

Sub-Acute Intestinal Obstruction: A Rare Presentation of Mucinous Cyst Adenocarcinoma of Pancreas

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Abstract

Pancreatic adenocarcinoma is one of the most aggressive malignant tumors documented as the fourth leading cause of cancer-related death with overall survival rate <4%. By the time, the clinical symptoms appear and the tumor has attained large size with local invasion leading to delayed diagnosis in the majority of cases. Here, we report a case of 51-year-old female presented with diffuse abdominal pain associated with nausea, vomiting and constipation since 15 days with clinically unremarkable abdominal findings. Abdominal X-ray (standing) s/o multiple air fluid level, and ultrasonography s/o heterogeneous lesion noted in tail of pancreas with central necrotic area. Other investigations brought to the diagnosis of mucinous cyst adenocarcinoma of the pancreas.

Keywords: Abdominal pain, Mucinous cystadenocarcinoma, Tail of pancreas

INTRODUCTION

Anatomical location of pancreas is retroperitoneal in the abdominal cavity and serves both exocrine and endocrine function. It is divided into the head, body and tail. Both the exocrine and the endocrine portion of the pancreatic tissue can turn malignant with majority (95%) developing in the exocrine portion. Only 2% of tumors from the exocrine pancreas are benign, the most common type being adenocarcinomas of the ductal epithelium. The other less common types of exocrine tumors are giant cell carcinoma, adenosquamous carcinoma, cystadenocarcinoma, papillary cystic carcinoma. Tumor of the body and tail of the pancreas constitute one-third of the pancreatic neoplasm. Mucinous cyst neoplasms accounts for approximately 15-30% of cystic neoplasm of the pancreas.¹ The location

of the mucinous cystic neoplasm (MCN) within the gland is entirely confined to the body and tail (97%).¹ The presentation of the symptoms and signs depends on where the tumor is growing and what structure, it is invading. The commonly encountered symptoms although nonspecific include anorexia, loss of appetite, weakness and lethargy. About 70% of the patients presented with a complaint of abdominal pain or discomfort.¹ Here, we present a patient, who reported generalized abdominal pain with nausea, vomiting and constipation with abdominal distension mimicking sub-acute intestinal obstruction.

CASE REPORT

A 51-year-old female presented to the outpatient department of our hospital with diffuse abdominal pain and distension. The pain was described as upper abdominal in location, dull in nature, intermittent for the last 15 days. The pain first presented in the epigastric region and migrated to right hypochondrium and left hypochondrium and finally to the whole of the abdomen. This was associated with nausea, vomiting, constipation and aggravated after by meals. On further enquiry, there was a h/o weight loss

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of approximately 5 kg within 1 month. A history of renal tubular acidosis and scoliosis since 15 years was obtained. The patient is postmenopausal with no h/o major medical or surgical illness in the past. No h/o addictions is present.

Clinically, the patient appeared cachectic and was in distress secondary to abdominal pain with diffuse mild tenderness. Initial laboratory work-up showed hemoglobin 11.3, thin-layer chromatography 13700, platelet 1.7 lacs/cum, erythrocyte sedimentation rate 79, serum sodium 139 meq/L, serum potassium 2.6 meq/L, serum chloride 108 meq/L, liver function test and renal function test within normal limits. Serum amylase and serum lipase were within normal range.

Radiological investigations: Abdominal X-ray (standing) shows multiple air-fluid levels (Figure 1). Ultrasonography findings s/o a 3.6 cm × 3.2 cm heterogeneous lesion noted in tail of pancreas with central necrotic area and focal calcification, mild ascites with cystitis. Hence, the patient was subjected to magnetic resonance cholangiopancreatography (MRCP) that showed a 35 mm × 28 mm sized peripherally enhancing cystic lesion in the pancreatic tail with debris within. No communication with the main pancreatic duct. Possibilities of cystic pancreatic neoplasm were noted (Figure 2). Ascitic fluid tapping was done, and fluid was sent for cytology that revealed adenocarcinomatous cells.

Her chromogranin A was 405.5 ng/mL (reference interval 0.00-100.00) and CA-19.9 was 6795.8 U/mL (reference interval 0-37).

