A RARE CASE OF SARCOMATOID CARCINOMA OF THE PANCREAS ASSOCIATED WITH PANCREATOLITHIASIS

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Abstract

Pancreatolithiasis is a risk factor for developing pancreatic cancer. We report here a rare case of sarcomatoid carcinoma of the pancreas in a 55-year old diabetic male associated with pancreatolithiasis. CT scan of abdomen revealed a large operable mass occupying the distal body and tail of the pancreas. Per-operative survey revealed a small metastatic nodule in the surface of hepatic segment IVa. Histopathology of the distal pancreatic lesion revealed sarcomatoid carcinoma. Hepatic nodule was a metastatic adenocarcinoma. Distal pancreatectomy and splenectomy was done en-mass, along with non-anatomical resection of the hepatic metastatic nodule. Combined with six cycles of chemotherapy, the patient survived a total of another fourteen months.

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Introduction

Our study on 120 cases of pancreatolithiasis has shown that risk of associated malignancy with pancreatolithiasis is >6%.[1] Therefore, a mass lesion of pancreas associated with long standing pancreatolithiasis should raise the high index of suspicion for a malignant lesion. If the lesion is located distally in the body and tail, the situation is more complex, because distal pancreatic malignancy is an aggressive disease and is associated with low resectibility and poor prognosis.[2] In the pancreas, about 90% of mass forming solid lesions are adenocarcinoma which has a poor prognosis with a 5-year survival rate of only 5%.[3] On the other hand, the anaplastic cancers of the pancreas (ACP) are rare aggressive tumors of duct cell origin and account for 2-7% of all pancreatic cancers.[4] The median survival rate of ACP is less than one year.[5] Sarcomatoid carcinoma is a subtype of ACP. We are reporting a rare case of sarcomatoid carcinoma which presented with a distal pancreatic solid mass in the background of diabetes mellitus and pancreatolithiasis.

Case report

A 55 year male college teacher, known diabetic for 25 years and on insulin for 6 years, was diagnosed as a case of pancreatolithiasis for last two years. Pancreatolithiasis was diagnosed by plain X-ray of the abdomen which showed multiple large stones in the pancreas (Fig:1a). He presented to Hepato-Biliary out-patient department with occasional colicky pain in the left hypochondrium, anorexia and weight loss for last one year. His built and nutrition was average, but he was anxious for pain because it was hampering his regular activities. He was not anemic or icteric, and vital signs and symptoms were stable. Abdominal examination revealed mild tenderness at left hypochondrium with no organomegally or ascites. Haematological and biochemical parameters including liver function tests and pancreatic enzymes were within normal limits. Ultrasonography of the abdomen revealed pancreatolithiasis, dilated major pancreatic duct (main pancreatic duct diameter 17mm) and a mass lesion occupying the distal body and tail of the pancreas. Computerized tomography (CT) scan of abdomen

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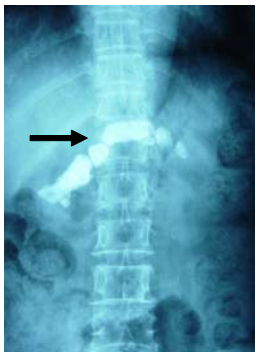

Fig. 1a: *Plain X-ray of abdomen showing pancreatolithiasis*

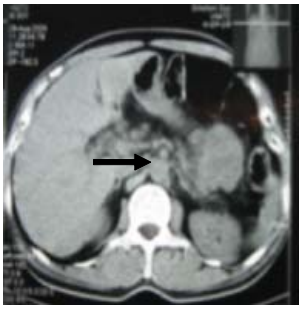

Fig. 1b: *CT scan of abdomen showing distal pancreatic mass*

showed a fairly large mass (6.5 X 5.0 X 5.0 cm) occupying the distal pancreas (Fig1b). CA 19-9 level was 183 u/L (n= <37 u/L). CT guided fine needle aspiration cytology (FNAC) was done and it was positive for malignant cells.

