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CASE REPORT

# Sarcomatoid carcinoma of the pancreas — a rare tumor with an uncommon presentation and course: A case report and review of literature

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**Author contributions:** Toledo PF reviewed the literature and drafted the manuscript; Berger Z was the patient's gastroenterologist who maintained medical follow-up and was responsible for critical revision of the article for important intellectual content; Carreño L performed the analyses and interpretation of the anatomopathological findings of the described tumor; Cárdenas G analyzed and interpreted the imaging findings; Castillo J and Orellana O were the patient's digestive surgeons; all authors issued final approval for the version to be submitted.

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## Abstract

### *BACKGROUND*

Sarcomatoid carcinoma of the pancreas (SCP) is a rare type of pancreatic neoplasm, and only a few cases have been described in the literature. Histologically, it is composed mostly of atypical spindle cells with apparent sarcomatous features.

### *CASE SUMMARY*

This is a report of a 61-year-old Chilean woman who underwent medical investigation for acute abdominal pain. Computed tomography identified a solid tumor in the tail of the pancreas with features suspicious of malignancy. *En-bloc* distal pancreatectomy and splenectomy were performed to excise the tumor. Histopathology and immunohistochemistry were confirmatory of sarcomatoid carcinoma with lymphovascular invasion. After surgery, the patient did not receive chemotherapy. Previous studies indicate a poor prognosis for this type of malignancy. However, our patient has survived for 35 mo with no recurrence to date.

The authors have read the CARE Checklist (2016), and the manuscript was prepared and revised accordingly.



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*CONCLUSION*
The case presented herein is a patient with an SCP with a rare presentation and long-term survival after surgery despite not receiving adjuvant chemotherapy.

**Key Words:** Pancreatic neoplasms; Sarcomatoid carcinoma; Pancreatic ductal carcinoma; Survival; Abdominal pain; Case report



**Core Tip:** Sarcomatoid carcinoma of the pancreas (SCP) is an extremely rare and aggressive histologic subtype of undifferentiated pancreatic carcinoma. The prognosis of this neoplasm is similar to or even worse than that of typical pancreatic ductal adenocarcinoma (PDAC). However, the clinical course and surgical outcomes of SCP remain poorly characterized owing to its rarity. Because there is no standard regimen for treating SCP, patients with this disease are administered the same regimens as those with more common PDACs. In the present study, we report a case of SCP; although some patients have a rapid recurrence and early death, long-term survival may be possible.

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## INTRODUCTION

Pancreatic cancer is considered a disease with uniformly poor outcomes[1]. The worldwide 5-year survival rate for pancreatic cancer patients is approximately 6%[2]. Pancreatic ductal adenocarcinoma (PDAC) is by far the most common solid pancreatic tumor, which represents 85 to 90% of all pancreatic neoplasms; thus, most attributes of pancreatic cancer are related to this tumor[3]. However, several morphological variants of PDAC are recognized in the latest (2019) World Health Organization (WHO) classification of pancreatic tumors based on distinctive histologic features[2,4]. Sarcomatoid carcinoma of the pancreas (SCP) is among these variants. SCP is an extremely uncommon tumor that accounts for 0.1% to 5.7% of all pancreatic malignancies[5]. It is an undifferentiated carcinoma that shares similar molecular pathogenesis with PDAC and therefore a similarly poor prognosis[2]. Despite aggressive surgical management, the median postoperative survival has consistently been reported as less than 1 year[6]. Most examples of SCP are found in the literature only as case reports.

We report an exceptional case of SCP detected in a patient who underwent consultation in our emergency room with acute abdominal pain. The patient has survived for a long time to date without disease recurrence despite not receiving chemotherapy. We, therefore, discuss this case and review the relevant literature.

## CASE PRESENTATION

### *Chief complaints*

A 61-year-old female was admitted to our hospital suffering from 48 h of acute abdominal pain, characterized by epigastralgia without radiation and no response to spasmolytics or analgesics.

### *History of present illness*

The patient had been suffering intermittent episodes of mild discomfort of the gastrointestinal tract such as bearable diffuse abdominal pain and feeling of flatulence that persisted for one year. The pain pattern was not related to defecation or eating,

there was no nausea, vomiting, weight loss, melaena, change in bowel habit, urinary symptoms, or fever. She was managed conservatively as thought to be a functional gastrointestinal disorder.

