CLINICAL CASE

Pancreatocolonic fistula complicating noninvasive intraductal papillary mucinous tumor of the pancreas

Fistule pancréaticocolique compliquant une tumeur intracanalaire papillaire et mucineuse du pancréas (TIPMP) non invasive

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Summary

Intraductal papillary mucinous tumors (IPMT) of the pancreas are a distinct clinicopathological entity that is increasingly recognized and whose natural history and clinical presentation are now better understood. Nevertheless, only rare cases of pancreatobiliary or pancreatodigestive fistulas complicating IPMT have been described so far and their clinicopathological significance and association with cancer remain controversial. We report a case of pancreatocolonic fistula complicating a noninvasive IPMT, and review the published literature. Unlike previous reports, IPMT complicated by fistula in nearby organs does not seem to be more often associated with invasive carcinoma: frequency is comparable in resected IPMT with or without internal fistula. Since fistulas are not a reliable clinicopathological predictor of invasive malignancy, en-bloc resection should not be routinely performed especially if extended resection increases the immediate risks or the long-term risks of surgery.

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Initially described in the 1980s in the Japanese literature, Intraductal Papillary Mucinous Tumor (IPMT) of the pancreas is currently accepted as a distinct entity. Natural history and clinical presentation of IPMT is now well-known [1–3], but only rare cases of pancreatobiliary or pancreatodigestive fistulas complicating IPMT have been described [4–9]. The clinicopathological significance of those internal fistulas remains controversial, and their association with cancer is unclear. We herein report the case of a 65-year-old patient presenting with a noninvasive diffuse IPMT complicated by an isolated pancreaticocolonic fistula.

Case report

A 65-year-old woman, with a medical history of non-insulin-dependent diabetes since 1998, presented in June 2000 with transient epigastric pain, vomiting and unstable diabetes. Abdominal ultrasonography (US) revealed a wide cystic mass involving the whole pancreatic gland. Computed tomography (CT), and endoscopic ultrasonography (EUS) confirmed a clear dilatation of the main pancreatic duct, and no neoplastic cells were found on biopsy performed through EUS-guidance. Because of suspected malignant IPMT, surgical treatment was performed. At laparotomy, there was neither metastasis nor vascular encasement, and the pancreatic tail was closely adherent to the splenic flexure (Figs. 1 and 2). EUS showed several intraductal polyloid lesions, suggestive of malignancy, but no neoplastic cells were found on biopsy performed through EUS-guidance. Of suspected malignant IPMT, surgical treatment was performed. At laparotomy, there was neither metastasis nor vascular encasement, and the pancreatic tail was closely adherent to the splenic flexure. An en-bloc resection of the whole duodenopancreas, the spleen, and the splenic flexure was performed. The continuity was reconstructed with gastrojejunostomy, hepaticojejunostomy and colonocolonic anastomosis. At gross examination, the pancreaticocolonic fistula was confirmed with issue of mucin in the colon (Fig. 3). The patient was discharged on postoperative day 22 after an uneventful recovery. With a 14-month follow-up, the patient is doing well without any sign of recurrence.

Pathological examination confirmed an IPMT invading almost diffusely the main and branched pancreatic ducts. Most of them were dilated and filled by large papillary or pseudopapillary structures lined by a dysplastic multilayered epithelium. Dysplasia was mainly severe and focally moderate (or borderline). No invasive carcinomatous component was present. The pancreatic parenchyma was atrophic and fibrous. The colonic fistula was lined by noninvasive IPMT lesions and the surrounding tissue was fibrous with inflammatory symphysis between pancreas and colon (Fig. 4).

Discussion

IPMT is a distinct clinicopathologic entity, presently recognized with an increasing frequency [1]. Those tumors are characterized by cystic dilatation of the main and/or branch ducts, mucin production, and intraductal papillary growth. Their natural history is now better known, mainly marked by progressive malignant transformation [2]. Indeed, 40 to 50% of resected specimens contain invasive carcinoma [2,3]. Internal fistula is a very rare feature of IPMT with less than 50 cases collected in a review published in 2005 [4]. Fistulas with surrounding organs involves, in decreasing order of frequency, duodenum [5,6], bile duct [5,7,8], stomach [4–6,9], or exceptionally pleural cavity. In about 30% of cases, internal fistulas are multiple [4,5,9]. To our best knowledge, only one case of pancreaticocolonic fistula has been previously described, and was associated with both pancreatogastric and CA 19.9 slightly increased: 49 IU/mL (normal < 37). CT scan and magnetic resonance cholangiopancreatography showed that the main pancreatic duct was globally dilated, with a maximum diameter of 36 mm, and revealed the presence of an internal fistula between the pancreatic tail and the splenic colon (Figs. 1 and 2). EUS showed several intraductal polyloid lesions, suggestive of malignancy, but no neoplastic cells were found on biopsy performed through EUS-guidance. Because of suspected malignant IPMT, surgical treatment was performed. At laparotomy, there was neither metastasis nor vascular encasement, and the pancreatic tail was closely adherent to the splenic flexure. An en-bloc resection of the whole duodenopancreas, the spleen, and the splenic flexure was performed. The continuity was reconstructed with gastrojejunostomy, hepaticojejunostomy and colonocolonic anastomosis. At gross examination, the pancreaticocolonic fistula was confirmed with issue of mucin in the colon (Fig. 3). The patient was discharged on postoperative day 22 after an uneventful recovery. With a 14-month follow-up, the patient is doing well without any sign of recurrence.

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Figure 1  CT scan showing a pancreaticocolonic fistula (yellow arrow) located between the dilated main pancreatic duct (white arrow) and the colonic lumen (white star). Scanner montrant une fistule pancréaticocolique (flèche jaune) située entre le canal pancréatique principal dilaté (flèche blanche) et la lumière colique (étoile blanche).
Pancreatocolonic fistula complicating noninvasive IPMT

It has been suggested that IPMT complicated by internal fistula was almost always associated with an invasive carcinoma [5,6]. Indeed, only around 50% of fistulas are associated with invasive cancer [4,9], which is comparable to cancer frequency in resected IPMT specimens free of fistula formation [1,3]. Moreover, even in the case of malignant IPMT, the fistula and its surrounding tissues are free of neoplastic infiltration in half of the cases [7], as in the previously published case of pancreatocolonic fistula [6]. In this latter case, malignant IPMT was associated with a non-neoplastic colonic fistula [6]. Physiopathology of non-neoplastic fistulas remains controversial, and could be related to mechanical phenomena such as high intraductal pressure due to mucin retention and inflammatory changes [9], as previously described in some cases of mucinous cystic neoplasm [4].

The present case of noninvasive IPMT complicated by a non-neoplastic pancreatocolonic fistula confirms the difficulty to establish preoperatively a diagnosis of invasive malignancy. Since fistula per se is not a reliable clinicopathological predictor of invasive malignancy, we advocate that an en-bloc resection should be performed only if malignancy is histologically proven, particularly if extension of resection increases the immediate risk or long-term drawbacks of surgery. Otherwise extent of resection should be the same than for a nonfistulised IPMT, without need for carcinologic resection of fistula. Since frozen section is reliable to grade severity of IPMT [10], it should be used to determine if a more radical resection is needed. Until now, internal fistula complicating IPMT have not been studied as a predictive factor of malignancy and long-term survival. Further studies are necessary to clarify the pathogenesis, significance and appropriate treatments of IPMT with internal fistula.
References


