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CASE REPORT | PANCREAS

Primary Pancreatic Liposarcoma: An Unexpected Cause of a Pancreatic Mass

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ABSTRACT

Liposarcoma is the most common type of soft-tissue sarcoma and typically occurs in the extremities or retroperitoneum. Primary liposarcoma of the pancreas is exceedingly rare, with only 10 cases reported since 1979. We present a patient who was incidentally discovered to have a pancreatic mass on imaging, which was ultimately diagnosed as dedifferentiated pancreatic liposarcoma. We review the clinical and histologic features of pancreatic liposarcoma in this case and in the 10 previously reported cases to increase awareness and knowledge of this rare disease.

KEYWORDS: pancreatic liposarcoma; liposarcoma; soft-tissue sarcoma

INTRODUCTION

Liposarcoma is the most common type of soft-tissue sarcoma and accounts for approximately 20% of all mesenchymal neoplasms.[1] Liposarcoma typically occurs in the extremities or retroperitoneum and rarely occurs within visceral organs such as the pancreas. Most pancreatic tumors arise from epithelial cells with the most common type of pancreatic cancer being adenocarcinoma, which comprises approximately 85% of all pancreatic cancers. In comparison, only 1% of pancreatic tumors arise from mesenchymal cells.[2] Of the mesenchymal tumors, primary pancreatic liposarcoma is one of the rarest with only 10 cases described in the literature since 1979.[3–12]

We present a patient who was incidentally found to have a pancreatic mass, which was ultimately diagnosed as dedifferentiated pancreatic liposarcoma. The aim of this case report is to increase awareness of pancreatic liposarcoma and to add to the body of available literature of this rare diagnosis.

CASE REPORT

A 52-year-old previously healthy man sought medical evaluation for right shoulder and right upper chest wall pain. The patient had never smoked and had no history of excessive alcohol use. He had no other symptoms, and physical examination was unrevealing. Laboratory test results were obtained and were all within normal range, including a comprehensive metabolic panel and a complete blood count. After a negative cardiac workup, a computed tomography (CT) scan of the chest was obtained for further evaluation of right shoulder and upper chest wall pain, which incidentally noted a pancreatic mass. CT of the abdomen and pelvis was then obtained and revealed a 10-cm lobulated mass in the pancreatic tail with extension into the splenic hilum and gastric fundus (Figure 1).

An upper endoscopic ultrasound was performed to obtain biopsies of the lesion and revealed a large, atypical mass in the pancreatic tail with marked acoustic shadowing and areas of calcification (Figure 2). The mass was noted to be in direct contact with the gastric wall and spleen and encased the splenic artery resulting in splenic vein occlusion and gastric varices. The uninvolved pancreatic

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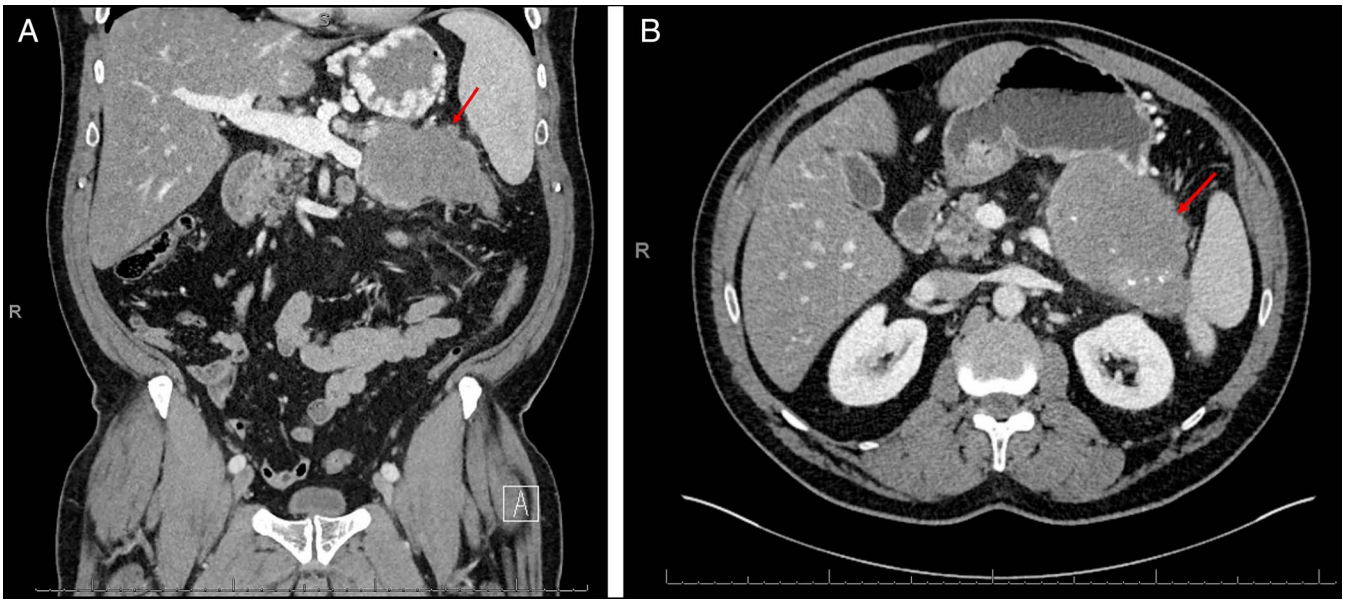