She describes the pain as aggravating suddenly and sharp in nature. She presented to the emergency department after 48 h of the pain acutely worsened. The pain was in the epigastrium and across the anterior abdomen, was sharp and constant without radiation.

### History of past illness

She had no antecedents of alcohol, tobacco, or drug abuse.

### Personal and family history

She had a medical history of arterial hypertension and trigeminal neuralgia and had no surgical history. In her family history, there were two cases of colorectal cancer (mother and sister) without other illnesses.

### Physical examination

The patient experienced epigastric tenderness upon palpation, although she had no rebound tenderness, muscle tension, or a palpable mass. She had no other relevant findings.

### Laboratory examinations

Laboratory test results including complete blood count, liver function tests, serum amylase and lipase, biochemistry, were within normal ranges.

### Imaging examinations

An abdominal computed tomography (CT) scan showed a solid mass of the tail of the pancreas that contacted the lesser curvature of the gastric body. Magnetic resonance imaging (MRI) showed a pancreatic head, uncinate process, neck, and body of normal morphology. A solid nodular mass 29 mm in diameter was confirmed in the pancreatic tail, hypointense in T1, heterogeneous with hyperintense areas in T2, with enhancement after the administration of i.v. contrast predominantly towards the latter phase. Severe atrophy of the tail of the pancreas and upstream dilation of the main pancreatic duct was observed. The intra- and extrahepatic bile ducts were of normal caliber. This hypovascular nodule was highly suspicious of malignancy, probably PDAC. No regional or distant metastases were visualized in the abdomen (Figure 1).

Complementary imaging studies for staging were performed. Thorax CT revealed 10 solid nodules between 3-6 mm distributed in both lungs, which, due to their distribution, were suspicious of secondary implants.

## FINAL DIAGNOSIS

Video-thoracoscopy was performed, and these nodules had the characteristic appearance of benign anthracotic nodules, a type of pneumoconiosis caused by repeated exposure to air pollution or coal dust particles[7]. Biopsies were performed, and the benign nature was confirmed by histology.

Given these findings of no extra-abdominal disease, surgery was performed. Distal pancreatectomy with *en bloc* splenectomy was performed. Following surgery, the patient recovered successfully and was discharged from the hospital after 5 d.

Gross examination of the resected specimen revealed the tumor was localized in the tail of the pancreas, measured 3.2 cm × 2.9 cm, and consisted of a solid mass. Margins of surgical resection were free of tumor. Microscopically, the tumor was consistent with ductal adenocarcinoma with sarcomatoid features (Figure 2). Immunohistochemistry showed that the tumor had both epithelial and mesenchymal markers that were positive for pan-cytokeratin (Figure 2D), vimentin (Figure 2C), and smooth muscle actin (SMA) and negative for CD68. Thus, a diagnosis of SCP was confirmed.

The tumor was confined to the tail of the pancreas with no invasion to the spleen. All surgical margins were free of tumor tissue. There was no evidence of perineural invasion but lymphovascular permeation of one of thirty peripancreatic lymph nodes were positive for metastatic cancer.

