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CASE REPORT

Intrahepatic Cholangiocarcinoma Presenting with Spontaneous Rupture

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ABSTRACT

Spontaneous non-traumatic hepatic rupture is an uncommon condition and most often due to underlying hypervascular hepatic lesions. Cholangiocarcinomas are usually hypovascular and spontaneous rupture is extremely rare, very few such cases having been documented in the literature. We present here a patient with spontaneous hepatic rupture secondary to intrahepatic cholangiocarcinoma.

Key Words: *Bile ducts, intrahepatic; Chemoembolization, therapeutic; Choangiocarcinoma; Hemorrhage; Rupture, spontaneous*

中文摘要

肝內膽管癌的自發性破裂

賈亦尊、梁禮賢、朱志揚、尹宇瀚、鄧偉倫

非創傷性的自發性肝破裂很少見，最常見的病因是其中隱含著高血供型的肝病變。膽管癌一般缺乏血供，所以自發性破裂極為罕見，而關於此病的報導亦相當少。本文報告一名肝內膽管癌患者併發自發性的肝破裂。

INTRODUCTION

Spontaneous non-traumatic hepatic rupture is uncommon and most often due to underlying hepatic lesions. Among these, the most frequent are hypervascular tumours such as hepatocellular carcinomas (HCCs), hepatic adenomas, and less commonly hypervascular liver secondaries.[1] Cholangiocarcinomas are usually hypovascular, thus resulting in spontaneous rupture is extremely rare and only a few such cases having been documented in the literature. We present here a patient with spontaneous hepatic rupture secondary to intrahepatic cholangiocarcinoma. This is of particular interest locally, because cholangiocarcinoma has a relatively high incidence in South East Asia.

CASE REPORT

A 62-year-old female with good past health was referred to the surgical outpatient department in 2003 for investigation of per rectal bleeding and unexplained weight loss. Physical examination was unremarkable, as were colonoscopy and oesophagogastroduodenoscopy. Abdominal ultrasound revealed a large solid mass in right lobe of liver. Computed tomography (CT) of the abdomen showed a large tumour with marked increase in vascularity occupying most of the right lobe and medial segment of left lobe of liver (Figure

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1a, 1b). Also there was evidence of right portal vein thrombosis and presence of subcapsular fluid adjacent to the tumour. The presumptive diagnosis was HCC with impending rupture. The patient then underwent celiac, superior mesenteric and hepatic angiography, which showed a large area of hypervascularity in right lobe of liver (Figure 1c). Embolisation of the common hepatic artery using gelfoam was performed. Post-embolisation angiography demonstrated successful embolisation of the tumour. A follow-up CT scan 10 days after embolisation demonstrated a large hypodense mass with central necrosis occupying almost the entire right lobe of liver. A rim of subcapsular fluid with increased density and stranding of the adjacent fat was seen around the tumour, which suggested recent tumour rupture with subcapsular haemorrhage (Figure 1d). The patient then underwent surgery for staging and right hemihepatectomy. Operation revealed a huge tumour involving the entire right lobe and part of caudate lobe. There was no evidence of liver cirrhosis or peritoneal metastases.

Grossly, the resected tumour was an 11-cm necrotic mass, with invasion into adjacent omental fat and extension to the raw resection surface. Microscopically, sections showed moderately differentiated adenocarcinoma with extensive necrosis. The tumour rupture site was covered by fibrinoid exudate (Figure 2a). Tumour cells were arranged in irregular glandular structures. Immunohistochemically, the cells expressed

