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Case Report

Cystic tumours of the pancreas – the importance of correct diagnosis and treatment

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Cystic tumours of the pancreas are relatively rare and are frequently misdiagnosed as pancreatic pseudocysts. The three cases described below demonstrate some of the typical presenting features of these lesions and emphasise the importance of correct diagnosis and treatment.

CASE REPORTS

Case 1

A 35 year old previously healthy woman presented with left subcostal pain. Ultrasound revealed a 10 cm cystic mass of the pancreas. This was confirmed by CT scan. CT-guided aspiration revealed a high amylase content. ERCP revealed normal pancreatic and bile ducts. The lesion was thought to be a pseudocyst and endoscopic drainage with stent insertion was performed. However, the lesion persisted. Surgical drainage (cystgastrostomy) was then performed. Biopsy of the cyst wall revealed gross dysplasia, highly suggestive of cystadenocarcinoma.

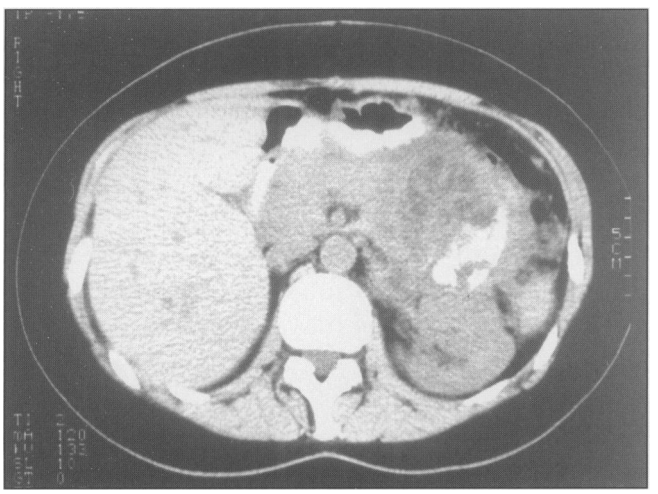

Fig 1. CT scan demonstrating a mucinous cystadenocarcinoma of the pancreas. Note the solid components. The contrast material within the cyst is due to the previous surgical drainage (cystgastrostomy), following misinterpretation as a pseudocyst.

She was transferred to our unit for further management. Repeat CT scan showed debris and fluid in the cyst cavity (Figure 1). In view of possible malignancy and the fact that the cyst was refractory to percutaneous, endoscopic and surgical drainage, resection (distal pancreatectomy and splenectomy) was undertaken. Histology confirmed a mucinous cystadenocarcinoma of the pancreas. She remains well 3 years post surgery.

Case 2

A 40 year old female presented with a history of epigastric pain radiating to the back. Of note, she had an attack of pancreatitis four years previously when seven months pregnant. On this admission investigation revealed a pancreatic cystic mass which was subsequently treated by cyst-gastrostomy for a presumed pseudocyst. The pseudocyst did not resolve and she was transferred to our unit.

Enhanced CT scan showed a cystic mass in the tail of the pancreas (Figure 2). ERCP revealed a normal pancreatic duct. Distal pancreatectomy and splenectomy was performed. Histology confirmed a benign mucinous cystadenoma of the pancreas. She remains well two years post surgery.

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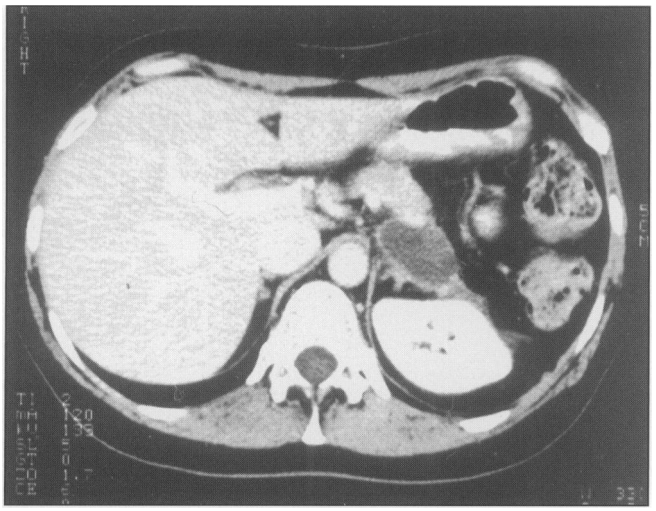

Fig 2. CT scan demonstrating a cystic mass (mucinous cystadenoma) in the tail of the pancreas.

Case 3

A 35 year old previously healthy housewife presented with a four month history of epigastric pain. Ultrasound revealed a 4.5 cm cystic mass in the distal body and tail of the pancreas. CT scan confirmed this. Surgical resection was planned but deferred as she became pregnant. Post-partum repeat CT scan showed the cyst had increased to 6 cm (Figure 3) and resection was performed (distal pancreatectomy and splenectomy). Histology confirmed a serous cystadenoma of the pancreas. She remains well one year post surgery.

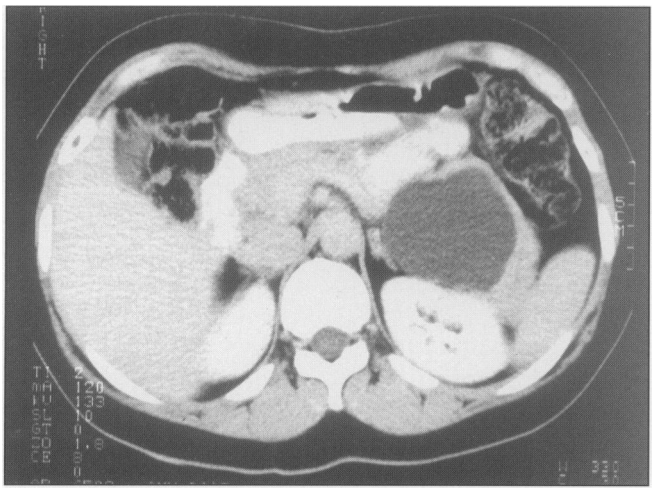

Fig 3. CT scan demonstrating a serous cystadenoma of the pancreas. Note the unilocular appearance with absence of any solid components.

