Case of mixed - type intraductal papillary - mucinous neoplasm of pancreas highlighting the need of multidisciplinary approach

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Summary

Mixed-Type Intraductal Papillary-Mucinous Neoplasms (MT – IPMNs) of pancreas are rare mucin producing papillary growths involving the Main Pancreatic Duct (MPD) and Branch Ducts (BD). Among other subtypes, MT-IPMNs are the most challenging in determining the ideal therapeutic strategy.

A 56-year-old female presenting with severe acute epigastric abdominal pain due to acute on chronic pancreatitis was diagnosed with a cystic lesion in the pancreatic head by Contrast Enhanced Computed Tomography (CECT) imaging. Further imaging, interventions or follow-up were not offered. The patient presented four years later with a similar presentation to be subjected to pancreaticoduodenectomy following diagnosis of IPMN through Magnetic Resonance Cholangiopancreatogram (MRCP) and Endoscopic Ultrasound (EUS) imaging. The report highlights the necessity for heightened consideration of the role of MRCP as a determinant of the management strategy and the awareness of indications for surgery in MT-IPMNs in the Sri Lankan surgical setting.

Keywords - pancreatic intraductal neoplasms, cholangiopancreatography, magnetic resonance.

Introduction

IPMNs are defined as pre-invasive, intraepithelial neoplasms[1] characterized by ductal dilation, intraductal papillary growth and thick mucus secretion[2] . IPMNs can be differentiated into three groups based on the sites involved: Branch Duct (BD-IPMN), Main Duct (MD-IPMN) and MT-IPMN which includes characteristics of both types[3].

Due to the increasing incidence, management of IPMN is an emerging problem[3]. Magnetic Resonance Imaging (MRI) and Endoscopic Ultrasound (EUS) are primary investigations in diagnosing and following up[2]. To improve the survival of IPMN patients, resection is necessary to prevent progression to invasive disease[4].

Case Report

A 56-year-old female presented with severe acute epigastric abdominal pain radiating to the back, associated with multiple episodes of vomiting in 2016. Persistent fever accompanied her complaints. The patient failed to recall further details of this episode as it had occurred over four years ago. No other findings were significant in her clinical history.

She was extensively investigated and a diagnosis of acute exacerbation of chronic pancreatitis was reached. CECT abdomen with pancreatic protocol had concluded the presence of a single ill-defined cystic lesion of 1.2 x 1.5 cm in the pancreatic head region. Partial obstruction and dilatation of the MPD with antero-posterior diameter of 5.4mm was observed in the proximal body region. The lesion projected into the second part of the duodenum. A differential diagnosis of benign cyst or cystic neoplasm was considered.

However, further imaging or surgical opinion was not taken. Furthermore, no surveillance imaging was performed.

In August 2020, the patient presented with a history of repeated attacks of pancreatitis. Examination revealed epigastric tenderness, but jaundice was not evident.

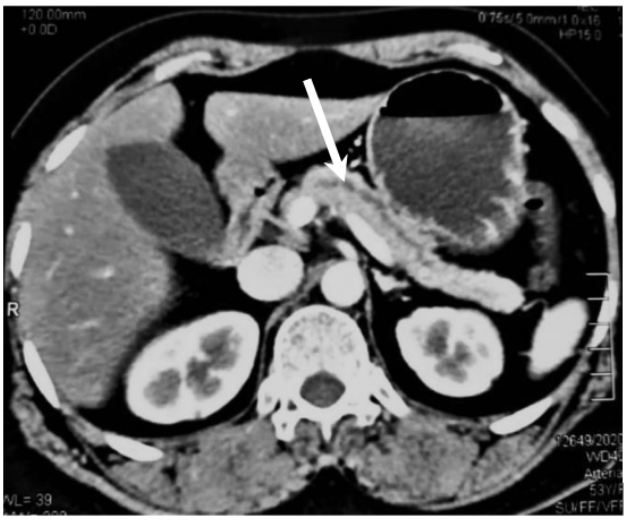

Figure 1 : Axial CT slice of CECT abdomen depicting ill-defined cystic lesion of proximal pancreas and prominent dilatation of MPD (white arrow) (Diameter of 7mm proximally and 5mm distally).

The patient was re-evaluated and the summary of findings of cross-sectional imaging is as follows: CECT of abdomen with pancreatic protocol depicted a cystic lesion of the uncinate process of the pancreas with dimensions of 1.5 x 1.3 x 0.8 cm. Disproportionate dilatation of the MPD was evident. The distal and proximal regions of the duct were 5mm and 7mm in diameter, respectively (Figure 1). Dilated branch ducts were detected in the uncinate process.

