Pancreatic cystic neoplastic disease with unusual clinical findings: a potential diagnostic dilemma

P. F. McGowan, G. R. Williams and R. G. Postier
Department of Surgery, University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma, USA

Three patients with cystic neoplastic disease of the pancreas who presented over a 6-year period are described. In contrast to the usual situation, these neoplasms were associated with underlying pancreatic disease or were in direct communication with the pancreatic drainage system. All were managed by pancreatic resection. One patient succumbed from locally invasive disease 15 months after surgery, whereas the others were free of disease at 30 and 56 months respectively following resection. Recognition of these tumours is essential, particularly as the macrocystic variety is inherently malignant and should be treated by excision.

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Most pancreatic cysts encountered in clinical practice are pseudocysts but pancreatic cystic neoplasms account for 5–10% of the total seen. Though it is widely reported that cystic neoplasms of the pancreas are neither associated with underlying pancreatic disease nor communicate with the pancreatic drainage system, they not uncommonly are mistaken for pseudocyst disease and are managed accordingly. Pancreatic cystic neoplasms are conveniently subdivided into two separate entities each with quite different histological and prognostic characteristics. One group, termed microcystic adenoma of the pancreas, consists predominantly of small cysts lined by flattened or cuboidal cells whose cytoplasm contains glycogen and little or no mucin. These tumours are benign. The second group, termed mucinous cystadenoma-cystadenocarcinoma of the pancreas (macrocystic pancreatic neoplasm) consists of large unilocular or multilocular cysts lined by tall columnar mucus-secreting epithelium which often displays a papillary morphology. The mucinous tumour is an aggressive neoplasm that invariably becomes malignant when left to its own course, though it may remain asymptomatic and benign over a long period.

Case reports

Case 1

A 60-year-old man presented in July 1985 with a 3-month history of epigastric pain and gastric flatulence. Previous history was non-contributory. On examination, a palpable slightly tender non-mobile cystic mass was present within the epigastrium. Laboratory investigations were entirely normal. Abdominal computed tomographic scanning identified two pancreatic lesions, one within the head and the second within the tail of the gland respectively. The former was suggestive of cystic neoplastic disease whereas the latter resembled a pseudocyst. On subsequent exploration, both cystic lesions were readily identified. There was no evidence of metastatic disease and the remainder of the laparatomy was normal. A Whipple’s procedure was employed in the removal of the proximal lesion whereas the distal lesion was simply excised with a surrounding rim of pancreatic parenchyma. On sectioning the proximal specimen, the head of the pancreas was seen to be virtually replaced by a mucus-filled unilocular cyst (7.0 × 5.0 × 3.0 cm) whose lining clearly elaborated a papillary morphology. Microscopically, it was a well differentiated macrocystic pancreatic neoplasm. The peripancreatic lymph nodes contained tumour metastases. Sections of pancreatic parenchyma free of tumour demonstrated histological features of chronic pancreatitis. Histological examination of the distal lesion confirmed that it was a pseudocyst. The patient eventually succumbed from locally invasive disease 15 months after surgery.

Case 2

A 33-year-old woman presented to another institution in December 1984 with a 10-month history of epigastric pain, abdominal swelling and weight loss. Previous history was non-contributory. On examination a palpable cystic mass was present within the epigastrium. A barium study showed that the stomach was displaced anteriorly by a large lesser sac swelling which was believed to be a pseudocyst. Percutaneous drain age was instituted and 1600 ml of a chocolate-coloured fluid rich in amylase (17 000 units/litre) was withdrawn within the first 24 h alone. Because the cyst failed to resolve, the patient was transferred to this institution for further management. Endoscopic retrograde pancreatography was performed which outlined a large cystic lesion that was clearly seen to communicate with the middle third of the pancreatic duct. Calcified...
gallstones were noted within the gallbladder. The preoperative diagnosis was that of pancreatic pseudocyst disease. At exploration, a large cantaloupe-sized mass (still regarded as a pseudocyst) was identified within the lesser sac. Having removed the calculous gallbladder, it was elected to manage the so-called ‘pseudocyst’ by performing a cystogastrotomy. Having made the posterior gastrostomy, however, it became clear that large papillary fronds were present within the lumen and only then was the true nature of the lesion suspected. Frozen section biopsy of the wall confirmed its cystic neoplastic nature and accordingly a 95% distal pancreatectomy was performed. Macroscopically, it was a thick-walled multiloculated cyst (17 × 15 × 5 cm) which contained numerous papillary fronds within a dark mucoid material. Microscopically, it proved to be a well-differentiated macrocystic tumour without evidence of invasion. It is now 2 years since excision and the patient is clinically free of disease and has long since regained her normal weight.

