

## CASE REPORT

# Carcinosarcoma of the Pancreas: How a Common Blood Disorder Can Hide an Extremely Rare Tumour

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### ABSTRACT

**Context** Sarcomas represent a relatively rare malignancy. Primary sarcomas of the pancreas represent an extremely rare pathology. **Case report** We report a case of primary pancreatic carcinoma that presented with anaemia. The patient underwent a Kausch-Whipple operation, and, 16 months after the operation, the patient is disease free. **Conclusion** This unique case describes an extremely rare gastro-intestinal tumour that was found during the patient's anaemia assessment.

### INTRODUCTION

Primary sarcomas of the pancreas represent an extremely rare pathology that accounts for less than 0.1% of pancreatic malignancies and only 11 cases have been described in the English literature. The histogenesis of carcinosarcomas is not clearly understood. It is an aggressive tumour associated with a poor prognosis. The treatment of choice is surgical resection followed by adjuvant chemotherapy based on gemcitabine and cisplatin.

### CASE REPORT

A seventy-year-old man was admitted to our department for an elective left inguinal hernia repair. During the preoperative assessment, the patient was found with low haematocrit.

The patient had been on simvastatin the last five years, the rest of his medical history was negative. There was no relevant family or social history. In his physical examination, the patient was pale with no evidence of jaundice. The patient lost 10 kg during the past year without dieting and was suffering from symptoms of depression during the same period. An abdominal examination revealed an indirect left inguinal hernia. A rectal examination was normal. Investigations confirmed low haemoglobin 8.3

g/dL and haematocrit 25%. The rest of the biochemical examinations were normal. In the tumour markers, only the CA (carbohydrate antigen) 19-9 was elevated with a value of 169.67 U/mL. An abdominal ultrasound and colonoscopy showed non-pathological results. The gastroscopy revealed a large tumour of the ampulla of Vater (Figure 1), while the histology was positive for malignant cells with sarcomatous characters and frequent mitotic figures (>40/10 high power fields). The computed tomography scan demonstrated a large mass of the ampulla of Vater, causing obstruction and dilatation of the common bile duct, the intrahepatic and the pancreatic duct (Figure 2). The magnetic resonance revealed a large mass of the uncinata process of the pancreas of 4.7x4.5 cm and the dilatation of the intrahepatic bile ducts, common bile duct and pancreatic duct with no invasion of the superior mesenteric vessels or the portal vein and no metastasis.



**Figure 1.** Gastro-duodenoscopy demonstrating a tumor at the ampulla of Vater.

**Received** January 07th, 2015 – **Accepted** March 28th, 2015

**Keywords** Anemia; Carcinosarcoma; Pancreas

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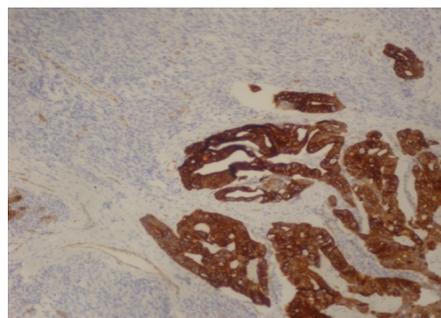
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**Figure 2.** CT of the abdomen which shows a mass at the ampulla of Vater with dilated common bile duct, pancreatic duct and intrahepatic ducts.



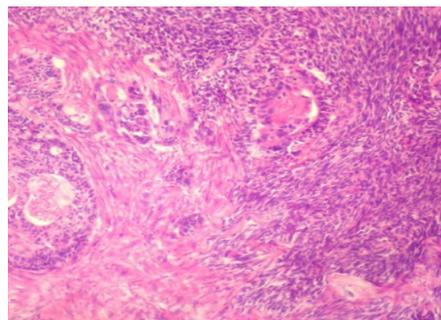
**Figure 3.** Immunohistochemical staining of the epithelial component showing positivity to CK 7 (cytokeratin) (magnification x 45).

The patient underwent a Kausch-Whipple operation. Upon gross examination, the head of the pancreas revealed an exophytically-growing tumour localised in the duodenum orally to the ampulla of Vater that measured 11x9x6 cm. The resection margins and lymph nodes were tumour free, from the 14 that were resected.