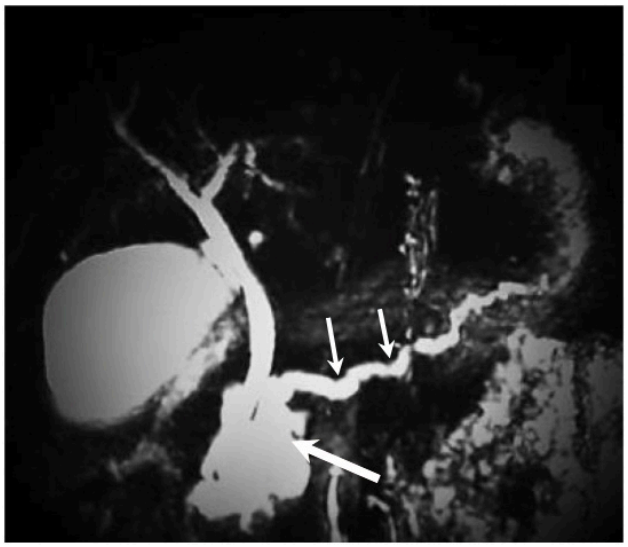

Figure 2: Coronal view of T2-weighted MRCP of the pancreatico-biliary tree indicating high intensity ill-defined multiloculated cystic lesion in the uncinate process region (large arrow) which communicates with the MPD. Dilatation of MPD (small arrows) is clearly visible.

MRCP indicated a T2 weighted high intensity multiloculated cystic lesion in the uncinate process which communicates with the MPD (Figure 2).

The diagnosis was acute on chronic pancreatitis supported by the radiological diagnosis of MT-IPMN in the uncinate process.

The patient was also subjected to an EUS with Fine Needle Aspiration Cytology (FNAC). The case was discussed in a multidisciplinary meeting (MDT) and pancreaticoduodenectomy was offered, given the history of recurrent pancreatitis and MT-IPMN in the uncinate process.

Discussion

IPMNs were formally described initially by Ohashi in 1982[1]. Since then, the evaluation of IPMNs has evolved in classification and management. Different international guidelines suggest varying indications for adopted management strategies .

Indications for surgery of five reputed guidelines are compared as follows (table 1). In accordance with them, this patient's initial presentation of acute pancreatitis and dilatation of the MPD warrants surgical intervention.

In large surgical series, MD-IPMNs have been reported to harbour malignancy in more than 60% of cases. BD-IPMNs are indolent in nature, portraying a malignancy rate in the same series of around 25%[1].

Patients with MT-IPMN meet the criteria for both MD-IPMN and BD-IPMN[6], therefore are managed accordingly.

MRI with MRCP is considered the imaging technique of choice, being more accurate than Computed Tomography (CT) in the evaluation of pancreatic cysts; its' sensitivity and specificity in assessing the presence of communication with the MPD are 91-100% and 89%, respectively[3].

However, as with the case of this patient, the misdiagnosis of IPMN as chronic pancreatitis results in serious delays in appropriate management resulting in the patient getting recurrent pancreatitis. Furthermore, curative surgery is only possible if detected early.

The patient was offered surgical intervention only four years after the initial presentation. MRCP was not utilized in the initial sequence of imaging as a diagnostic investigation. The possibility of a malignancy was overlooked without surveillance imaging. The case was not discussed in an MDT meeting with multiple inputs. This signifies, shortcomings in the awareness of the management of IPMNs and the need for collaboration of different specialities when managing a complex patient in the Sri Lankan setting.

Conclusion

With the increasing incidence of IPMNs, it is of timely importance to highlight and develop MRCP as a diagnostic utility in the Sri Lankan surgical setting supported with increased awareness on the management, of the central clinical entity of discussion.

Table 1: Indications for surgery: Variables considered in the initial evaluation of pancreatic cystic neoplasms[5]

		EU	ACG	AGA	IAP	ACR
Symptoms	Jaundice	AI	HR	HR	HR	HR
	Pancreatitis	RI	HR		WF	
Imaging based cyst characteristics	MPD dilation	>10 mm AI 5-10 mm RI	>5 mm HR	HR	>10 mm HR 5-10 mm WF	>10 mm HR 7-10 mm WF
	Associated mass		HR	HR	HR	HR
	Mural nodule	>5mm AI <5mm RI	HR	HR	>5mm HR <5mm WF	WF
	Cyst size	>=4cm RI	>=3 cm HR		>3cm WF	>3cm WF
	Parenchymal atrophy				WF	
	Lymphadenopathy				WF	
Serum based	CA 19-9	RI	HR		WF	
	New onset diabetes	RI				

EU –European based guidelines, ACG –American College of Gastroenterology, AGA –American Gastroenterological association, IAP –International Association of Pancreatology, ACR –American College of Radiology, AI – Absolute indication, RI –Relative indication, HR –High risk, WF –Worrisome features

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