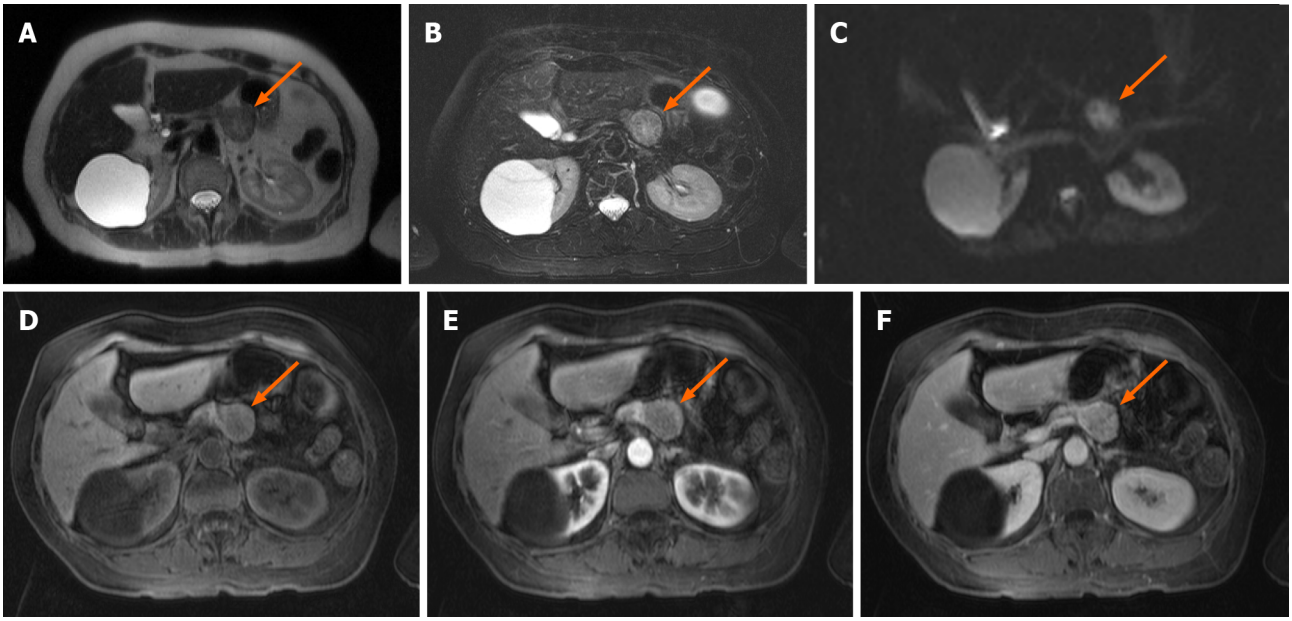


**Figure 1 Magnetic resonance imaging scan of the abdomen showing a distal pancreatic mass of 29 mm.** A: On the T2-weighted image, the lesion contained mixed signals (orange arrow); B: T2 fat saturation; C: Diffusion-weighted; D: T1-weighted fat sat gadolinium; E and F: T1-weighted image during arterial and portal phase that shows a hypovascular lesion.

