Case report

Benign inflammatory pseudotumour of the biliary tract masquerading as a Klatskin tumour

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Background

In the presence of obstructive jaundice, irregular strictures high in the common hepatic duct are usually due to bile duct cancer or to metastatic infiltration from other malignant tumours. Postoperative strictures and a variety of benign tumours also occur in this region but usually have a distinctive appearance on retrograde cholangiography. Obstruction of the bile duct due to an impacted calculus in Hartmann’s pouch (Mirizzi syndrome) and sclerosing cholangitis also have characteristic radiological appearances and are supported by the coexistence of gallstones and inflammatory bowel disease respectively.

Case report

A 57-year-old woman presented with a one week history of jaundice associated with pruritus, nausea and mild abdominal pain. She also complained of anorexia and had noticed some weight loss. Twelve months earlier she had undergone an uneventful laparoscopic cholecystectomy for repeated episodes of biliary colic and had made a full recovery. Liver function tests at the time were normal with no operative cholangiogram performed. Relevant past history consisted of a left mastectomy for breast cancer 17 years earlier without adjuvant therapy. Endoscopic retrograde cholangiopancreatography (ERCP) showed a 3-cm-long stricture extending distally from the confluence of the left and right hepatic ducts (Figure 1).

The patient was admitted electively for resection of a presumed cholangiocarcinoma of the confluence of the hepatic ducts (Klatskin tumour) [3]. On admission she was noted to be slightly obese and deeply jaundiced with no stigmata of chronic liver disease. The remainder of the physical examination was essentially normal with, in particular, no evidence of recurrent, local or metastatic disease from her previous breast cancer. Her liver function tests were markedly raised: serum bilirubin 346 µmol/L, alkaline phosphatase 245 u/L and a gamma-glutamyl-transferase 105 µmol/L. A preoperative CT scan of the abdomen showed no obvious local or distant spread of the tumour.

At operation an obviously thickened common hepatic duct was found with tumour spreading to encase the right hepatic artery. The tumour necessitated division of the right hepatic duct, the left hepatic duct at the level of the ducts to segments II and III, the common bile duct at a level below the duodenum and ligation and division of the right hepatic

Introduction

Neoplastic strictures at the confluence of bile ducts pose a difficult diagnostic and management problem. Benign non-traumatic inflammatory strictures in this location are extremely rare [1] but should be considered in the differential diagnosis to help avoid potentially inappropriate ‘palliative’ intubational therapy with its relatively high morbidity rate in the setting of benign disease [2].

Case outline

This case report describes a much rarer entity, an inflammatory pseudotumour of the bile duct, which was only diagnosed after histological evaluation of a specimen resected for presumed bile duct cancer.

Discussion

Clinicians should be aware of the possibility of a benign cause for a cholangiogram showing obstruction due to an apparent Klatskin tumour and of the good long-term outcome of surgical excision of these lesions.

Keywords

bile duct, jaundice, pseudotumour.
artery. Biliary drainage was restored with a Roux-en-Y loop of jejunum anastomosed to the right hepatic duct and to a combined opening of the ducts to segments II and III. The patient progressed well postoperatively with a gradual return to normal of her liver function tests.

Histopathology confirmed an inflammatory pseudotumour of the common hepatic duct (Figure 2). Histopathology of the fusiform stricture of the common hepatic duct near its bifurcation showed an inflammatory infiltrate composed of polymorphs, eosinophils, lymphocytes and plasma cells surrounding bile ductules and vessels within the wall of the duct and extending out into adjacent adipose tissue. Myofibroblastic proliferation was evident, which on immunoperoxidase staining showed positivity for actin and vimentin and negativity for S100 protein. The appearances were those of an inflammatory pseudotumour.

**Discussion**

Since ERCP and percutaneous transhepatic cholangiography (PTC) have been in regular clinical use, the localisation of the site of extrahepatic biliary tract obstruction in patients with obstructive jaundice has become more accurate. However, an extrahepatic biliary tract obstruction should not be automatically assumed to be malignant. ERCP can sometimes obtain a histological diagnosis, but often the clinician is left without a definite answer.

Benign neoplasms of the extrahepatic biliary tract are rare and can be of many different histological subtypes [1,4]. Benign inflammatory pseudotumour (BIP) of the bile duct is extremely rare; only two cases have been reported in the literature [5,6]. In addition, a small number of cases of benign extrahepatic biliary strictures have been reported attached to histological diagnoses including localised sclerosing cholangitis [2], non-traumatic inflammatory stricture [7], Mirizzi syndrome [8] and, most recently, inflammatory myofibroblastic tumour [9]. On review of the reports, the differentiation between the histological diagnoses seems unclear, and perhaps many of these cases represent a spectrum of the same pathological process under different names. Inflammatory myofibroblastic tumour would seem to be the most acceptable pathological description to encompass all these reports. It is described microscopically as a tumour comprising varying proportions of spindle cells.
and plasma cells, lymphocytes, eosinophils and macrophages. The spindle cells usually stain positively for actin and vimentin but negatively for S100 and cytokeratin. The aetiology is uncertain, but local trauma (e.g. previous cholecystectomy) does not appear to be an explanation in the majority of cases.

An inflammatory pseudotumour (synonym plasma cell granuloma, inflammatory myofibroblastic tumour) is a benign localised proliferation of plasma cells, lymphocytes, eosinophils and neutrophils together with myofibroblastic proliferation. As well as in the liver, the liver this lesion has been described in spleen [10], lymph node [11], lung [12], stomach [13], thyroid [14], skin and other sites.

Irrespective of the nomenclature, the condition remains rare. More importantly, the reported long-term outcome for these patients when managed by surgical excision and biliary-enteric anastomosis appears to be excellent [2,5,7]. Only one report has raised the possibility of local or distant ‘recurrence’ with long-term follow-up [9].

In conclusion, benign extrahepatic biliary strictures are rare, and benign inflammatory pseudotumour is a small subset of this cause of obstructive jaundice. Clinicians must remain vigilant and aware of the possibility of a benign cause for the cholangiogram showing obstruction due to a presumed classical Klatskin tumour. Surgical excision of these lesions appears to carry a very good long-term outcome and avoids the recurrent problems associated with ‘palliative’ intubational stenting techniques.

References