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(CASE REPORT)



# A rare case of sarcomatoid carcinoma of the pancreas

A. Snoussi\*, F. Lamri Zeggar, C. Sabat and C. Locher

Department of Hepato-gastro-enterology Meaux hospital Paris, France.

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## Abstract

Sarcomatoid carcinoma of the pancreas is a rare malignancy characterised by a predominant sarcomatoid component associated with a well-differentiated adenocarcinomatous component. We report the case of an 84-year-old patient with a history of cardiac arrhythmia who was admitted to hospital with jaundice of one month's duration. Ultrasound and abdominal computed tomography revealed a solid and locally advanced pancreatic head mass. The patient underwent endoscopic ultrasound with biopsy of the mass and placement of a biliary stent. Pathological examination of the biopsy revealed a sarcomatoid carcinoma of the pancreas. In view of the patient's bedridden condition and comorbidities, it was decided at a multidisciplinary consultation meeting to withhold treatment.

The patient died 1 month later.

Keywords: Sarcomatoid carcinoma; Echo-endoscopy; Pancreas; Prognosis; Epithelial differentiation

## 1. Introduction

Sarcomatoid carcinoma of the pancreas is a rare biphasic and aggressive tumor with a poor prognosis, characterised by the coexistence of a predominantly sarcomatoid component associated with a well-differentiated epithelial component.

## 2. Patient and observation

We report the rare case of an 84-year-old hypertensive man with a history of cardiac arrhythmia who presented with jaundice of 1 month's duration without fever and associated with an altered general condition. The clinical examination was unremarkable. Abdominal ultrasound showed the presence of a fairly well circumscribed heterogeneous mass measuring  $4 \times 2.5 \text{ cm}$  at the level of the head of the pancreas and moderate dilatation of the intrahepatic bile ducts and the common bile duct. Abdominal CT showed a hypodense lesional process of the head of the pancreas measuring 4 cm by 3 cm with surrounding adenopathy and most likely vascular extension (Fig.1).

<sup>\*</sup> Corresponding author: A. Snoussi

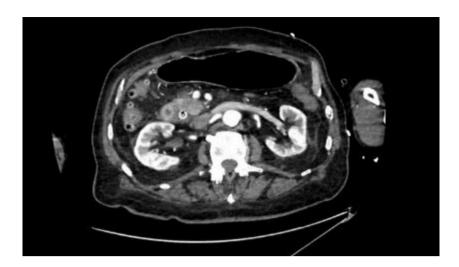
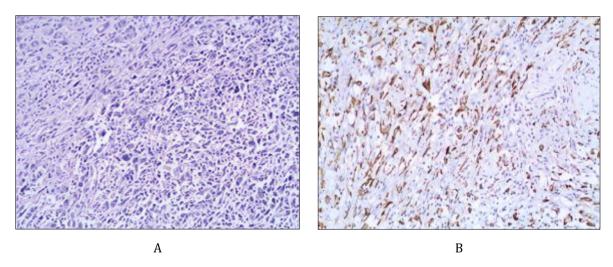


Figure 1 Abdominal CT scan showing pancreatic head tumor

Biologically, there was a normocytic normochromic anemia of 10.2 g/dl, cholestasis with a total bilirubin of 195 mmol/l, CRP was 40 mg/l and tumor markers (ACE and CA 19-9) were negative. Locoregional and distant extension studies showed no liver lesions or secondary lung lesions.

The patient underwent endoscopic ultrasound with biopsy of the pancreatic mass and placement of a biliary stent. Histological analysis revealed a biphasic proliferation. The first component was sarcomatoid, consisting of a diffuse layer of spindle-shaped and oval cells with elongated nuclei with vesicular chromatin showing multiple atypical mitotic patterns. The cytoplasm was sparsely eosinophilic with indistinct cytoplasmic borders. The second component was a well-differentiated adenocarcinoma (Figure 2). Immunohistochemistry showed positive and intense labelling of both spindle cells and well-differentiated foci with epithelial markers (cytokeratin AE1 and AE3, cytokeratin 19, EMA). The tumor cells were negative for mesenchymal markers (desmin, CD 117 and CD34). The diagnosis was sarcomatoid carcinoma of the pancreas.



**Figure 2** Tumor proliferation with a sarcomatoid component (A) and positivity of the sarcomatoid component for cytokeratin AE1/AE3 (B)

The case was discussed in a multidisciplinary consultation and the patient opted for therapeutic abstinence.

The patient died 1 month later.

#### 3. Discussion

Sarcomatoid carcinoma of the pancreas is a rare, aggressive tumor with a poor prognosis and an estimated median survival of 6 months. It affects men and women equally and often presents in the sixth decade of life [1]. The clinical

presentation depends on the location and size of the tumor and is no different from that of ductal carcinoma of the pancreas [2]. In our case, transfixing epigastralgia dominated the clinical picture. However, tumor markers (ACE and CA19-9) are negative in over 80% of cases, as in our patient [3].

Radiologically, the tumor is hypodense on CT and hypointense on MRI due to the resulting desmoplastic stroma, which is poorly perfused compared to normal pancreatic parenchyma. After contrast injection, the lesion is hypodense with peripheral contrast enhancement in the portal venous phase. They may have a cystic appearance, simulating a cystic lesion of the pancreas, which is reflected histologically by central necrosis with viable peripheral tumor tissue [3].

Pathologically, sarcomatoid carcinoma has a solid cross-sectional appearance with hemorrhagic and necrotic changes. Histological examination reveals a proliferation of atypical spindle cells with a hyperchromatic nucleus, sometimes nucleolated, with a few atypical mitoses. After careful sampling, this proliferation is associated with foci of well-differentiated adenocarcinoma. These cells do not express mesenchymal markers (desmin, vimentin, CD117) and consistently express epithelial markers (cytokeratin AE1 and AE3, cytokeratin 19, EMA) [4]. In our case, this tumor proliferation was indeed of epithelial origin, as it was positive for cytokeratins and EMA. The constant search for the K-Ras mutation suggests that this tumor component is derived from pancreatic ductal cells [5].

Treatment of sarcomatoid carcinoma of the pancreas follows the same goals and procedures as for ductal adenocarcinoma of the pancreas [6].

Sarcomatoid carcinoma has a poor prognosis, with median survival estimated at 6 months [6].

#### 4. Conclusion

Sarcomatoid carcinoma of the pancreas is a rare tumor. Its management is identical to that of ductal adenocarcinoma, with which it shares a relatively poor prognosis.

#### Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

The authors appropriate statement of ethical approval.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study

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