Patient was managed conservatively and later started on gemcitabine and is on regular follow-up since last 2 months.

DISCUSSION

Mucinous cystic adenocarcinoma is a rare entity. They are

spherical, thick-walled, septated or unilocular cysts with a tall columnar mucin-producing epithelium accompanied by a subendothelial ovarian-type stroma that appears as a dense layer of spindle cells with sparse cytoplasm and uniform, elongated nuclei.¹ WHO and Armed Forces Institute of Pathology has defined the presence of this ovarian like stroma as a requirement for the diagnosis of this tumor.¹ Mucinous cystic adenocarcinomas are almost exclusively seen in perimenopausal female patients with the mean age of 48 years with one of the most common sites being the tail of pancreas.²

Patients with tumors arising in body or tail of pancreas usually do not develop jaundice or gastric outlet obstruction.³ Weight loss and abdominal pain are usually the only presenting features. This causes the neoplasm to be diagnosed late and therefore leads to a poorer prognosis.¹ Only 10% of cancers involving body and tail of pancreas are resectable at diagnosis. 5 year survival in patients who have resectable tumors is 8-14%.³ The evaluation of MCN can be done by ultrasound, computed tomography (CT) and/or magnetic resonance imaging (MRI) as they contain large septated cysts with thick irregular walls. MRI may distinguish MCN from other lesions i.e., pseudocyst, intraductal papillary mucinous neoplasms. The carcinoembryonic antigen (CEA) level <800 ng/mL has a specificity of 98% for predicting MCN.⁴ Elevated CEA levels in the fluid (>200 ng/mL) may suggest malignant transformation.⁵

The treatment of choice for early-stage or resectable pancreatic adenocarcinoma is surgery. Depending on

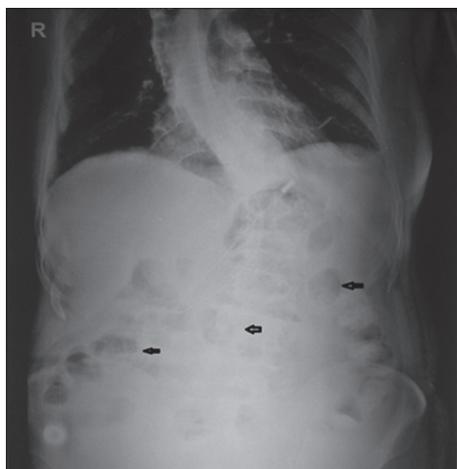


Figure 1: Multiple air-fluid levels



Figure 2: Cystic lesion on tail of pancreas

the location of the tumors, surgical procedures may. For tumors of the head of the pancreas, Whipple's procedure is the preferred procedure. For tumors involving the body of the pancreas, a pancreatectomy is performed and for tumors involving tail of pancreas, a pancreatectomy with splenectomy.

Pancreatic body and tail tumors are less resectable than those of the pancreatic head due to the often earlier presentation of these tumors with obstructive jaundice.⁶ Brennan *et al.* reported that only 10% of patients with tumors of the body and tail of the pancreas are suitable for pancreatic resection.⁶ CT, MRI (MRCP) might helpful in the early diagnosis of the disease but not in the survival. The 5 years survival rate of pancreatic body and tail tumors after surgical resection ranges from 0% to 25%, and the median survival time is 10-15.9 months.^{7,8} Most patients will require chemotherapy and radiation therapy after the surgery.

Gemcitabine is the first-line therapy for patients with metastatic pancreatic adenocarcinoma with a median overall survival of 5.65 months, progression-free survival of 9 weeks and clinical benefits response of 23.8%.⁹

CONCLUSION

The majority of the carcinoma tail of pancreas is mucinous cyst adenocarcinoma and presents with unusual clinical features like in our case where patient presented with sub-acute intestinal obstruction. When patient presents to us,

it is already in the late stage of the disease, so the death ratio is too high compared to other tumors. Currently, available approach for carcinoma tail of pancreas is surgical resection and palliative care. Hence, we conclude that a high index of suspicion is required to diagnose such condition at the initial stage to have a positive impact in terms of management and overall survival.

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