International Surgery Journal
Anandi A et al. Int Surg J. 2023 Jun;10(6):1106-1109
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20231748

Case Report

Carcinosarcoma of pancreas: an unusual pancreatic tumour: case report

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Received: 07 April 2023
Revised: 09 May 2023
Accepted: 10 May 2023

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ABSTRACT

Carcinosarcoma of pancreas is a rare tumour with limited clinical and pathological data reported in literature. Although carcinosarcoma predominantly found in uterus, it has been identified in breast, liver, prostate, kidney, salivary glands and pancreas. Carcinosarcoma is a biphasic tumour with epithelial and mesenchymal components. About less than 40 cases had been reported in literature. The treatment of choice in most reported cases is surgery. Given such rarity, we report a case of pancreatic carcinosarcoma diagnosed in our institution, review tumour clinicopathological characteristics, and describe our surgical and medical management strategy.

Keywords: Carcinosarcoma, Epithelial, Spindle cell, Pancreas

INTRODUCTION

Carcinosarcoma of the pancreas is a rare malignancy with high mortality. Diagnosis is based on pathologic demonstration of adjacent malignant epithelial and mesenchymal tissue. Due to inherent limitations of biopsy sampling, tumor heterogenesity is rarely recognized until definitive surgical resection. Although carcinosarcoma predominantly found in uterus, it has been identified in breast, liver, prostate, kidney, salivary glands and pancreas.[1] The most common pancreatic tumour is ductal adenocarcinoma. Undifferentiated carcinoma is one of its subtypes and it has three distinct patterns: anaplastic undifferentiated carcinoma, sarcomatoid undifferentiated carcinoma and Carcinosarcoma.[2] Most of the carcinoma components are well-differentiated ductal adenocarcinoma with small areas of moderately to poorly differentiated ductal adenocarcinoma. The sarcomatous component is a high-grade highly cellular spindle cell tumor with frequent mitosis and apoptosis. Here, we present a case of pancreatic carcinosarcoma, including the clinical and histopathological features and their management strategies.

CASE REPORT

A 53 year old male with history of hypertension presented to our OPD with complaints of upper abdominal pain for past 3 months on and off, dull aching which was radiating to back for past 1 week. Patient had no history of abdominal distension, vomiting, anorexia, jaundice. He had no history of loss of weight and appetite. He had no significant past, personal and family history. His bowel and bladder habits were normal. On general examination ECOG score was 1, with no pallor, icterus. Supraclavicular fossa was empty. Per abdominal examination showed ill-defined mass of size 3×2 cm over epigastrium which does not moves with respiration. Tenderness present. The mass was firm in consistency and dull to percuss. No evidence of free fluid present over abdomen. Per rectal examination showed no deposits. Patient was admitted in our unit and was further evaluated. Routine blood investigations showed normal parameters (including LFT) except raised total count

(26,600 cells per cu.mm). Triple phase contrast enhanced CT abdomen and pelvis showed well defined lobulated hypodense lesion with exophytic component noted in neck and body of pancreas with areas of necrosis with displacement of splenic vein and artery with no surrounding fat stranding. Upper GI scope with Side view scope showed prominent ampulla with ulcerations from which biopsy taken. Biopsy was not suggestive of malignancy. Tumour markers (CA 19-9 and CEA) were within normal limits. MRCP showed heterointense lesion of size 5×6×6 cm lesion in body of pancreas with abutment of splenic vein posteriorly and peripancreatic fat planes preserved. Staging workup was negative for metastasis.

