

Primary Pancreatic Undifferentiated Pleomorphic Sarcoma

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Introduction

- Primary pancreatic sarcomas are rare malignancies with an incidence of 0.1% [1, 2].
- Sarcomas are frequently seen in the surrounding pancreatic tissue, hence it's important to differentiate primary pancreatic sarcomas from other peripancreatic tumors [3].
- We report a case of high-grade primary undifferentiated pleomorphic sarcoma (UPS) of the pancreas.

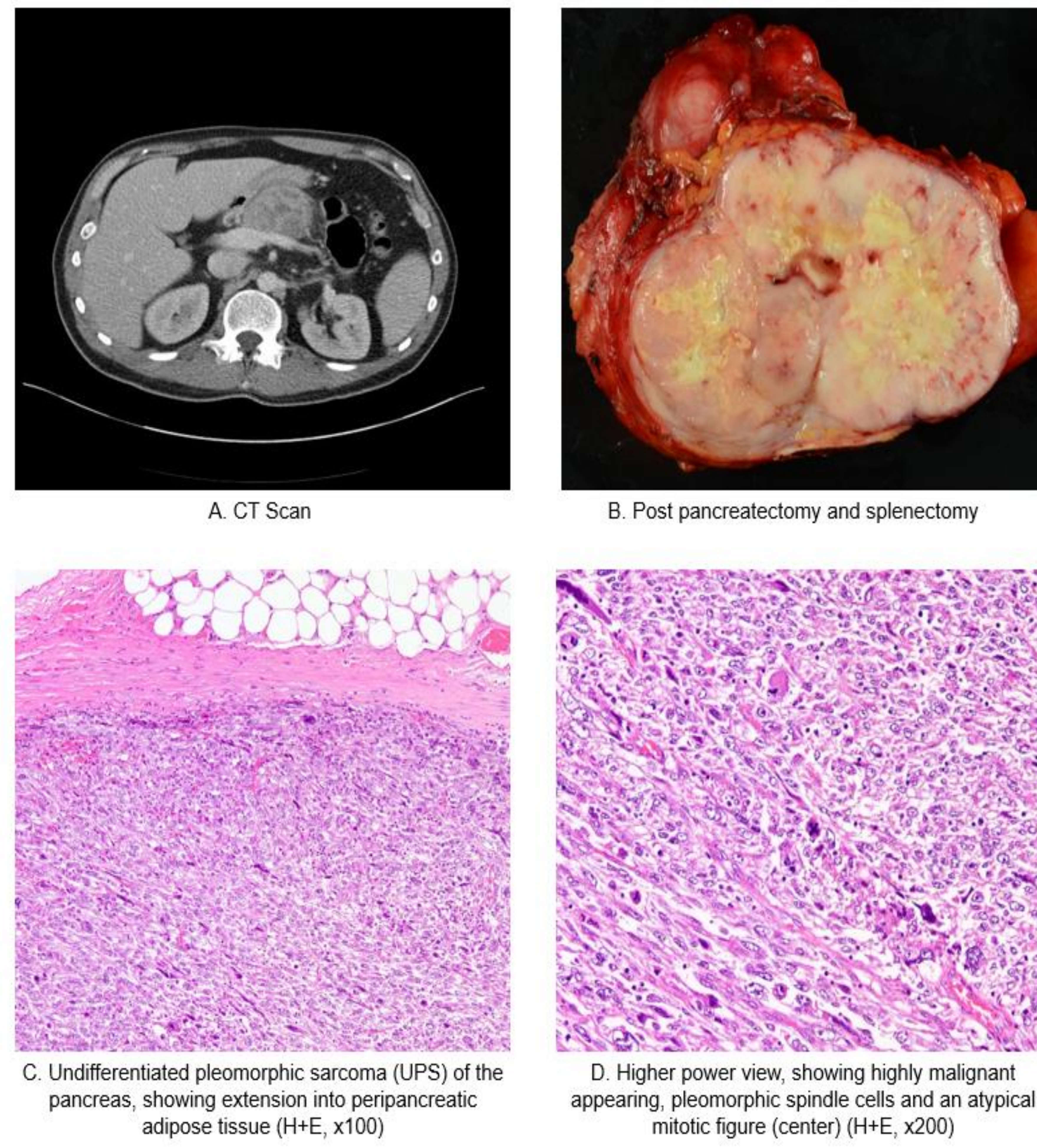
Case Presentation

- A 48-year-old man presented with acute LLQ abdominal pain and weight loss.
- He had a family history of colorectal cancer.
- Laboratory studies showed hemoglobin of 11.0 g/dL and a white blood cell count of 13.3/mm³.
- Liver panel and CA 19-9 were normal.
- Abdominal CT revealed a 6.8 cm complex solid-cystic pancreatic lesion (Figure 1A).
- Abdominal MRI showed a 9.2 cm macrolobulated mass involving the pancreas without local extension or metastasis.
- PET CT showed large fluorodeoxyglucose (FDG) avid centrally necrotic pancreatic mass without metastasis.
- Endoscopic ultrasound revealed a 6.5 cm solid-cystic mass in the pancreatic body with no vascular involvement.
- Fine needle biopsies revealed malignant spindle cells.
- Immunohistochemistry (IHC) failed to characterize lineage and differential included carcinoma or sarcoma.
- He underwent distal pancreatectomy and splenectomy (Figure 1B).
- Intraoperative frozen section showed epithelioid and spindled malignancy with IHC negative for EMA, broad spectrum keratins (AE1/AE3 and OSCAR), and high molecular weight keratins.
- CKAE1AE3, CD163, Synaptophysin, CD117, CD34, CD31, SMA, S-100 and SOX-10 were also negative.
- Final pathology revealed high grade undifferentiated pleomorphic sarcoma measuring 10.5 cm with negative margins (Figure 1C&D).
- He underwent adjuvant chemotherapy with 4 cycles of doxorubicin/ifosfamide.
- Surveillance with CT chest/abdomen/pelvis every 3 months was recommended.
- Follow-up imaging has been unremarkable.

Discussion

- This case highlights a rare case of primary pancreatic UPS in a middle-aged man.
- A search of the literature revealed 16 cases of UPS with a similar clinical presentation (Table 1).
- A step-wise approach with cross-sectional imaging and biopsy is important to appropriately manage these rare malignancies [4].

Figure 1:



References

2. Ambe, P., et al., *Primary sarcoma of the pancreas, a rare histopathological entity. A case report with review of literature.* World J Surg Oncol, 2011. **9**: p. 85.