Figure 1. (A) A 10-cm lobulated mass is seen in the tail of the pancreas (arrow). (B) The mass can be seen extending into the hilum of the spleen and abuts the wall of the gastric fundus. Scattered internal calcifications are also seen.

parenchyma was hyperechoic suggestive of fatty infiltration, but there was no evidence of chronic pancreatitis. Biopsies of the mass were obtained. An enlarged lymph node adjacent to the left adrenal gland was noted, which was also biopsied. No other masses or enlarged lymph nodes were visualized. A large gallstone was seen in the gallbladder, which was suspected to be the underlying cause of his shoulder pain.

Biopsies of the mass and adjacent lymph node showed atypical spindle cells with immunohistochemical staining positive for CDK4 and MDM2 (Figure 3). This was strongly suggestive of liposarcoma, although excisional biopsy was recommended for definitive diagnosis. After a multidisciplinary tumor board presentation and discussion with the patient, the patient decided to undergo radical surgical resection of the mass and surrounding structures (distal pancreatectomy, partial gastrectomy, splenectomy, left hemicolectomy, and left adrenalectomy) and the adjacent enlarged lymph node with atypical spindle cells on biopsy. Gross pathology showed a tan-white, whorled, ill-defined mass with focal areas of calcification. The final pathology report confirmed dedifferentiated primary pancreatic liposarcoma with invasion into the spleen, gastric wall, and left colon wall (pT3). Tumor extended to the resection margins. Adjuvant chemotherapy with pembrolizumab was discussed, although the patient has not yet started this because of insurance issues. Surveillance PET-CT scan was obtained 3 months after surgical resection and did not show any clear evidence of metastatic disease or recurrence. To date, there has been no evidence of recurrence on surveillance imaging after 12 months.

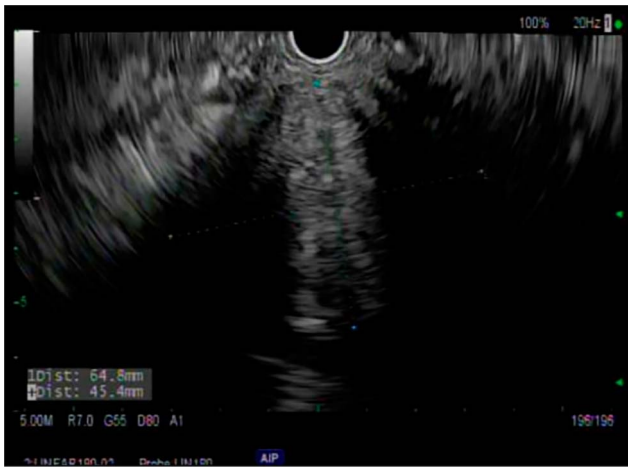

Figure 2. A large mass measuring 65 mm × 45 mm in diameter was seen in the pancreatic tail. It had scattered shadowing regions and poor through-transmission, precluding examination of deeper structures.