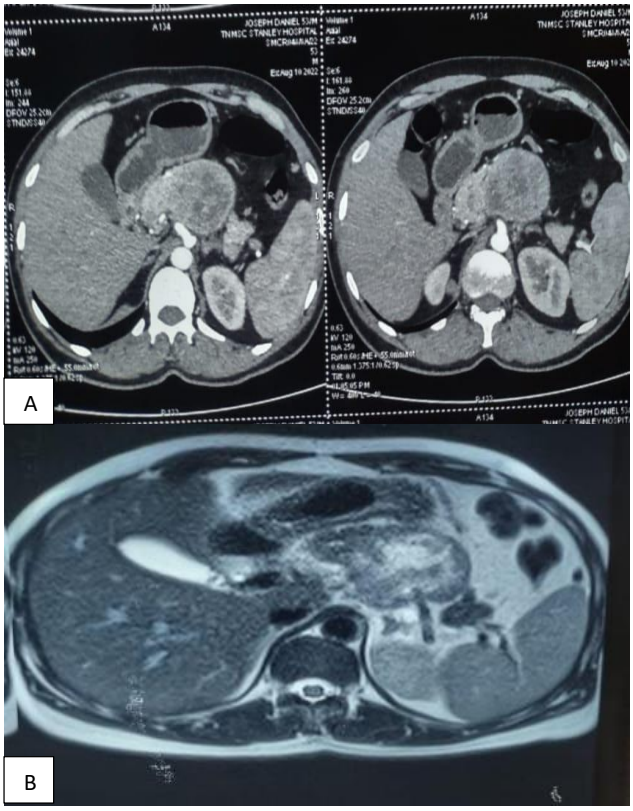

Figure 1 (A and B): CECT abdomen of hypodense lesion in neck and body of pancreas with areas of necrosis. MRCP showing heterointense lesion in body of pancreas with MPD displaced anteriorly.

After discussing the case with surgical oncologist and surgical gastroenterologist, patient was planned for trial resection. After complete preoperative evaluation, patient underwent radical antegrade modular pancreaticosplenectomy (RAMPS) on 6/9/2022. Intraoperatively an irregular tumour of size 10×12 cm found in body of pancreas extending superiorly up to common hepatic artery, inferiorly up to left gerota fascia and distally to tail of pancreas. Tumour was found to adherent to common hepatic artery. SMA, SMV and portal vein was free of tumour. In view of intraoperative difficulty, surgical gastroenterologist team was called and procedure went uneventful. Patient was extubated on pod 1, postoperative period were uneventful. CBG monitoring was done periodically and there were no fluctuations. DT removed on pod 10 and sutures were removed on pod 14. Patient was given post splenectomy vaccine prophylaxis.

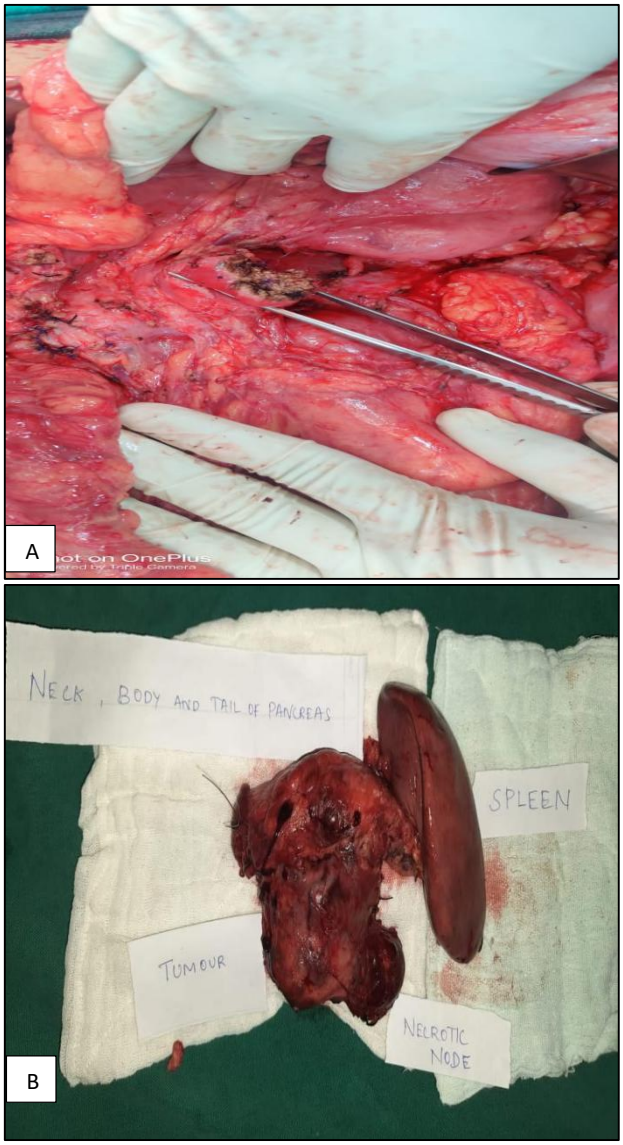

Figure 2 (A and B): Intraoperative picture with remnant pancreatic head after pancreaticosplenectomy; Postoperative specimen showing resected tumour along with necrotic node.

