoma of the pancreas, suspected cases of pancreatic carcinoma should have histological assessment for a proper definitive diagnosis.

References