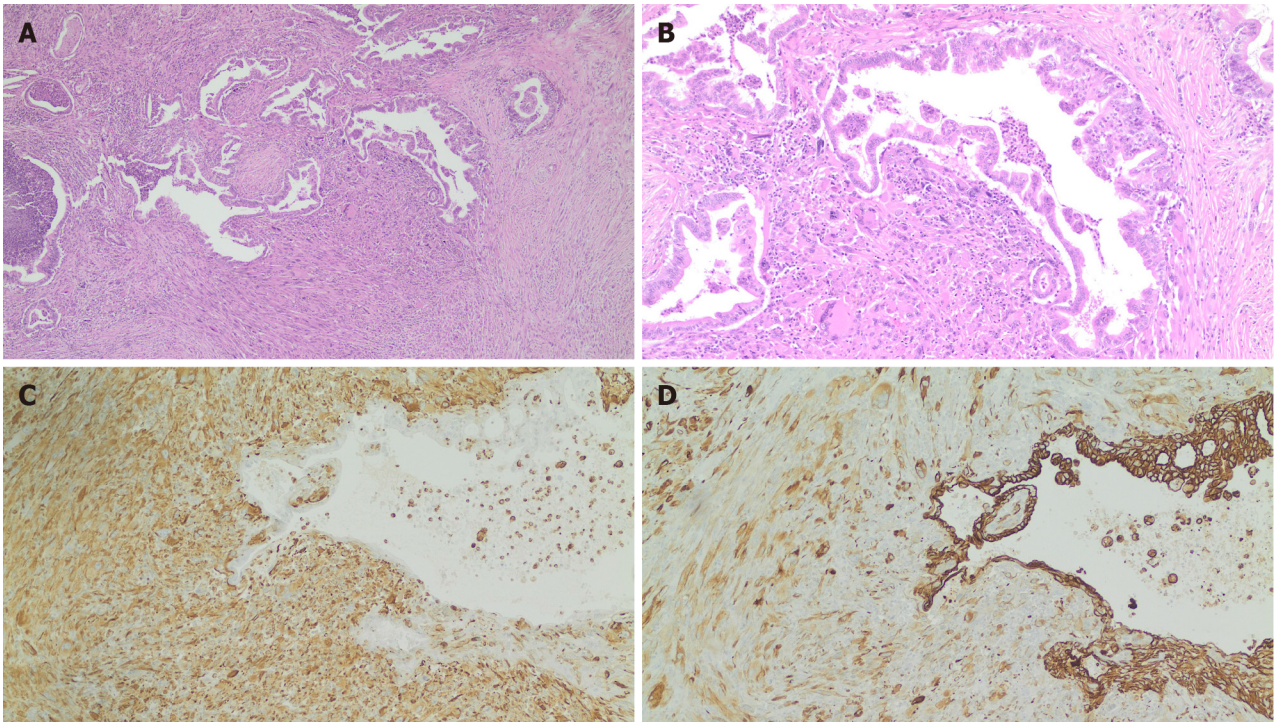


**Figure 2 Histological examination and immunohistochemical staining.** A and B: Histological examination of the pancreatic neoplasm reveals infiltration by malignant cells displaying a glandular and spindle-cell pattern. Hematoxylin and eosin, 4 × (A); Hematoxylin and eosin, 10 × (B). Glandular component lined by atypical epithelium and sarcomatous spindle cell component with pleomorphic giant cells; C: Immunohistochemical staining for pan-cytokeratin, 10 × (C). Glandular and sarcomatoid components are positive for this epithelial marker; D: Immunohistochemical stain for vimentin, 10 × (D). Vimentin is the most common mesenchymal marker. The epithelial glandular component is negative, and the sarcomatoid component is strongly positive.

## TREATMENT

The oncologic committee disclosure was that the patient should receive postoperative adjuvant chemotherapy with gemcitabine and capecitabine. Unfortunately, this could not be carried out for extra medical reasons since the patient's medical insurance did not cover this treatment.

## OUTCOME AND FOLLOW-UP

As our patient did not have access to adjuvant chemotherapy, we performed follow-up every 6 mo with general laboratory exams and imaging of the abdomen. The last image obtained was an abdominal CT after 35 mo of curative surgery, which did not reveal tumor recurrence.

## DISCUSSION

Sarcomatoid carcinomas are uncommon aggressive histologic variants of carcinoma. Although they may rarely arise in almost any organ, the lung, breast, and kidney are the most common primary sites[8]. Several terms have been used to describe this malignancy, including carcinosarcoma, pseudosarcoma, pseudocarcinoma, and spindle cell carcinoma[9]. The multiple names demonstrate the varied understanding of this disease, which these terms have been often used interchangeably, and their definitions vary among the reports[10]. According to the WHO classification (5$^{th}$ edition, 2019) of pancreatic tumors assigns SCP under the category of undifferentiated carcinomas (UCP)[4]. UCP is a subtype of PDAC representing a set of rare tumors that accounts for as many as 5% of all pancreatic malignancies[11]. UPC is categorized into two different types: undifferentiated carcinoma [with three variants: anaplastic undifferentiated carcinoma, sarcomatoid carcinoma (SCP), and carcinosarcoma] and undifferentiated carcinoma with osteoclast-like giant cells[5,12]. Hence, we present a case of SCP that is an extremely rare type of tumor, with only a few cases reported in the literature[5,9,10,13-21,31-36]

SCP is defined histologically as a poorly differentiated tumor composed by the proliferation of spindle cells with evidence of epithelial differentiation. Sarcomatoid carcinomas can exhibit a monophasic or biphasic appearance. The monophasic pattern often referred to as spindle cell carcinoma, is akin to a soft tissue sarcoma without epithelioid areas. The biphasic pattern features a mixture of mesenchymal-like and epithelial-like cells with a transition zone. The sarcomatous tissue of these tumors shows evidence of epithelial differentiation, such as epithelial markers and epithelial ultrastructural features, rather than a specific line of mesenchymal differentiation[14].

The diagnosis often represents a clinicopathologic challenge, and immunohistochemistry plays a key role in the histopathological diagnosis where an epithelial immunohistochemical profile assembles PDAC[6,22]. In immunohistochemistry, undifferentiated cells often express both broad lineage carcinoma (pan-cytokeratin) and sarcoma (vimentin and desmin) markers and display a loss of E-cadherin[12]. Its pathogenesis remains unclear[23,24].

Owing to the rarity of the disease, the clinical course, surgical outcomes, and optimal treatment strategies for SCP are poorly characterized[5].

To date, the largest study to analyze the histological spectrum of pancreatic carcinoma with sarcoma-like transformation was reported in 1977 by AlguacilGarcia and Weiland[25] who identified four distinctive histological types of sarcoma-like carcinoma based on light microscopic analysis only. Of twelve cases they reported an average survival of 8.3 mo for patients with nonresectable lesions.

