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## Pancreas angiosarcoma—Case report of a rare cause of abdominal pain

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## ABSTRACT

**INTRODUCTION:** Pancreas angiosarcoma is a very aggressive malignant neoplasm. The symptoms are nonspecific and it is usually diagnosed at an advanced stage, which confers a poor prognosis.**PRESENTATION OF CASE:** We present a 56-year-old woman with abdominal epigastric pain and nausea. The abdominal CT-scan showed a 7 cm mass within the head of the pancreas and the pathology and immunochemistry analysis were positive for pancreas angiosarcoma. Intra-operatively the tumor was irresectable.**DISCUSSION:** Pancreas angiosarcoma is an extremely rare neoplasm with non-specific diagnosis. The histology has a wide range of presentations and immunohistochemistry is required. The surgery appears to be the only effective treatment.**CONCLUSION:** We report the seventh case of pancreas angiosarcoma in the English literature. Despite its irresectability, the patient was asymptomatic two months after surgery, initiating chemotherapy with paclitaxel, with good tolerance.© 2020 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 1. Introduction

Angiosarcomas are highly malignant endothelial-cell tumors of vascular or lymphatic origin [1,2], representing less than 2% of all soft tissue sarcomas [3]. Angiosarcomas of the pancreas are extremely rare, with only six cases reported in the literature [4,5]. They are fast growing and very aggressive tumors with a poor prognosis [6,6]. Since the symptoms are nonspecific, they are usually diagnosed at an advanced stage [7]. Surgery appears to be the only effective treatment approach in localized disease [4,6]. We report the seventh case of pancreas angiosarcoma in the English literature. This work is reported in line with SCARE 2018 criteria [8].

## 2. Presentation of case

A 56-year-old woman was admitted in our hospital with two months history of abdominal epigastric pain and nausea. She was a housewife with no known exposure to carcinogens and drugs and with no relevant family and psychosocial history. Her medical and surgical history was unremarkable, except for appendicectomy and tubal ligation decades before. Laboratory tests showed

a diminished haemoglobin level (11 g/dL), CEA 1,8 ng/mL (Reference range: < 5), CA 19.9 8,9 U/mL (RR: 0–37), chromogranin A 21,7 ng/mL (RR: <102) and NSE 10,9 ug/L (RR: 0–16,3). A contrast enhanced CT-scan (Fig. 1) revealed a 7 cm mass within the head of the pancreas with no apparent vascular or surrounding organ invasion. There was no distant spread location. Pancreatic biopsy through ultrasound was performed and demonstrated atypical and pleomorphic malignant endothelial cells, that are the hallmark of angiosarcoma. Kocher laparotomy, revealed an 9 × 9 cm head pancreatic mass, with superior mesenteric artery invasion and compression of the third portion of duodenum, thus revealing itself intra-operatively irresectable. A new biopsy was done and a gastro-jejunostomy with Braun's jejunostomy was performed by a general surgery with 20 years of experience in hepatobiliopancreatic surgery. The histological examination of the pancreatic biopsy (Fig. 2) showed a solid pattern neoplasm consisting of spindle and epithelioid cells, richly vascularized with moderate to intense nuclear pleomorphism and the presence of atypical endothelial cells. Immunohistochemical studies (Fig. 3) demonstrated positive staining for vimentin and endothelial markers CD34, CD31, factor VIII and Fli-1, but negativity for AE1/AE3, SMA, desmin, DOG-1, CD117 and pS100, thus establishing the diagnosis of pancreatic angiosarcoma.

The patient was discharged on postoperative day 4 with no complications. Based on multidisciplinary discussion, she was proposed for palliative chemotherapy with paclitaxel beginning on

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Fig. 1. Contrast enhanced CT scan showed the tumor in the head of the pancreas.