To our surprise, histopathological report showed features suggestive of carcinosarcoma of pancreas with resected margins were free of tumour. The tumour showed two components composed of epithelial component and spindle cell component that were intermingled together. The majority of epithelial component was round to polygonal cells, well differentiated with low mitotic rate with tumour giant cells. The spindle cell component was arranged in fascicles and sheets with extensive areas of necrosis with pale eosinophilic cytoplasm and pleomorphic nuclei. Lymphovascular invasion was present. Out of six regional lymph nodes examined three

were found to be positive and pathologic stage classification was PT3N1. To confirm the immunohistochemistry (IHC) was done in which Pan CK marker was positive in tumour cells.

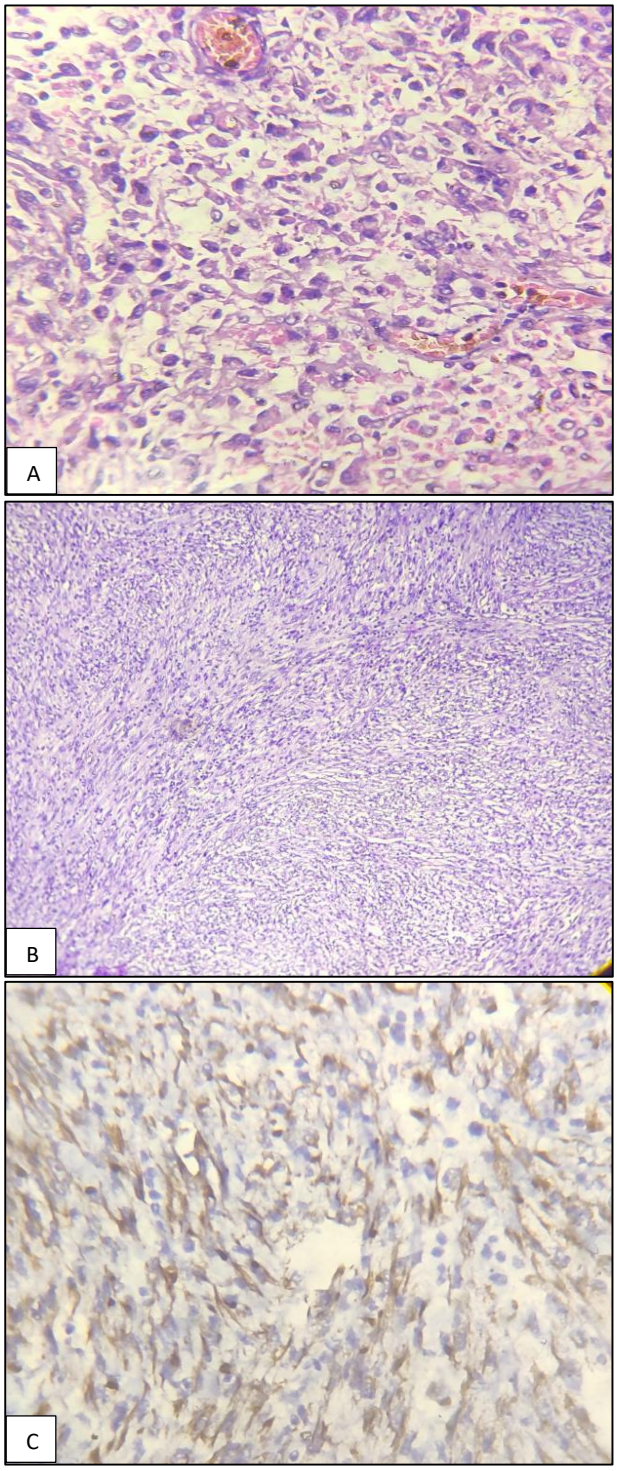

Figure 3 (A-C): H and E showing carcinomatous component with epitheloid cells; H and E showing sarcomatous component with high grade spindle cells; IHC showing pan cytokeratin markers positive on tumour cells.

