Case Report

Total Pancreatectomy for Malignant Intraductal Papillary Mucinous Neoplasm (IPMN) Complicated by Gastropancreatic Fistulae

Oshan Basnayake, Pradeep Wijerathne, Umesh Jayarajah, Nilesh Fernandopulle, and Sivasuriya Sivaganesh

1Professorial Surgical Unit, National Hospital of Sri Lanka, Colombo, Sri Lanka
2Department of Surgery, Faculty of Medicine, University of Colombo, Colombo, Sri Lanka

Correspondence should be addressed to Sivasuriya Sivaganesh; sivaganesh@srg.cmb.ac.lk

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Background. Intraductal papillary mucinous neoplasms (IPMN) of the pancreas complicated by fistula formation to adjacent organs are an uncommon phenomenon. We present an IPMN of the pancreas with malignant transformation and multiple fistulae to the stomach and duodenum.

Case Presentation. A 50-year-old female was referred for investigation of recent epigastric pain and a past history of recurrent pancreatitis. Imaging with computed tomography showed a gross dilatation of the entire pancreatic duct with a heterogeneous enhancement of the periductal parenchyma. A passage of oral contrast was noted from the greater curvature and pylorus of the stomach into the dilated duct suggestive of fistulae formation. Gastroduodenoscopy demonstrated these fistulae in the stomach and the proximal duodenum and an exophytic growth at the ampulla obliterating the view of ampullary opening. Endosonography- (EUS-) guided fine-needle aspiration cytology (FNAC) showed cells with high-grade atypia. A total pancreatectomy, distal gastrectomy, and splenectomy were performed, and recovery was uneventful. Histology revealed a ductal adenocarcinoma arising from an intestinal type intraductal papillary mucinous neoplasm with high-grade dysplasia. A year and a half after surgery, she is healthy with good glycaemic control and nutritional status.

Conclusion. This case highlights the importance investigating patients for the aetiology in recurrent acute pancreatitis and their follow-up. Awareness of cystic pancreatic neoplasms including IPMN is important to avoid misdiagnosis or delayed diagnosis. Referral of these patients to centres with facilities for multidisciplinary input and specialised management is strongly recommended.

1. Introduction

Intraductal papillary mucinous neoplasms (IPMN) of the pancreas are cystic neoplasms composed mainly of mucous-secreting columnar cells with variable cellular atypia [1]. The usual age of presentation is between the fifth and seventh decades with equal distribution among the sexes and a prevalence of 2-4% in general population [1]. Depending on the duct involvement, these tumours are classified as main duct type, branch duct type, or mixed type. Histologically, these tumours are classified into intestinal, pancreaticobiliary, oncocytic, and gastric types. The risk of invasive carcinoma in high-risk IPMN was 12% over a mean follow-up period of 60 months [2]. Fistula formation to adjacent organs in IPMN with invasive carcinoma is an uncommon phenomenon. A retrospective study using computed tomography (CT) and magnetic resonance imaging (MRI) of 423 patients with IPMN showed fistulae involving the duodenum, stomach, common bile duct, and colon in 1.9% (n = 8). Furthermore, fistulae appeared to predominate in malignant main duct IPMN [3]. We present a case of a colloid type ductal adenocarcinoma of intestinal variety occurring...
in the background of IPMN with multiple fistulae to the
greater curvature of the stomach, pylorus, and duodenum.

2. Case Presentation

A 50-year-old female was referred for investigation of
recurrent epigastric pain. She was otherwise well with no
constitutional symptoms, anorexia, or weight loss and was
normoglycaemic. She had a past history of recurrent pancre-
atitis with elevated amylase levels. The liver enzymes and
abdominal ultrasonographic studies were normal. She has
been asymptomatic for the preceding 3 years. A contrast-
enhanced CT scan which was performed 3 years ago had
been unremarkable except for evidence of acute pancreatitis
and a 6 mm main pancreatic duct dilatation. The contrast-
enhanced CT showed a gross dilatation of the entire pancre-
atic duct (diameter = 23 mm) with a heterogeneous enhance-
ment of the periductal parenchyma of the whole gland with
an irregular outline. A passage of oral contrast was noted
from the greater curvature and pylorus of the stomach into
the dilated duct suggestive of fistulae formation (Figure 1).
Gastroduodenoscopy demonstrated these fistulae in the
stomach and also the proximal duodenum and exophytic
growth at the ampulla obliterating the view of ampullary
opening. Endosonography (EUS) revealed a mixed echogenic
collection in relation to pancreatic head and body (Figure 2),
and the EUS-guided fine-needle aspiration cytology (FNAC)
showed cells with high-grade atypia. The volume of aspirate
was inadequate to perform carcinoembryonic antigen
(CEA) and amylase assays. Her preoperative CA 19.9 level
was 21 U/mL, and faecal elastase was not performed.