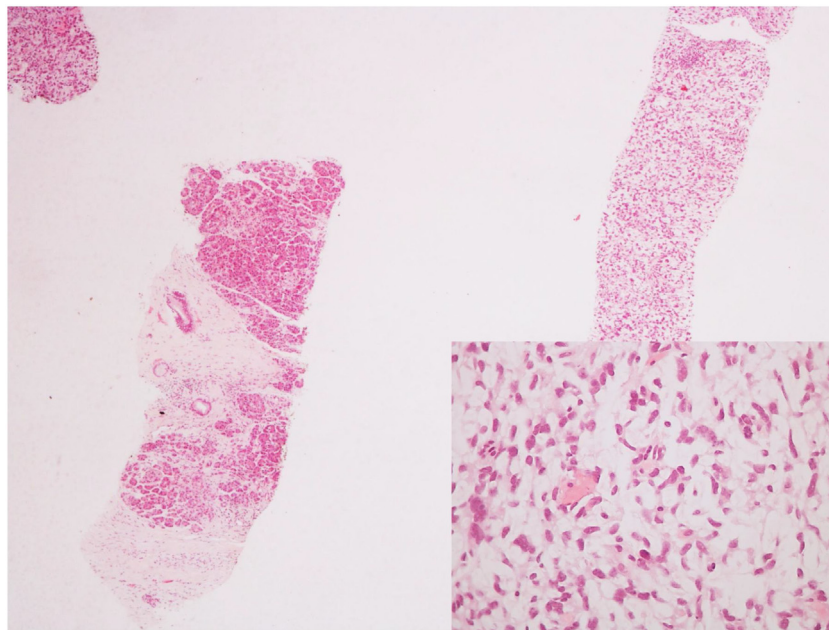


Fig. 2. Biopsy specimen showing pancreatic tissue with a solid, vessel-rich pattern and high grade mesenchymatous neoplastic cells (HE x50 and HE x400).

day 26 after surgery. The patient understood and adhered fully to the treatment plan. At two months after surgery she maintains chemotherapy and has no tumor-related symptoms.

### 3. Discussion

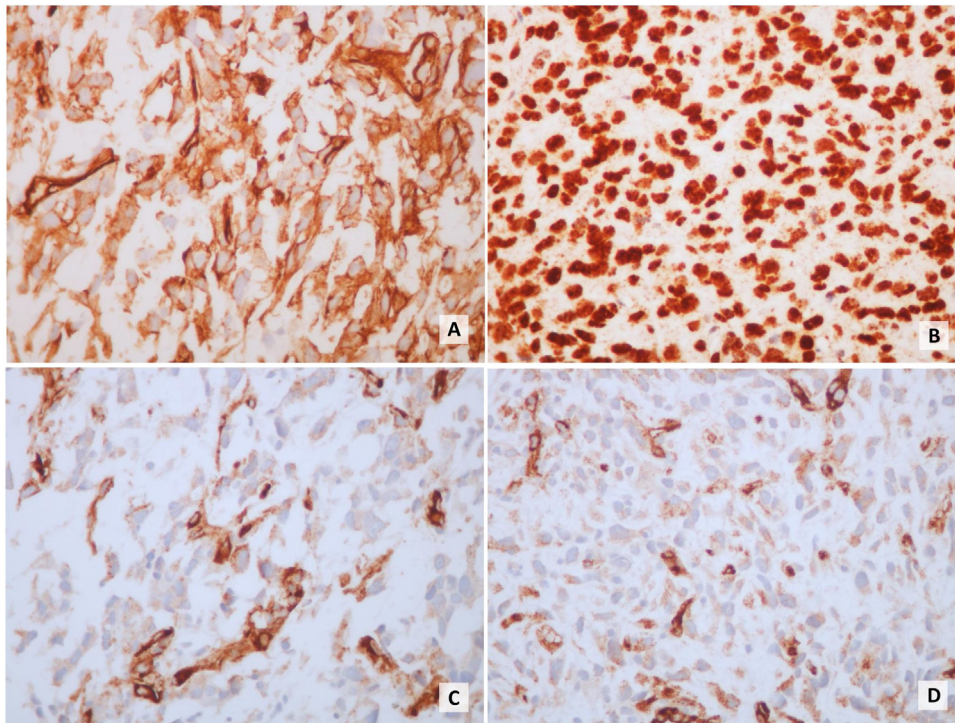
Angiosarcomas represent vascular neoplasms with a very low incidence, even within sarcomas [9]. A pancreatic primary origin is especially rare, with the present case being the seventh accounted in the English literature. Table 1 summarizes the most important features of the primary pancreatic angiosarcomas published to date. More common origins of angiosarcomas are the head, neck and breast. In the abdominal compartment, angiosarcomas develop mostly in the liver and the spleen [1]. Regarding of the low number of cases worldwide, this is still a rather unknown entity. However, risk factors seem to include age, radiation, chemical carcinogens