Medical oncologist expert opinion was obtained and patient was started on adjuvant chemotherapy with paclitaxel 21 days post-surgery. Patient was discharged and was on our regular follow up.

DISCUSSION

Carcinosarcoma of pancreas is an extremely rare and complicated malignant neoplasm with the characteristics of high invasiveness and rapid progression, early metastasis and poor prognosis. Carcinosarcoma composed of carcinomatous and sarcomatous cellular components. Each component should account at least for 30% of the tumour. With scanty number of cases, it was seen that carcinosarcoma of pancreas had a gender predilection towards female. The mean age of the patients was 65 years with a range of ages from 46 to 90. As per WHO classification of tumours it comes under the category of undifferentiated carcinoma of pancreas along with sarcomatoid and anaplastic carcinoma. It is a single tumor consisting of two distinct malignant tissues with both epithelial and mesenchymal morphology. The epithelial component can be adenocarcinoma, squamous cell carcinoma, or basal cell carcinoma, among others. The mesenchymal component can be spindle cell sarcoma, rhabdosarcoma, osteosarcoma, chondrosarcoma, or undifferentiated sarcoma.[3]

Most malignant tumors of pancreas are adenocarcinomas (85%) and show a male predilection. However, carcinosarcomas showed a greater incidence in females (10:9). They are usually seen in elderly patients with mean age of 65 years (46 to 90). The usual presenting symptom were upper abdominal pain and jaundice.[4]

Carcinosarcoma is best characterised by IHC staining. Three histogenic theories have been proposed to explain the mechanism of their development-collision, combination and conversion. In combination theory both cellular components develop from a single progenitor cell. In the collision theory, carcinomatous and sarcomatous components arise independently. In the conversion theory, the sarcomatous component evolves from its epithelial counterpart.[5]

The IHC diagnosis of carcinosarcoma is established by visualising positive reactivity of the carcinomatous elements to cytokeratin and of sarcomatous elements to vimentin or Desmin. In the current study, the tumour tested positive for pan cytokeratin even in sarcomatous components.[6] Histologically the tumour demonstrated well differentiated carcinomatous component with high grade spindle cell sarcoma.

Almost all the cases have been primarily treated by surgical therapy. To date, systemic treatment for carcinosarcoma of the pancreas has been extrapolated from treatment guidelines for pancreatic ductal adenocarcinoma (PDAC).[7] Scanty information is available about chemotherapy and radiotherapy in

management of this rare tumour. Gemcitabine delays disease progression when given adjuvant to a patient with pancreatic cancer, who has undergone curative-intent surgery.[8] Surgery with lymphadenectomy, adjuvant therapy, and neoadjuvant therapy are thought to help improve survival outcomes. Modern treatment approaches for conventional pancreatic ductal adenocarcinoma could be applied to pancreatic carcinosarcoma.[9]

Other potential second-line treatments include capecitabine or fluorouracil (5-FU), oxaliplatin with CAP or 5-FU, or gemcitabine with erlotinib. When comparing the overall survival, the use of second-line therapies shows a statistically significant increase compared with best supportive care.[10] A locally advanced or metastatic pancreatic cancer warrants the use of adjunct systemic therapy.

CONCLUSION

Patients with pancreatic carcinosarcoma manifests nonspecific preoperative signs and symptoms and definitive diagnosis is based on post operative pathological findings. Despite surgery and adjuvant chemotherapy, recurrence rates are high and prognosis is poor. Our goal in presenting this case is to bolster the literature regarding this rare neoplasm. Without treatment guidelines available, we encourage reporting of long-term survival in these patients.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Anandi A, Praveen PA, Rani SR. Carcinosarcoma of pancreas: an unusual pancreatic tumour: case report. Int Surg J 2023;10:1106-9.