Our impression was pancreatolithiasis with a malignant lesion in the distal pancreas. Plan of management was to perform distal pancreatectomy and splenectomy en-mass along with pancreatolithotomy and Roux-en-Y pancreatojejunostomy. Patient was prepared accordingly and was vaccinated against *Streptococcus pneumoniae*, *Haemophilus influenzae* and *Neisseria meningitides* as per schedule.

Laparotomy was done by midline incision with combined thoracic epidural and general aneaesthesia.

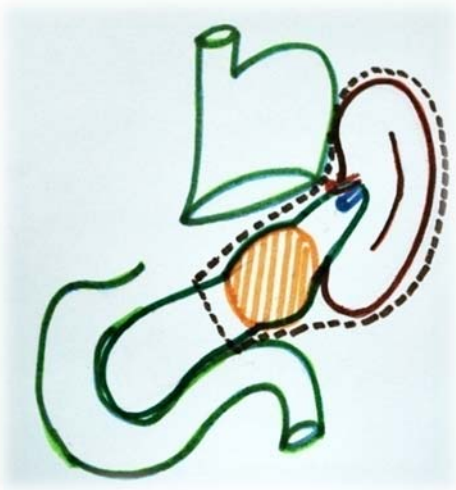

Fig. 2a: *Line of en-mass resection*

There was no ascites and peritoneal cavity appeared to be healthy. Large stones could be felt in the pancreatic duct and there was a tumor in the distal body and tail of the pancreas, free from surrounding structures. There was no enlarged regional lymph node, but there was a metastatic nodule (1.0 X 1.25 cm) found in the hepatic segment IVa. Operative ultrasound revealed no other metastasis in any other part of the liver. Repeat survey in the abdomen revealed no other detectable metastatic lesion in any part of the peritoneal cavity or any abdominal organ. The spleen, tumor and the distal pancreas were mobilized and removed en-mass as shown in Fig 2a and 2b. Hepatic nodule was removed by non-anatomical hepatic resection. (Fig.2c)

Histopathology of the specimen revealed sarcomatoid carcinoma characterized by plenty of spindle cells along with glandular structures and areas of clear-cut glandular differentiation in the distal pancreatic lesion (Fig.2d). Resection margin was free of tumor invasion. Hepatic nodule was a metastatic adenocarcinoma with hepatic margin free of tumor invasion.

Postoperatively, the patient developed deep vein thrombosis (DVT) on the 12th post operative day (POD) and was managed conservatively. Good glycaemic control was maintained. Patient was discharged on 18th POD with advice for oncological consultation.

First follow up was after one month and subsequently the patient was followed up for next one year. During this period he received six cycles of chemotherapy and was under the care of an oncologist. After one year, there was recurrence of the tumor in the operative bed. With consultation of the oncologist, chemotherapy