(thorium dioxide and vinyl chloride), chronic lymphedema and some hereditary syndromes [10].

The histology findings are non-specific, with a wide range of histological appearances, often mimicking carcinomas, especially when showing epithelioid-cell morphology. Therefore, immunohistochemical staining is mandatory. Positive staining for vascular markers, which include CD31, CD34, Factor-VIII, Ulex europaeus agglutinin 1 (UEA-1), Friend leukemia integration 1 (Flt-1), endothelin-1, vascular endothelial growth factor receptor (VEGFR) and von-Willebrand factor (vWF), is typical. CD31, vWF and UEA-1 are believed to be positive in less differentiated tumors. The expression of epithelial markers, such as low-molecular weight cytokeratins or the epithelial membrane antigen (EMA) may be found both in angiosarcomas and carcinomas and is thereby unhelpful in their distinction. Lymphoid markers (CD45 or CD30) and melanoma markers (S100) are typically negative [1].

**Table 1**  
Review of reported cases of literature.

Article	Age	Sex	Symptoms	Laboratory	CT scan	Surgery	Histology	Immunohistochemical profile	Follow-up
Seth et al. [5]	83	Woman	Recurrent GI bleeding	CA 19.9 284.7 (0–36) U/mL	Mass in the head of the pancreas	Pylorus-preserving pancreaticoduodenectomy	4.5 × 3.5 × 2.7 cm solid tumor → high-grade epithelioid angiosarcoma	CD31 CD34	Postoperative sepsis and death at day 15 post-op
Maeyashiki et al. [3]	72	Man	Dizziness and black stools	–	Multiple masses	Endoscopic splenectomy and partial enterectomy	Gastrointestinal and pancreatic angiosarcoma	CD31 CD34 Factor VIII	Hemorrhagic shock and death at day 103 post-op
Csiszkó et al. [4]	58	Man	Upper abdominal pain, nausea and fever	Normal CA 19.9	Acute haemorrhagic necrotizing pancreatitis with multiple peripancreatic fluid collection	Necrosectomy and peripancreatic drainage	Angiosarcoma with acute haemorrhagic pancreatitis.	Vimentin CD31 Cytokeratin Factor XIII	Multiorgan Failure, sepsis and dead at day 5 postoperative
Meeks et al. [2]	65	Man	Severe acute abdominal pain	CA 19.9 167 U/mL (0–35)	2.6 cm low attenuation mass within the proximal pancreatic body	Pancreaticoduodenectomy	3.0 × 1.8 × 1.6 cm mass located in the head of the pancreas suggestive of epithelioid angiosarcoma	CD31 CD34 Cytokeratin CK7 Pancytokeratin AE1/AE3	Abdominal compartment syndrome with bowel ischemia and death at day 6 post-op
Worth et al. [7]	78	Woman	Swollen inguinal lymph nodes	–	3.1 × 2.4 cm lesion on the anterior surface of the pancreatic tail.	Robotic-assisted distal pancreatectomy and splenectomy	2.4 × 1.6 × 1.6 cm vascular neoplasm with cystic change	Moderate increase in Ki-67 proliferation CD31 CD34	One-year follow-up: no evidence of recurrence
Darré et al. [6]	41	Man	Epigastric pain, jaundice, nausea and vomiting	–	Heterogeneous tumor of the pancreas body, measuring 63 × 46 mm	Impossibility of tumor resection	Tumor proliferation of vascular layers	CD31 CD34 Factor VIII	The patient died two weeks after the diagnosis