DISCUSSION

Liposarcomas comprise approximately 50% of soft-tissue sarcomas and typically occur on an extremity.[13] They are also one of the most common types of retroperitoneal sarcomas; however, primary liposarcoma of the pancreas is exceedingly rare, with only 10 cases reported since 1979.

We present a patient who was incidentally diagnosed with dedifferentiated pancreatic liposarcoma after initially presenting with referred shoulder pain believed to be secondary to symptomatic cholelithiasis. Treatment involved distal pancreatectomy and surgical resection of adjacent structures with evidence of tumor invasion. To date, the patient has no evidence of recurrence on surveillance imaging after 12 months.

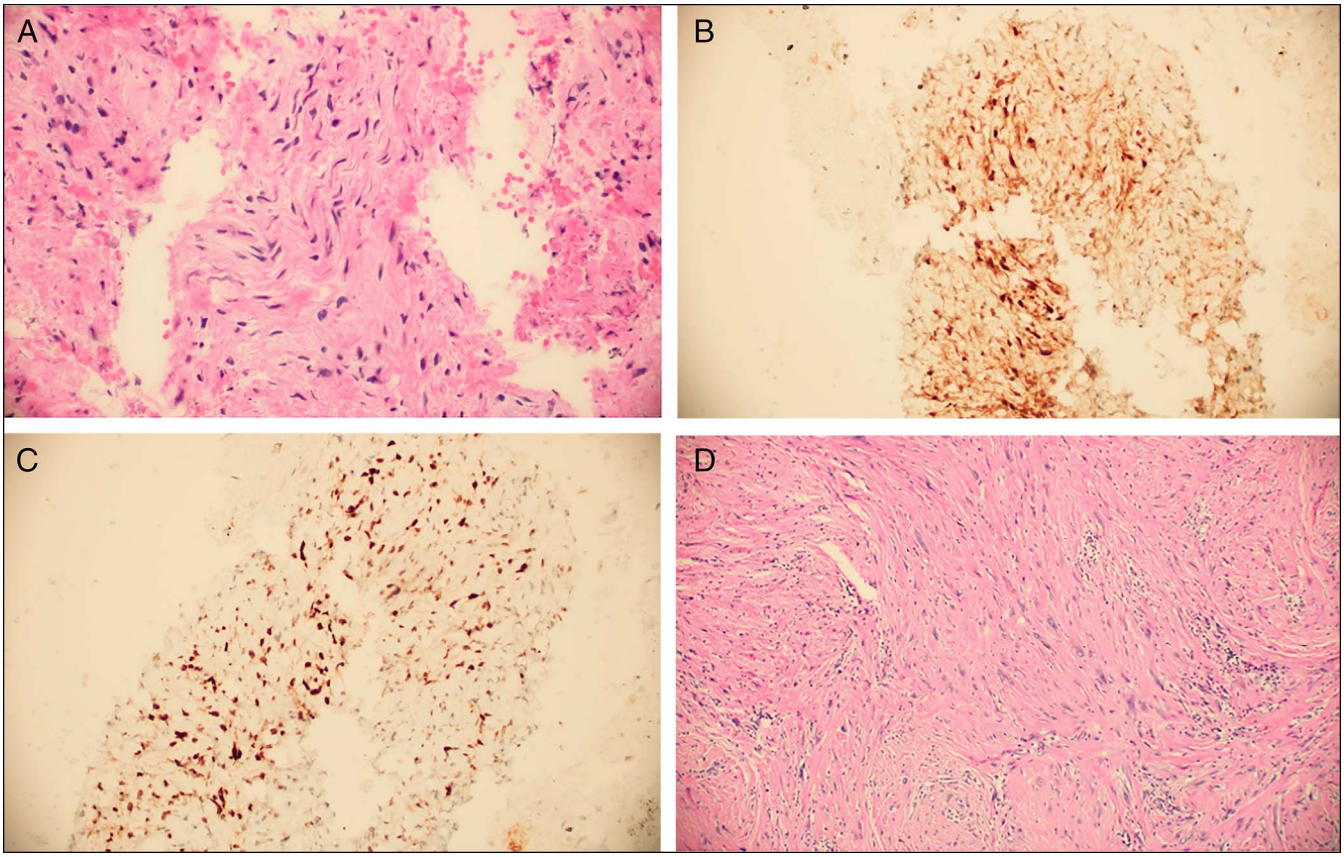

Figure 3. (A) Cell block cytology specimen showing a spindle cell proliferation composed of bundles of spindle cells with relatively mild nuclear atypia and absence of well-differentiated liposarcoma component. This morphology is nonspecific (H&E stain, 20x magnification). (B) CDK4 immunostain on the cell block tissue showing positive staining of the tumor cells (20x magnification). (C) MDM2 immunostain on the cell block tissue showing positive nuclear staining of the tumor cells supporting the diagnosis of liposarcoma (20x magnification). (D) Resection specimen showing dedifferentiated component of liposarcoma with significant nuclear pleomorphism characterized by nuclear size variability, irregular nuclear membranes, and prominent nucleoli (H&E stain, 20x magnification). H&E, hematoxylin and eosin.

Liposarcoma is classified histologically into 4 subtypes: well-differentiated, dedifferentiated, myxoid/round cell, and pleomorphic liposarcoma. Together, well-differentiated and dedifferentiated subtypes make up most cases of liposarcoma. Dedifferentiated sarcoma arises from transformation of well-differentiated liposarcoma in 10% of cases but can also occur de novo or as a recurrence of well-differentiated liposarcoma.[1] Dedifferentiated liposarcoma is associated with higher rates of metastasis and local recurrence compared with well-differentiated liposarcoma. Both well-differentiated and dedifferentiated liposarcoma are highly associated with amplification of MDM2 and CDK4 on immunohistochemistry analysis, as was seen in this case.[1]