In addition to our patient, 16 cases of SCP with confirmed both epithelial and sarcomatoid elements have been reported (Table 1).

Although SCP and "Carcinosarcoma" have different pathologic features, both share similar clinical features. Carcinosarcomas are considered to be "truly" biphasic neoplasms composed of intermingled carcinomatous and sarcomatous components, which have epithelial and mesenchymal differentiation. These two components are typically separated without a transition zone[14].

In previously published reports, the terms SPC and Carcinosarcoma have been often used interchangeably, and their definitions vary among the reports. On this basis, we excluded some articles in our summary of case reports (Table 1), when the terminology of "carcinosarcoma", "sarcoma-like" or "carcinosarcomatous histology" was used.

Recent publications have described the clinical and radiological features of UCP. Shiihara *et al*[6] aimed to identify the detailed clinicopathological features of UCP and revealed that these patients likely have abdominal pain or discomfort as an initial symptom, whereas jaundice was less common. It tends to present more commonly in men *vs* women with a ratio of 2.5:1 and occurs more frequently in the head of the pancreas[25]. Zhao *et al*[26] reported the radiologic features of SCP and found that the mean size of SCP was 5.1 cm, and most of the lesions appeared to be round or

**Table 1 Summary of reported cases of sarcomatoid carcinoma of the pancreas**

| Ref. | Age (yr)/ gender | Involved part of the pancreas | Tumor extension | Therapeutic schedule | Tumor size, cm | Sarcomatoid component | Follow-up time/results |
|---|---|---|---|---|---|---|---|
| Cresson *et al*[31], 1987 | 69/male | Head and tail | NA | Mitomycin, adriamycin, and 3000 rads of external radiation to the stomach | NA | Tubular structures, desmosomes, and hemijunctions under electron microscope | 5 mo/hemorrhage after surgery of metastasis in the jejunum |
| Higashi *et al*[19], 1999 | 74/male | Head | Head of the pancreas and the adjacent duodenum, with blood vessel and perineural sarcomatoid | Pylorus preserving pancreatoduodenectomy | 4.5 × 4 × 3 | CK AE1/AE3 (+), EMA (+), MUC1 (-), ARA (+), S100 (+), SMA (+), desmin (–), vimentin (–) | 3 mo/died after surgery of peritoneal carcinomatosis |
| Darvishian *et al*[32], 2002 | 74/male | Head | Peripancreatic adipose tissue and the duodenal wall. | Pancreatoduodenectomy | 4.0 × 3.0 | Vimentin (+), CK (+), CEA (+), SMA (+), desmin (+) and CD68 (-) | 4 mo/alive and well |
| De la Riva *et al*[33], 2006 | 72/female | Head | NA | NA | NA | CK and vimentin (+) | 9 mo/deceased with hepatic metastasis |
| Kim *et al* [21], 2006 | 73/female | Body and tail | Local invasion. With retroperitoneal lymph node with metastasis | Pancreatectomy with splenectomy and colonic segmental resection | 20 ×15 ×13 | CK (-), Vimentin (+), CD68 (+) | 4 mo/deceased secondary to hepatic and peritoneal metastases |
| Ren *et al* [13], 2013 | 48/male | Tail | Free surgical margins | Surgery N/A. Digital subtraction angiography interventional chemotherapy was then implemented. Gemcitabine, oxaliplatin, and floxuridine were intravenously injected *via* the superior mesenteric artery and celiac trunk artery. | 10 cm × 8 cm × 3.5 | Vimentin, α-1-antichymotrypsin, CK-19, CK-18, and pan-CK (+). CD68 and lysozyme (-) | 36 mo/alive and well |
| Yao *et al*[15], 2013 | 48/male | Tail | Free surgical margins | Laparoscopic spleen-preserving left pancreatectomy, adjuvant gemcitabine 1 cycle | 10 × 8 × 5 | CK 18 and vimentin (+) | 3 mo/tumor recurrence and death |
| Kane *et al*[9], 2014 | 85/male | Body | Local invasion with free surgical margins | A distal pancreatectomy, splenectomy, and partial gastrectomy | 3.3 × 3.0 × 2.6 | Pan-CK, CK5.2 (+), S100, SMA, EMA (-) | 26 mo/alive and well |
| Lai *et al* [34], 2015 | 55/male | Body and tail | NA | Distal pancreatectomy, splenectomy, and colonic segmental resection | 14 | CK, CK7, and vimentin (+) | NA |
| Nambiar *et al*[35], 2017 | 41/male | Head and uncinate | Liver metastasis | Gemcitabine | 2.2 × 2.1 | CK (+) and vimentin (+) | 1 mo/on chemotherapy when reported |
| Ruess *et al* [36], 2017 | 73/female | Head of pancreas | Free surgical margins | Extended pylorus-preserving pancreatoduodenectomy | 4.2 | Pan-CK1/3 (+), CK7 (+), CK19 (+). Vimentin (+). S100 (+) | 4 mo/death after surgery |
| Xie *et al* [16], 2018 | 63/male | Head of pancreas | Invasion of the distal common bile duct. Local invasion of the peripheral nerves. The lymph nodes, blood vessels, and resection margins were free from tumor tissue. | Pancreatoduodenectomy. 15 d of thymopeptides (1 mg per day). | 2.5 × 2 × 1.8 cm | Vimentin (+), CK7 (+), and CK19 (+) | 16 mo/hepatic metastasis |
| Bukhari and Joudeh[17], 2019 | 64/male | Head | Free surgical margins | Pancreatoduodenectomy with cholecystectomy and adjuvant gemcitabine | 2.4 × 2 × 1.9 | CAM 5.2 (-), vimentin (+) | 19 mo/alive and well |
| Zhou *et al* [14], 2019 | 59/male | Head | Pancreatic head with extension into the | Pancreatoduodenectomy | 2.5 × 2.5 × | CK19 (+) and vimentin (+) | 6 mo/liver metastasis and |