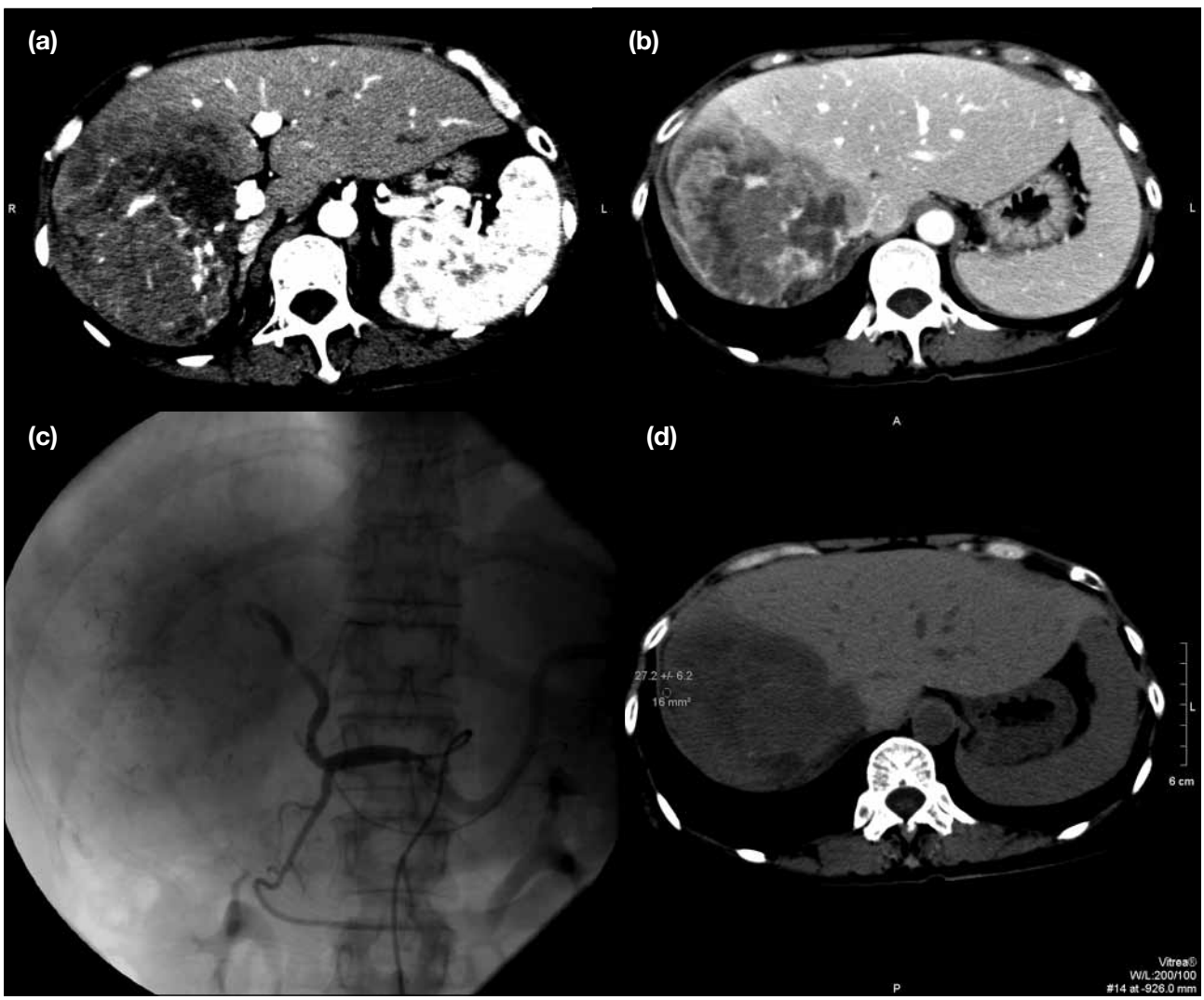

Figure 1. Post-contrast axial computed tomographic (CT) scan of the liver in (a) the arterial phase and (b) the portovenous phase showing a large hypervascular mass lesion in the right lobe and medial segment of the left lobe of the liver. (c) Conventional angiography with cannulation of the common hepatic artery shows an area of hypervascularity in the right lobe of the liver. (d) Non-contrast CT shows a rim of subcapsular fluid with a slight increase in density, suggestive of subcapsular haemorrhage.

CK7 and CK19 (markers for bile duct differentiation) [Figure 2b and 2c]. They did not express HepPar-1 (marker for hepatocytic differentiation) [Figure 2d] or CK20 (marker to suggest colorectal origin). These features were consistent with cholangiocarcinoma.

Postoperatively, the patient developed multiple complications and succumbed about 1 month after the operation.