She underwent a total pancreatectomy, distal gastrec-
tomy, and splenectomy. The fistula in the proximal stomach
was removed with a cuff of posterior gastric wall using a
linear stapler (Figure 3). The postoperative recovery was
uneventful, and glycaemic control was achieved with a
basal-bolus insulin regime. A histological analysis revealed
a ductal adenocarcinoma (colloid type) arising from an intes-
tinal type intraductal papillary mucinous neoplasm with
high-grade dysplasia and pathological stage of pT3N0Mx.
She was prescribed insulin and pancreatic enzyme supple-
mentation, received postsplenectomy vaccination, and is on
penicillin prophylaxis. She received FOLFIRINOX (folinic
acid, fluorouracil, irinotecan, and oxaliplatin) 2 weekly for
12 cycles as adjuvant chemotherapy. At 18 months after sur-
gery, she is disease-free with good glycaemic control and
nutritional status.

3. Discussion and Conclusion

Most patients with IPMN are asymptomatic and are often
diagnosed incidentally. Others present with symptoms of
pancreatitis due to ductal obstruction by mucin or nonspe-
cific abdominal symptoms. Late diagnosis is not uncommon
due to the insidious onset and slow progression of the disease
and also lack of awareness among clinicians who do not
infrequently mistake abnormalities on imaging to be due to
pancreatitis. Proper evaluation of patients with cystic lesions
in pancreas should be considered especially in patients with
late-onset pancreatic type pain. Evaluation includes cross-
sectional imaging, biochemical assay, and cytology of aspi-
rate obtained at endosonography. Contrast CT has an accu-
rlacy of 40-81% [4, 5] while MRI has a higher accuracy of
40-95% but with equivalent specificity [6, 7]. Endosono-
graphic differentiation of benign and malignant lesions is
subject to expertise but has the advantage of obtaining sam-
ple for cytology and biochemistry [8]. The side-viewing
endoscopic appearance of bulging ampulla extruding thick mucus is a characteristic finding of IPMN [8].

IPMN complicated by fistula formation to the adjacent viscera has been reported. The viscera involved include the duodenum, stomach, and bile duct, and 39% were multiple [9]. Direct tumour invasion and mechanical pressure in the setting of noninvasive tumours are proposed mechanisms of pathogenesis of fistula formation [9].

The risk of invasive carcinoma in high-risk IPMN was 12% over a mean follow-up period of 60 months [2]. The established risk factors for malignancy include the presence of jaundice, enhancing mural nodules, solid components, and a main pancreatic duct (MPD) diameter of >10 mm. Because of the time-dependent risk of progression to malignancy, surgery is indicated in high-risk patients [10]. A duct diameter of >10 mm is an absolute indication for surgical
resection of MPD IPMN while a diameter of 5-9.9 mm is considered a relative indication [11]. Options for surgical resection include pancreaticoduodenectomy with intraoperative frozen section assessment of the resection margin and total pancreatectomy. Data on the optimal type of surgical resection for main duct IPMN are conflicting [12]. Patients who undergo pancreaticoduodenectomy require close surveillance for early detection of malignant recurrence followed by completion pancreatectomy when suspected. Relative preservation of pancreatic function is achieved with pancreaticoduodenectomy but at the cost of surveillance. Total pancreatectomy is recommended in patients with involvement of entire MPD because of the elevated risk of high-grade dysplasia and cancer [10]. Loss requiring replacement therapy of both endocrine and exocrine functions is the downside of total pancreatectomy.

Irrespective of lymph node status, adjuvant systemic chemotherapy is often recommended because of the associated aggressive behaviour of these malignancies [13].

4. Conclusion

This case highlights the importance of investigating those with acute relapsing pancreatitis for aetiology and their follow-up. Awareness of cystic pancreatic neoplasms including IPMN is crucial in clinical practice to avoid misdiagnosis or delayed diagnosis. In the management of these patients, a multidisciplinary approach is strongly recommended. This is the first reported case of successful surgical management of a malignant main duct IPMN by total pancreatectomy in Sri Lanka.

Abbreviations

IPMN: Intraductal papillary mucinous neoplasm
EUS: Endosonography
CT: Computed tomography
MRI: Magnetic resonance imaging
CEA: Carcinoembryonic antigen.

Consent

Informed written consent for publication and accompanying images was obtained from the patient prior to collecting information.

Conflicts of Interest

The authors declare that they have no competing interests.

Authors’ Contributions

Authors OB, PW, and UJ contributed to the collection of information and writing of the manuscript. Authors NF and SS contributed to the writing and final approval of the manuscript. All authors read and approved the final version of the manuscript.

References