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| | | | main pancreatic duct. Free surgical margins. Three out of 23 lymph nodes were positive for metastasis | | 2.0 | | peritoneal metastasis |
| Kimura *et al*[10], 2020 | 58/male | Body | Three lymph nodes out of 40 with direct invasion | Distal pancreatectomy with splenectomy. A six-month course of gemcitabine | 5 | CK (+) and vimentin (+). PSmad2/3, snail, and fibronectin | 120 mo (10 yr)/alive and well |
| Omrani *et al*[18], 2020 | 73/male | Tail | NA | *En bloc* resection of the tail of the pancreas, spleen, a part of the stomach, and postoperative adjuvant chemotherapy with gemcitabine | 10 | OCG were positive for CK19 and CK7 | 120 mo (10 yr)/colonic metastasis |
| Our case | 61/female | Tail | Free surgical margins, one lymph node compromised | Distal pancreatectomy and *en-bloc* splenectomy | 3.2 × 2.9 | Pan-CK (+) and vimentin (+) | 35 mo/alive and well |

CEA: Carcinoembryonic antigen; (-): No positivity; (+): Positivity; CK: Cytokeratin; EMA: Epithelial membrane antigen; NA: Not available; OCG: Osteoclastic giant cells; SMA: Smooth muscle actin; MUC1-ARA: Apoprotein MUC1.

ellipsoidal in shape and were ill-defined. Vascular invasion by CT and MRI was reported in 5 of 10 lesions[26]. At the time of diagnosis, a bulky tumor is frequently detected, with the involvement of organs in the vicinity[27]. One of the imaging key signs for PDAC is the abrupt "cut-off" of the main pancreatic duct (MPD), with upstream MPD dilatation and substantial pancreatic atrophy[28]. Zhao *et al*[26] reported that eight of ten patients with SCP had upstream dilatation of the MPD. Among them, in three patients MPD was compressed by the lesions and no atrophy of the distal pancreatic parenchyma. In the other five patients, upstream MPD dilatation and distal pancreatic parenchyma atrophy were detected synchronously in only two patients while no atrophy was detected in the remaining three patients[26].

Because there is no standard regimen for treating SCP, patients with this disease are administered the same regimens as those with more common PDACs. Gemcitabine has been reported to be effective in the event of portal vein thrombosis or tumor recurrence, whereas a cisplatin/etoposide/ifosfamide (VIP) regimen was also found to produce notable results[6]. Imaoka *et al*[29] conducted a multicenter retrospective cohort study to investigate the efficacy of chemotherapy in patients with UCP ($n$ = 50) showing a median overall survival (OS) of 4.08 mo. The most frequently used first-line treatment regimens were gemcitabine, S-1, and gemcitabine plus nab-paclitaxel. Although there was no significant difference in OS among these first-line regimens, gemcitabine plus nab-paclitaxel significantly improved median progression-free survival compared with gemcitabine alone[29].