DISCUSSION

Spontaneous hepatic rupture is an uncommon life-threatening condition. Common culprits include HCC, hepatic adenoma, focal nodular hyperplasia, haemangioma, and metastasis. Other conditions such as amyloidosis, connective tissue disease and infections can also result in hepatic rupture in the absence of underlying tumour.[1]

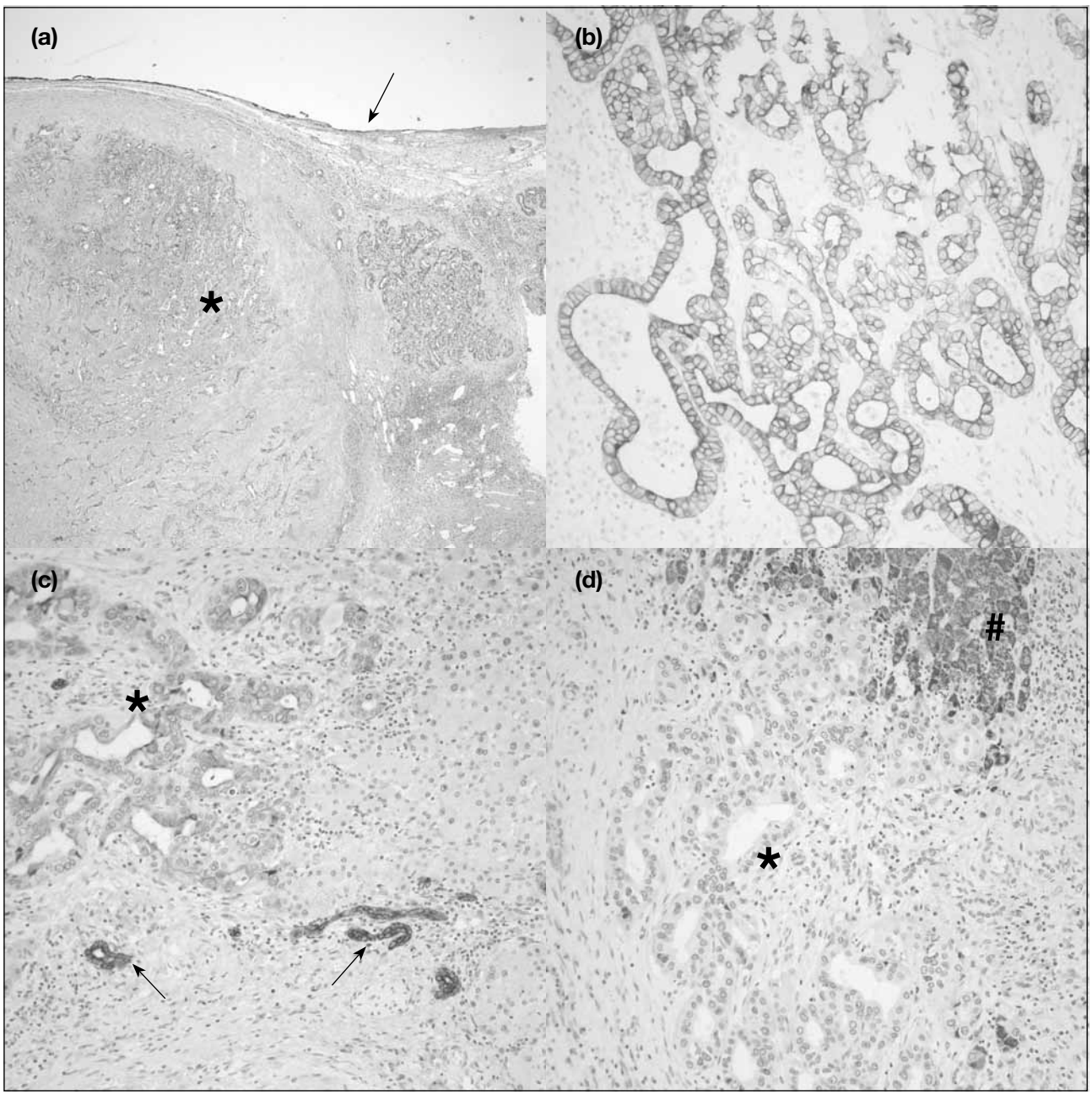

Figure 2. (a) Histology shows the tumour (*) with rupture site covered by fibrinous exudate (arrow). (b) Immunohistochemical staining shows the tumour to be CK7 positive. (c) CK19 immunohistochemical staining shows bile duct differentiation in the tumour (*). Normal intrahepatic ducts are also positively stained (arrows). (d) HepPar-1 stain reveals negative staining of the tumour (*), while the normal hepatocytes (#) are positively stained.