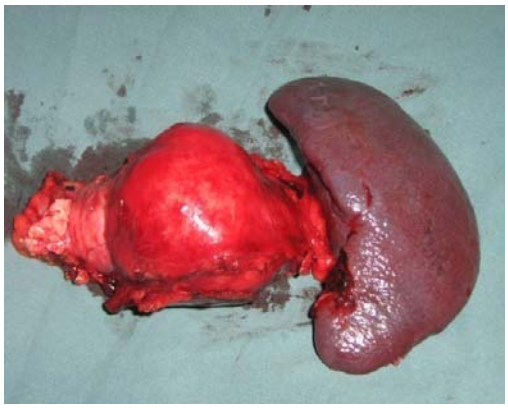

Fig. 2b: *Resected en-mass specimen of distal pancreas and spleen*

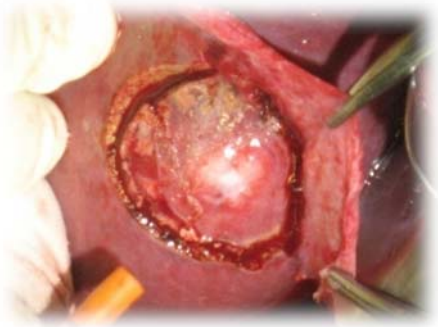

Fig. 2c: *Removal of hepatic nodule*

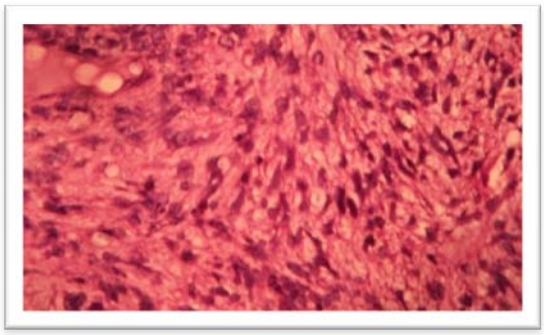

Fig. 2d: *Histopathology of distal pancreatic lesion showing sarcomatoid carcinoma characterized by plenty of spindle cells along with glandular structures and areas of clear-cut glandular differentiation.*

was restarted, but the patient died of disseminated disease after two months.

Discussion

Anaplastic cancers of the pancreas (ACP) are rare aggressive tumors of duct cell origin and account for 2-7% of all pancreatic cancers.[4] Sarcomatoid carcinoma is a subtype of ACP – other subtypes are pleomorphic and spindle cell carcinomas. It was first described by Sommer and Meissner in 1954.[6] ACP has been described in different names, including undifferentiated carcinoma, pleomorphic carcinoma, pleomorphic giant cell carcinoma, anaplastic carcinoma, undifferentiated carcinoma with osteoclast-like giant cells (OCGCs) and mixed osteoclast and pleomorphic-type giant cell tumor.[7-10]

The overall survival for ACP is poor when compared with pancreatic duct adenocarcinomas (PDA) but several studies suggest that operative resection provide little benefit.[11,12] A Mayo Clinic Institutional Review Board approved cohort study using the Surveillance, Epidemiology and End Results (SEER) cancer registry database has shown that males are more affected by ACP than females, the size of ACP is larger at presentation than other PDA (median tumor size 6.0 cm vs. 3.5 cm) and PDA are located more in the head region of the pancreas whereas ACP are located more distally in the body and tail region.[13]

Sarcomatoid carcinoma is rare and an aggressive form of cancer and the resultant tumors are frequently symptomatic, locally advanced and have high rates of recurrence. Non-surgical treatments are usually recommended for patients with clinically advanced lesions.[12] In our case the hepatic metastasis was undiagnosed in the imaging studies. The tumor was well capsulated and easily separable from surrounding structures. There was a hepatic metastasis (1.0 X 1.25 cm in size) in the hepatic segment IVa. Peroperative ultrasonography revealed no other detectable metastatic lesion in any other part of the liver. In this situation, radical resection of the primary lesion and non-anatomical resection of the solitary hepatic metastasis was done which might produce a durable palliation.

However, local recurrence of the disease is very high in ACP and Strobel and colleagues (2011)[5] has shown in a study that after curative resection of ACP, median survival is only 7.1 months. Yamaguchi *et al.* (1998) has shown a zero one-year survival for patients with ACP.[12] Therefore, it is suggested that pre-diagnosed case of ACP should not undergo surgery. In our case, the decision of operation was influenced by FNAC report, but subtype categorization of pancreatic carcinoma was difficult with FNAC because of the small sample examined microscopically.[14] Recently, Clark *et al.* (2012)[11] has mentioned in his study that patients of ACP who underwent pancreatic resection had an overall survival comparable to patients with PDA. Therefore, they recommended that patients with ACP should be offered pancreatic resection when technically feasible. So the operation was justified in our case even with the hepatic metastasis and the patient got a total of fourteen months survival benefit.

Pancreatolithiasis is a risk factor for developing pancreatic cancers. Solid pancreatic lesion with associated pancreatolithisis should be regarded as malignant. Meticulous preoperative evaluation, planning, adequate surgery when technically feasible,

along with proper adjunct oncological care, may offer a durable palliation.

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