On histopathologic analysis, well-differentiated liposarcoma resembles mature adipose tissue but contains abnormal fibrous septations and atypical lipoblasts, whereas dedifferentiated liposarcoma usually has the appearance of high-grade spindle cell carcinoma or malignant fibrous histiocytoma or undifferentiated pleomorphic sarcoma.[1] In the case described here, initial fine needle biopsies were obtained with endoscopic ultrasound and showed a spindle cell lesion with dense fibrous tissue and mild to moderate cytologic atypia. Excisional biopsy then confirmed the diagnosis of dedifferentiated liposarcoma.

Of the previously reported cases of pancreatic liposarcoma, 5 cases were of the dedifferentiated subtype (including the current study), 4 were well-differentiated, one was pleomorphic, and one was myxoid (Table 1). The average age at diagnosis was 55 years, and most cases occurred in female patients (7/11). The average size of the lesion at the time of diagnosis was 12.6 cm. Six patients presented with abdominal pain or distension, whereas the rest of patients were asymptomatic at initial presentation. All patients underwent surgical resection, which is the mainstay of treatment. Long-term (>12 months) follow-up data were available in 6 cases, and short-term follow-up data were available in 2 cases. Of the cases with follow-up data available, only one case of recurrence was noted after 44 months.

In conclusion, primary pancreatic liposarcoma is rare with only 11 documented case reports, including this study. Surgical resection is the mainstay of treatment. Ongoing efforts should be made to report cases of pancreatic liposarcoma to increase awareness and improve knowledge of this rare diagnosis.