Although treatment for PDAC remains challenging, complete R0 surgical extirpation is the only chance of cure[5]. Although SCP shares similar molecular carcinogenesis with PDAC, its prognosis is much worse[6]. Despite aggressive surgical management, the median postoperative survival has consistently been reported as less than 1 year, and almost all recurrences involve unresectable multiple metastases[6,17,21,23,30]. Of the previously reported cases of SCP (Table 1), we calculated the mean survival time of the patients using the Kaplan-Meier method, showing a median OS of 9 mo (range 0-27), with 5-year and 10-year survival rates of 41.25% and 20.63% respectively (Figure 3).

Furthermore, the impact of adjuvant chemotherapy on the survival of SCP has not been well defined. Imaoka *et al*[29] report that a paclitaxel-containing regimen would offer relatively longer survival in patients with unresectable UCP.

Given its aggressive biological behavior and poor prognosis, it is of prime importance to make early diagnoses for patients with SCP[22]. Although some patients have a rapid recurrence and early death, long-term survival has been reported[5,10]. Blair *et al*[5] reported 8 cases of SCP, of which two experienced long-term survival (> 5 years), with the longest surviving nearly 16 years despite the presence of lymph node metastasis representing the longest survival time of SPC patients in the literature. Nevertheless, both long-term survivors had the tumor in the body/tail of the pancreas, underwent R0 resections, and received adjuvant therapy.

There are two reports of exceptional survival after ten years of follow-up. One of them received adjuvant chemotherapy with gemcitabine who remained free of tumor recurrence and metastasis for 10 years but after this period the patient presented a colonic obstruction due to metastatic disease[18]. In the other case of SCP with a stage

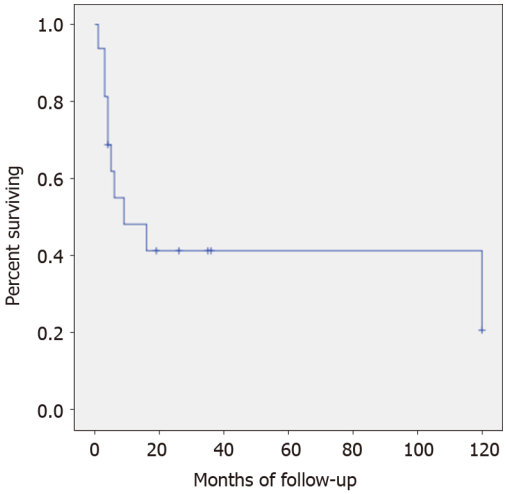


**Figure 3 Survival curves using the Kaplan-Meier method showing the overall survival of reported cases of sarcomatoid carcinomas.** The median overall survival times in patients with sarcomatoid carcinoma of the pancreas were 9 mo (range 0-27). The 5-year and 10-year survival rates were 41.25% and 20.63% respectively.

T3N1M0, after surgery the patient completed a 6-month course of adjuvant chemotherapy with gemcitabine and was then followed up with abdomen CT. At 10 years after the operation, the authors report he is alive with no recurrence[10].

SCP reported in the present paper is a very rare case of primary pancreatic neoplasm. Based on the limited number of reported cases, the prognosis is poor. To our knowledge, the good evolution of our patient, tumor-free survival of 35 mo after surgery despite not receiving adjuvant chemotherapy treatment, is rather exceptional particularly after having lymphovascular invasion. Although our patient had a smaller tumor size compared to the other long-term survival cases, Paal *et al*[25] reported in 35 cases of UCP that overall tumor size is not a reliable prognostic indicator. In this case, the clinical presentation with acute abdominal pain aided in obtaining a relatively early diagnosis and better surgical results.

## CONCLUSION

Sarcomatoid carcinoma is a rare aggressive tumor with a poor prognosis. With an early diagnosis with early surgical eradication of the tumor and adjuvant chemotherapy, evolution may be exceptionally favorable with long survival. The patient described in this case study is alive and without metastasis 35 mo after surgery despite not receiving chemotherapy.

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