DISCUSSION

When a cystic lesion of the pancreas is encountered the first priority is to determine whether it is a pseudocyst or a neoplasm.[1] The majority of these lesions are pseudocysts, which are of inflammatory or traumatic origin. Pseudocysts are collections of pancreatic secretions surrounded by a fibrous wall with no epithelial lining. Neoplasms account for 10-15% of pancreatic cystic lesions.[2,3] Ninety percent of neoplastic cysts are of epithelial origin. The commonest epithelial tumours are benign cystadenomas (serous or mucinous) and cystadenocarcinomas, which collectively account for 75% of all cystic tumours. Recently defined intraductal papillary and mucinous tumours represent 11% of cystic tumours whilst pseudopapillary tumours represent 4%.[4]

The definitive diagnosis of a pancreatic cystic lesion is often difficult. A history of antecedent factors or events that could generate a pseudocyst, such as pancreatitis or trauma, is extremely important. In the absence of such a history a pancreatic lesion should not be labelled as a pseudocyst and a neoplasm should be suspected. Cystic neoplasms are most commonly found in middle-aged women although the benign serous cystadenoma often occurs in elderly women. Abdominal pain is the most common symptom as our three cases demonstrate. Weight loss is also a significant feature.[3] Serum amylase level is usually normal in patients with a neoplasm but in 50-75% of patients with a pseudocyst it is increased.[3]

Some physical characteristics are helpful in diagnosis. Loculation and solid components on CT scan are indicative of a neoplasm. Calcification is common in cystic tumours. Hypervascularity on arteriography may indicate a neoplasm but not its type.[5,6] Size and location are not of much diagnostic value although most neoplasms occur in the body and tail of the pancreas.[6,12] ERCP usually demonstrates no communication between cystic tumours and the pancreatic duct, as in our cases, but communication with the ductal system is often found in pseudocysts.[3,7,8]

Percutaneous aspiration of cystic tumours for measurement of amylase, CEA, CA 19.9 and for cytology has not been evaluated sufficiently to allow determination of how sensitive or reliable these parameters might be for diagnosis of a neoplasm.[6] Cystic aspiration has been limited by the theoretical concern of spillage and seeding of tumour cells. Biopsy of the cyst wall, at the time of surgery, will usually make the differentiation between pseudocyst and neoplasm although the

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absence of an epithelial lining in a limited biopsy does not preclude the diagnosis of a cystic tumour. In cases where surgical drainage of a pseudocyst is undertaken biopsy of the wall should always be performed if there is any clinical or radiological doubt about the diagnosis.

Although discrimination of cystic neoplasms from pseudocyts may be difficult it is imperative to make this distinction because the treatment for each is obviously different. Internal drainage is the treatment of choice for an uncomplicated mature pseudocyst.[9] Surgical options include drainage via the stomach, duodenum or jejunum. Pseudocysts can be drained endoscopically (with endosonography) via the transpapillary approach or transmurally (endoscopic cystgastrostomy or cystduodenostomy) . The former is preferred if cystenterostomy is not possible, as in the absence of cyst-gut wall apposition, and if pancreatic duct morphology identifies cyst-duct communication.[10, 11] However, as our first case demonstrated, percutaneous or internal drainage of a misdiagnosed 'pseudocyst' will fail to alleviate symptoms, may convert a sterile cystic tumour to an infected one or, more importantly, leave behind a curable cancer.[2, 5] This problem is likely to increase with the increasing use of endoscopic drainage techniques as the definitive management of pseudocysts.

The treatment of epithelial cystic tumours of the pancreas usually involves surgical resection. One exception is for unilocular benign serous cystadenomas in asymptomatic elderly patients, where conservative management is justified provided there is no pancreatic duct or vascular obstruction.[4, 12] Such patients should be followed by yearly ultrasonography. All other cystic tumours have the potential for malignant degeneration or may be malignant at the time of diagnosis and hence should be managed by resection.[1, 6, 13, 14] The majority of cystadenocarcinomas of the pancreas (up to 70%) are resectable [4, 6] as these tumours can be exceedingly slow growing, indolent and tend not to invade adjacent structures.[3] These factors and the relatively low incidence of metastasis permits curative surgery, even after previous drainage or bypass procedures.[3] Resection should be attempted even in advanced cases as the five year survival rate may exceed fifty percent.[3, 4] Hence, large locally invasive tumours should not be dismissed as inoperable without evaluation.[4] Neither tumour size nor previous intervention should preclude an attempt at curative resection.[3] Removal of an intact cyst should be the aim of operative resection because cystic rupture may disseminate malignant cells intraperitoneally.[1, 15]

In conclusion, cystic tumours of the pancreas are rare. Failure to recognize the true nature of such a tumour will lead to misdiagnosis as a pseudocyst and incorrect treatment. Given the difficulties of accurate pre-operative diagnosis, the high incidence of potential malignancy and the good outcome with resection, a review of the literature suggests that all suspected cystic tumours of the pancreas (apart from asymptomatic serous cystadenomas in elderly patients) should be managed primarily by resection.

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