Table 1. Summary of the 11 reported cases of primary pancreatic liposarcoma

Study	Year	Age	Sex	Presentation	Location	Size (cm)	Subtype	Outcome
Elliott et al[5]	1980	59	F	Abdominal distension	Body	16	Pleomorphic	No recurrence (6 y)
Dodo et al[4]	2005	76	M	Abdominal pain	Body/tail	9	Well differentiated with area of dedifferentiation	No recurrence (26 mo)
Kuramoto et al[3]	2013	24	M	Abdominal distension	Body	25	Myxoid	Recurrence (44 mo)
Machado et al[9]	2016	42	M	Abdominal pain	Head	6.8	Dedifferentiated with high-grade components	No recurrence (5 y)
Matthews et al[8]	2016	65	F	Asymptomatic	Tail	4	Well differentiated	N/A
Han et al[7]	2017	29	F	Abdominal distension	Tail	20	Dedifferentiated	No recurrence (12 mo)
Cao et al[6]	2019	72	F	Asymptomatic	Body/tail	10.3	Well differentiated	N/A
Liu et al[12]	2019	28	F	Abdominal pain	Tail	28	Dedifferentiated with high-grade components	No recurrence (26 mo)
Tanabe et al[10]	2022	81	F	Asymptomatic	Tail	2.6	Dedifferentiated with well-differentiated component at the margin	N/A
Kim et al[11]	2014	78	F	Asymptomatic	Body	7.3	Atypical lipomatous tumor/ well-differentiated liposarcoma	No recurrence (2 mo)
Present case	2023	52	M	Asymptomatic	Tail	10	Dedifferentiated	No recurrence (12 mo)

M, male; F, female; N/A, not available.

DISCLOSURES

Author contributions: N. Wilson, A. Issak, and S. Mallery wrote the manuscript. All authors were involved in drafting and editing the manuscript. S. Mallery is the article guarantor.

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Informed consent was obtained for this case report.

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REFERENCES

1. Thway K. Well-differentiated liposarcoma and dedifferentiated liposarcoma: An updated review. *Semin Diagn Pathol* 2019;36(2):112–21.
2. Zhang H, Yu S, Wang W, et al. Primary mesenchymal tumors of the pancreas in a single center over 15 years. *Oncol Lett* 2016;12(5):4027–34.
3. Kuramoto K, Hashimoto D, Abe S, et al. Education and imaging. Hepatobiliary and pancreatic: Large pancreatic liposarcoma. *J Gastroenterol Hepatol* 2013;28(12):1800.
4. Dodo IM, Adamthwaite JA, Jain P, Roy A, Guillou PJ, Menon Kv. Successful outcome following resection of a pancreatic liposarcoma with solitary metastasis. *World J Gastroenterol* 2005;11(48):7684–5.
5. Elliott T, Albertazzi V, Danto L. Pancreatic liposarcoma case report with review of retroperitoneal liposarcomas. *Cancer* 1980;45(7):1720–3.
6. Cao D, Wang J, Guo L. Pancreatic liposarcoma: A rare cause of pancreatic mass in adult. *J Gastroenterol Hepatol* 2019;34(8):1275.
7. Han T, Luan Y, Xu Y, et al. Successful treatment of advanced pancreatic liposarcoma with apatinib: A case report and literature review. *Cancer Biol Ther* 2017;18(9):635–9.
8. Matthews M, Nelson S, Hari D, French S. Well differentiated liposarcoma, sclerosing type, of the pancreas a case report. *Exp Mol Pathol* 2016;101(3):320–2.
9. Machado MCC, Fonseca GM, de Meirelles LR, Zacchi FFS, Bezerra ROF. Primary liposarcoma of the pancreas: A review illustrated by findings from a recent case. *Pancreatology* 2016;16(5):715–8.
10. Tanabe M, Matsui H, Higashi M, Tokumitsu Y, Nagano H, Ito K. Pancreatic liposarcoma: A case report. *Abdom Radiol* 2022;47(6):1912–6.
11. Kim JY, Song JS, Park H, et al. Primary mesenchymal tumors of the pancreas: Single-center experience over 16 years. *Pancreas* 2014;43(6):959–68.
12. Liu Z, Fan WF, Li GC, Long J, Xu YH, Ma G. Huge primary dedifferentiated pancreatic liposarcoma mimicking carcinosarcoma in a young female: A case report. *World J Clin Cases* 2019;7(11):1344–50.
13. Dei Tos AP. Liposarcoma: New entities and evolving concepts. *Ann Diagn Pathol* 2000;4(4):252–66.