**Fig. 3.** Immunohistochemical staining, with positivity for CD34 (A), Fli-1 (B), CD31 (C) and Factor VIII (D).

Unlike the majority of the cases reported so far, which had a severe clinical presentation, ours was discovered only with mild abdominal pain. In this case, the resection of the tumor proved to be impossible intra-operatively, because of superior mesenteric artery invasion. Due to the strong duodenal adhesion and probable invasion, a gastrojejunostomy was performed. The alternative therapeutic options (chemo- and radiotherapy) have shown poor results in this type of neoplasms. Surgery remains the only known curative treatment. However, given the lack of success of the performed surgery, paclitaxel-based chemotherapy was proposed in a disciplinary discussion, since this agent seems to have a relevant activity against angiosarcomas of other origins [11].

At the date of submission of this article, four months after the diagnosis, no signs or symptoms of tumor expansion or metastasis have shown up, and no complications of chemotherapy have been reported.

#### 4. Conclusion

Pancreatic angiosarcoma is a very rare entity. Consequently, little is known about risk factors, tumor behavior and efficacy of the therapeutic options available. Imaging features are non-specific, with the diagnosis being only established through histology and immunohistochemistry analysis. The prognosis is dismal, especially when symptoms are already present. Surgical resection seems to be the only curative option, with radio- and chemotherapy being palliative alternatives.

#### Declaration of Competing Interest

The authors report no declarations of interest.

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#### Ethical approval

Not applicable.

#### Consent

The patient gave informed consent for publication of the case report with all accompanying images.

#### Author contribution

Aldara Faria, Filipe Lopes – study design, data collection, data analysis and interpretation and writing paper.

Alberto Figueira, Carlos Miranda, João Coutinho – data interpretation and corrections.

#### Guarantor

The guarantor of the case report is Aldara Faria.

#### Provenance and peer review

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**References**

- [1] R.J. Young, N.J. Brown, M.W. Reed, D. Hughes, P.J. Woll, Angiosarcoma, *Lancet Oncol.* 11 (2010) 983–991.
- [2] M. Meeks, et al., Primary angiosarcoma of the pancreas, *J. Gastrointest. Cancer* 48 (2017) 369–372.
- [3] C. Maeyashiki, N. Nagata, N. Uemura, Angiosarcoma involving solid organs and the gastrointestinal tract with life-threatening bleeding, *Case Rep. Gastroenterol.* 6 (2012) 772–777.
- [4] A. Csiszkó, et al., Primary angiosarcoma of the pancreas mimicking severe acute pancreatitis - case report, *Pancreatology* 15 (2015) 84–87.
- [5] A.K. Seth, Angiosarcoma of the pancreas - discussion of a rare epithelioid neoplasm, *Pancreas* 37 (2008) 230–231.
- [6] T. Darré, et al., Primary angiosarcoma pancreas: a case report of an exceptional localization, *J. Gastrointest. Cancer* 50 (2018) 935–938.
- [7] P.J. Worth, M. Turner, C.W. Hammill, Incidental angiosarcoma of the pancreas: a case report of a rare, asymptomatic tumor, *J. Pancreat. Cancer* 3 (2017) 24–27.
- [8] R.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, For the SCARE Group, The SCARE 2018 statement: updating consensus Surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 60 (2018) 132–136.
- [9] A. Cioffi, S. Reichert, C.R. Antonescu, R.G. Maki, Angiosarcomas and other sarcomas of endothelial origin, *Hematol. Oncol. Clin. North Am.* 27 (2013) 975–988.
- [10] A.H. Gaballah, et al., Angiosarcoma: clinical and imaging features from head to toe, *Br. J. Radiol.* 90 (2017).
- [11] N. Penel, et al., Phase II trial of weekly paclitaxel for unresectable angiosarcoma: the ANGIOTAX study, *J. Clin. Oncol.* 26 (2008) 5269–5274.

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