 3. Varela, A.B., et al., *Primary pancreatic sarcoma.* REVISTA ESPAÑOLA DE ENFERMEDADES DIGESTIVAS, 2009. **2009**(101/4): p. 285-287.
 4. Sanei, B., et al., *Undifferentiated pleomorphic sarcoma of pancreas: a case report and review of the literature for the last updates.* Case Reports in Medicine, 2018. **2018**.

Table 1. Summary of all Undifferentiated Pleomorphic Sarcoma of Pancreas report to date

Author	Age	Sex	Histologic type	Location	Treatment	Preoperative diagnosis	Postoperative therapy	Follow-up (months)
Ishiguchi, et al. ⁴	44	M	Pleomorphic	Body-tail	Left pancreatectomy, splenectomy	Pancreatic neoplasms	NA	15, NED
Garvey, et al. ⁵	77	M	Storiform-pleomorphic	Uncinate lobe	Enucleation	Pancreatic head mass	NA	48, NED
Pascal, et al. ⁹	39	M	Storiform-pleomorphic	Head	Pancreaticoduodenectomy	Mesenchymal tumor	NA	0, DOC
Allen, et al. ¹⁰	46	M	Storiform-pleomorphic	Body-tail, local invasion	Pancreatectomy, splenectomy, subtotal gastrectomy	NA	Chemotherapy	5, DOD
Tsujimura, et al. ¹¹	43	F	Storiform-pleomorphic	Tail	Pancreatectomy, splenectomy	Pancreatic cystadenoma	Chemotherapy	5, NED
Ben Jilani, et al. ¹²	72	M	Storiform-pleomorphic	Body-tail	Left pancreatectomy, splenectomy	Pancreatic mass	NA	12, DOD
Belen, et al. ¹³	37	M	Pleomorphic	Body-tail	Extended left pancreatectomy	Pancreatic mass	Radiotherapy and chemotherapy	7, DOD
Haba, et al. ¹⁴	70	M	Storiform-pleomorphic	Head	Pancreaticoduodenectomy	Tumor of pancreatic head	Chemotherapy	22, NED
Bastian, et al. ¹⁵	87	M	Storiform-pleomorphic	Body	Left pancreatectomy, splenectomy, transverse colectomy, subtotal gastrectomy	Pancreatic cancer	NA	34, NED
Darvishian, et al. ⁶	74	M	Storiform-pleomorphic	Head	Pancreaticoduodenectomy	Pancreatic head cancer	NA	4, NED
Akatsu, et al. ¹⁶	67	M	Storiform-pleomorphic	Body-tail	Left pancreatectomy, splenectomy, transverse colectomy, total gastrectomy	Pancreatic cancer	NA	35, NED
Mizukami, et al. ¹⁷	44	F	Pleomorphic	Body-tail	Total gastrectomy, left pancreatectomy	Pancreatic tumor	NA	20, NED
Yu, et al. ¹⁸	67	M	Storiform-pleomorphic	Head	Pancreaticoduodenectomy	Pancreatic head cyst	NA	11, DOD
Jamy, et al. ¹⁹	45	M	Storiform-pleomorphic	Head	Pancreaticoduodenectomy	Pancreatic cancer	Radiotherapy and chemotherapy	36, NED
Sanei, et al. ²	72	F	Pleomorphic	Head and neck	Pancreaticoduodenectomy	Pancreatic head cancer	NA	22, NED
Liang, et al. ⁷	37	F	Pleomorphic	Body-tail	Distal pancreatectomy	Pancreatic mass	NA	12, DOD
Own case	48	M	Pleomorphic	Body	Pancreatectomy with splenectomy	Pancreatic mass	Chemotherapy	12, NED



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