Cholangiocarcinoma is a malignant tumour arising from the epithelium of bile ducts. Although it is the second most common liver cancer after HCC, it remains relatively rare worldwide, accounting for about 5 to 30% of all liver cancers in most areas. Its annual incidence in the United States and the United Kingdom is about 1 to 2 per 100 000 inhabitants.[2,3] It is, however, relatively more common in certain parts of South East and Far East Asia, where infestation by liver flukes and choledocholithiasis was thought to be relatively common.

Cholangiocarcinoma is usually classified into intrahepatic and extrahepatic, while the former is further classified as either peripheral or hilar. Cholangiocarcinoma can also be categorised on a morphological basis into mass forming, periductal infiltrating, and intraductal growing.

Usual clinical presentations of extrahepatic cholangiocarcinoma are jaundice, abdominal discomfort, anorexia, weight loss, and fever.[4,5] Intrahepatic cholangiocarcinomas are usually asymptomatic until late in the course of disease. It is extremely rare for cholangiocarcinoma to present with rupture.[6,7] In which case, it may give rise to acute abdominal pain and upper gastrointestinal bleeding. As in our case, some patients may complain of less specific symptoms and be diagnosed incidentally.

A search of the literature revealed only 2 reported cases of ruptured peripheral cholangiocarcinoma. Akatsu et al[6] suggested that factors such as a peripheral tumour location and presence of venous obstruction can increase the chance of tumour rupture. Both of these factors were present in our patient, in whom the tumour showed cells arranging in glandular structures. This was similar to the case reported by Chong et al,[7] but differed from the predominant papillary pattern described by Akatsu et al.[6] Nevertheless, all 3 cases did not demonstrate the typical hard texture and abundant fibrous stroma of cholangiocarcinoma, which is believed to account for the distinctively low rupture rate of this tumour.

As there are very few reports on ruptured cholangiocarcinoma, it is generally believed that its management should follow guidelines used for ruptured HCC. These entail immediate treatment to achieve haemostasis whilst preserving as much functioning liver as possible, and may include conservative management, surgical haemostasis, transcatheter arterial embolisation (TAE), and open surgery.[8,9] Conservative management is reserved only for patients who are stable at presentation or already terminal. Many of the former may subsequently be deemed suitable for other treatment options if conservative management fails. In some centres, emergency hepatectomy is recommended as a definitive treatment in the acute setting,[10] but has a relative high mortality (reported to be 35% in one review[11]). Common procedures to attain haemostasis surgically include: hepatic artery ligation, perihepatic packing, suturing of the bleeding tumour, and resection of the ruptured component.[8,9] Most of these procedures can achieve a high rate of haemostasis but are associated with a high in-hospital mortality.[8,12] TAE has been shown to be an effective immediate treatment for ruptured HCC. It has a high success rate (53-100%) and a relative low 30-day mortality rate (0-37%).[8] As demonstrated by our case, emergency TAE can also avoid the need for emergency resection, and allow time for staging, neoadjuvant therapy, and ultimately a better planned operation. As compared with one-stage emergency liver resection, staged liver resection has a much lower in-hospital mortality rate (0-9%) and higher 1-year survival rate (54-100%).[8,13,14] Notably, mortality of ruptured HCC is not dependant on the modality of treatment, rather it was largely determined by the pre-rupture disease state, including liver function at the time of rupture as well as the severity of haemorrhage from rupture.[9] In some patients at high risk for surgery, TAE can achieve haemostasis without a major operation. The most common complication of TAE is post-embolisation syndrome (26-85%),[15] which consists of fever, abdominal pain, and elevated liver enzymes. The syndrome usually resolves within 1 to 2 weeks. The major life-threatening complication is liver failure, which ensues in 12 to 29% of TAE-treated patients and is the most common cause of death.[9,16]

CONCLUSION

This report describes an uncommon case of cholangiocarcinoma presenting with rupture. Clinicians should be aware of this rare and potentially fatal condition, especially in patients with compatible clinical signs and symptoms. Immediate management of the rupture should follow that as for ruptured HCC, one of the recognised effective options being TAE.

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