Microscopically, the tumour yielded two components. The first component was a relatively well-differentiated adenocarcinoma, while the second component showed poorly-differentiated solid tissue of mesenchymal origin (Figure 3). Immunohistochemically, the epithelial component was strongly reactive for antibodies to CK7 and CD34, while the sarcomatoid component was negative for all CK antibodies but strongly reactive for the antibody to vimentin (Figure 4). The postoperative period was uneventful. The patient was discharged the ninth postoperative day. A venous port implanted six weeks after the operation and he received eight cycles of gemcitabine. Despite the poor prognosis, the patient is still alive and without evidence of the disease 16 months after the operation.

## DISCUSSION

Carcinosarcomas are biphasic neoplasms consisting of two components: the malignant epithelial and the mesenchymal, both demonstrating distinct immunohistochemical and molecular features pertaining to their different lines of differentiation [1, 2]. The histogenesis of carcinosarcomas is not clearly understood. Four main hypotheses have been proposed: the collision, the combination, the conversion and the composition theories [3, 4]. ‘Conversion’ indicates that a section of the carcinoma transforms into a sarcomatous element, ‘combination’ describes the development of



**Figure 4.** Histological examination revealed duct-like and spindle-shaped atypical cells (left) and carcinosarcoma appearance (right) after hematoxylin and eosin stain (magnification x45).

a tumor from a single stem cell that differentiates into epithelial and mesenchymal tissues ‘collision’ refers to the invasion of carcinomatous and sarcomatous elements into each other, and finally the ‘composition’ theory when the sarcomatous component is an exuberant, non-neoplastic overgrowth of connective tissue in reaction to the presence of neoplastic carcinoma.

Regarding the pancreas, only a few cases of carcinosarcoma have been reported in the literature (Table 1) [5-14]. According to Feather *et al.* [15], sarcomas of the pancreas occur frequently in younger patients with the pancreatic head being most commonly involved, followed by the tail and the body of the organ. Because of its extreme rarity, the histogenesis of pancreatic carcinosarcoma remains unclear. The identification of identical DNA sequences in multiple genes supports the hypothesis that carcinosarcomas derive from a single stem cell. Several studies have suggested a monoclonal origin for

**Table 1.** Review of the literature of patients with carcinosarcoma of the pancreas.

Author	Age	Histology of non-epithelial component	Mucinous cystic component	Survival (months)
Millis <i>et al.</i>	50	Leiomyosarcoma	No	3
Watanabe <i>et al.</i>	76	Osteoclastic giant cells	No	3
Wenig <i>et al.</i>	48	Spindle shaped stroma	Yes	12
Wenig <i>et al.</i>	65	Spindle shaped stroma	Yes	9
Wenig <i>et al.</i>	67	Spindle shaped stroma	Yes	15
Darvishian <i>et al.</i>	74	Malignant fibrous histiocytoma	No	-
Yamazaki <i>et al.</i>	90	Spindle shaped stroma	No	-
Barkatullah <i>et al.</i>	67	Spindle shaped stroma	No	8
Bloomston <i>et al.</i>	67	Spindle shaped stroma	Yes	4
Gelos <i>et al.</i>	61	Spindle shaped stroma	Yes	9
Ambe <i>et al.</i>	52	-	-	-
Our case	70	Spindle shaped stroma	Yes	16

carcinosarcomas using diverse immunohistochemical and molecular analyses [2].

An immunohistochemical diagnosis is established by the reactivity of the carcinomatous and sarcomatous elements to CK and vimentin, respectively. Carcinosarcomas must be differentiated from sarcomatoid carcinomas, which immunohistochemically are positive only to CK [16].

Carcinosarcoma of the pancreas is an aggressive tumour that carries a poor prognosis. The treatment of choice is surgical resection followed by gemcitabine as an adjuvant treatment [17]; a combination with cisplatin has been associated with a high resection rate and improved survival in the neoadjuvant setting in a preliminary study [18].

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### Conflicting Interest

The authors had no conflicts of interest

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