Case 3

A 63-year-old man (non-drinker) was admitted in October 1982 with biochemically proven pancreatitis which subsequently became complicated by pseudocyst disease within the pancreatic head. Serial ultrasound examinations demonstrated complete resolution of the pseudocyst over an 8-week period. Fifteen months later, he was re-admitted with pancreatitis. When the initial inflammation had subsided, endoscopic retrograde pancreatography was performed which outlined a cystic lesion within the pancreatic tail that clearly was in communication with the pancreatic duct. Calcified gallstones were also noted within the gallbladder during this procedure. The pancreatic lesion was regarded as a pseudocyst and close follow-up by serial ultrasonography was advised, but on this occasion the patient was unwilling to comply with our recommendations. Because of persistent abdominal pain, however, he returned 4 months later and was re-admitted for renewed investigation and management. Physical examination at this time was unremarkable apart from slight left upper quadrant tenderness. Laboratory investigations (including serum amylase) were normal. Endoscopic retrograde pancreatography was again performed and on this occasion showed that the distal pancreatic duct was extrinsically compressed and distorted apparently by the aforementioned space-occupying lesion with which it was no longer in communication. The lesion was now regarded as a cystic neoplasm and exploration was advised. At laparotomy, the calculous gallbladder was removed. The pancreas felt hard and nodular and a localized cystic swelling was identified within the tail of the gland. There was no evidence of metastatic disease and accordingly a 45% distal pancreatectomy was performed thereby removing the lesion. Macroscopically, the tumour was a unicellular cyst (7 × 4 × 3 cm) that contained multiple papillary fronds and tumour excrescences within a mucus-filled lumen. Microscopically, it had the typical features of a macrocystic tumour without evidence of malignant transformation. The remainder of the resected material showed histological features of chronic pancreatitis. It is now almost 3 years since excision and the patient is totally asymptomatic and free of disease.

Discussion

The vast majority of pancreatic cysts encountered in clinical practice are pseudocysts but pancreatic cystic neoplasms account for between 5 and 10% of the total seen. The symptomatology associated with these cystic neoplasms is non-specific and is usually due to compression of adjacent structures. The most common symptom is epigastric or left upper quadrant pain often accompanied by a palpable abdominal mass. Though pancreatic cystic neoplasms occur predominantly in females in otherwise normal glands, confusion with pseudocyst disease is not uncommon, often resulting in inappropriate management. Since the advent of computed tomographic and ultrasonographic scanning, however, the identification of cystic neoplasms pre-operatively has become much more common. These sophisticated techniques can identify specific cystic neoplastic characteristics such as the presence of multiple septa and/or intracystic tumour excrescences which are altogether absent in pseudocyst disease.

The management of pancreatic cystic neoplasms, particularly with regard to the macrocystic variety, is surgical resection of the tumour with a surrounding margin of healthy pancreatic tissue. Even when malignant transformation has occurred within a macrocystic tumour, the 5-year survival following excision approached 70% whereas without resection or in the presence of metastatic disease only 30% are alive at 2 years. Since these lesions for the most part are well circumscribed and have a definite capsule, most are easily resectable. Whipple’s procedure is adequate for lesions in the head and neck whereas distal pancreatectomy will suffice for lesions within the pancreatic tail. Since the microcystic adenoma (benign lesion) usually presents in the elderly female and is located most commonly in the pancreatic head, its removal may not always be indicated, particularly if such a patient happens to be a poor surgical risk or is asymptomatic. In contrast, the macrocystic tumour, which presents most commonly in the 40-60-year-old woman, should be resected because of its overt or latent malignancy.

The coexistence of a pancreatic cystic neoplasm in a gland with underlying pancreatic disease (such as pancreatitis or pseudocyst disease) is extremely unusual (cases 1 and 3) and to date only similar cases have been reported in the English literature. It is also uncommon for such a lesion to communicate with the pancreatic drainage system as was the situation in two of our patients (cases 2 and 3). In the light of this experience, it is now our policy to have all cysts in relation to the pancreas examined by ultrasonographic or computed tomographic scanning before embarking on any ‘proposed’ surgical treatment. If the results of
these examinations are equivocal (particularly with regard to cystic neoplastic disease) or if a lesion exhibits all the hallmarks of a typical unresolved mature pseudocyst, then at surgery we submit a portion of the cyst wall for frozen section (biopsy) to establish the precise diagnosis12. Using this approach, it is unlikely that any cystic neoplasm will be mistaken for pseudocyst disease and inappropriately drained. Recognition of pancreatic cystic neoplasms is the key to correct management as early appropriate treatment, particularly with regard to the macrocystic tumour, offers the best chance of cure to these patients.

References


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