Malignant Peripheral Nerve Sheath Tumor of the Pancreas: A Case Report and Updated Review of Related Literature

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ABSTRACT

Malignant peripheral nerve sheath tumors (MPNSTs) are rare soft tissue sarcomas with poor prognosis due to their high recurrence rates. The prevalence of MPNST in the general population is 0.001%, with tumors arising from the retroperitoneum accounting for only 1% of all MPNSTs. In this report, we present a case of a 59-year-old male with pancreatic MPNST. To the authors' knowledge, this is the first documented case of pancreatic MPNST in the Philippines.

The patient initially presented with a 3-month history of abdominal pain, weight loss, and anorexia. On abdominal computed tomography (CT) scan, a large cystic mass involving the pancreatic head and body, with an enhancing peripheral solid component in the superior region was seen. The patient underwent distal pancreatectomy, en bloc splenectomy and excision of duodenal cyst. Post-operative histopathology and immunohistochemistry staining were consistent with pancreatic MPNST with tumor very near the margin of resection adjacent to the portal vein. Adjuvant systemic chemotherapy and radiotherapy were not performed due to lack of evidence of benefit over risk for this population. Disease recurrence (nodal-peritoneal metastases) was noted six months post-operatively and he was given palliative chemotherapy with single-agent doxorubicin. However, disease progression was noted after five cycles of chemotherapy. Second-line regimen was planned but the patient died of a pulmonary embolism prior to the initiation of chemotherapy.

Due to the rarity and highly aggressive nature of MPNSTs, furthering knowledge on these tumors is important, particularly in their inclusion among the differential diagnoses for pancreatic tumors. Prompt diagnosis and histopathologic confirmation by a pathologist specializing in sarcomas are crucial in the treatment planning and prognostication of these tumors. Lastly, further studies are needed to establish more effective treatments in unresectable or metastatic disease.

Keywords: malignant peripheral nerve sheath tumor, sarcoma, pancreas

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INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are rare forms of soft tissue sarcomas that are locally aggressive with high rates of metastases. The prevalence of MPNST in the general population is 0.001%, with around 50% of patients having neurofibromatosis type 1 (NF1). The most common sites of involvement include the extremities, trunk, and head and neck regions. Meanwhile, retroperitoneal MPNSTs account for only 1% of all MPNSTs. Patients with MPNSTs usually present with a history of progressively enlarging soft tissue masses that present with compressive symptoms on its surrounding structures that may cause pain or neurologic symptoms (hypoesthesia or dysesthesia).[1,2]

Treatment usually entails adequate surgical resection to achieve negative margins, with poorer survival outcomes among patients with positive margins. The role of chemotherapy in the adjuvant setting is not yet established and systemic chemotherapy is usually used in locally unresectable or metastatic disease. Reported chemotherapeutic regimens in literature include doxorubicin and ifosfamide-based regimens. Radiotherapy is another treatment option for patients with MPNSTs, however its role has not yet been shown to improve survival.[1,2]

Here we report one case of a resected malignant peripheral nerve sheath tumor arising from the pancreas that progressed six months postoperatively and was shown to be refractory to doxorubicin monotherapy. To the authors' knowledge, this is the first documented case of pancreatic MPNST in the Philippines.

CASE PRESENTATION

A 59-year-old man with good performance status presented in the outpatient department with a 3-month history of left-sided flank pain, bloating, undocumented weight loss, and anorexia. At the time, there were no bowel movement changes and jaundice. He had no comorbidities, prior surgeries or hospitalizations and maintenance medications. He had no family history of cancer, cardiovascular diseases, and neurofibromatosis, and he did not have any vices. Physical examination revealed unremarkable findings, with no grossly palpable abdominal mass and no cutaneous lesions.

An abdominal computed tomography (CT) scan with contrast enhancement was done showing splenomegaly and an abdominal cystic mass at the head and body of the pancreas, measuring 9.2 x 13.8 x 10.4 centimeters (cm) (Figure 1). An enhancing, peripheral solid component of the mass was also noted along the superior aspect, measuring 1.3 x 1.7 x 1.7 cm (Figure 2). Considerations were a pancreatic pseudocyst versus a neoplasm. Initial laboratories showed normal serum lipase and amylase levels, and a normal CA19-9 at 17 U/mL (units per milliliter) (reference value less than 37 U/mL). He was seen by a hepatobiliary surgeon who performed a distal pancreatectomy, en bloc splenectomy, and excision of duodenal cyst, done five months from the onset of symptoms. Intraoperatively, the mass was noted to be closely adherent to the portal vein.

The specimen received in surgical pathology was a tan, lobulated, unicystic mass measuring 11.5 x 6.0 x 3.5 cm, compressing the adjacent rubbery, tan, lobulated pancreatic parenchyma. Cut sections of the mass showed a 0.2 cm-thick cream grey, fibrous capsule, lined by a tan to hemorrhagic, spongy to fibrous tissue, measuring 0.2 to 0.5 cm in thickness. A 2.8 x 1.5 x 2.5 cm solid nodule was seen along the superior area, showing tan, spongy cut surfaces. The pancreatic duct was patent, compressed, and grossly uninvolved. Microscopy showed a rather well-circumscribed tumor surrounded by a fibrous stroma. All inked resection margins were noted to be free of tumor although tumor cells were seen to be very near [less than 1 millimeter (mm)] from the margin of resection near the portal vein. On low power magnification, the tumor was composed of spindle cells arranged in long fascicles interspersed with thin-walled vessels (Figure 3A) and

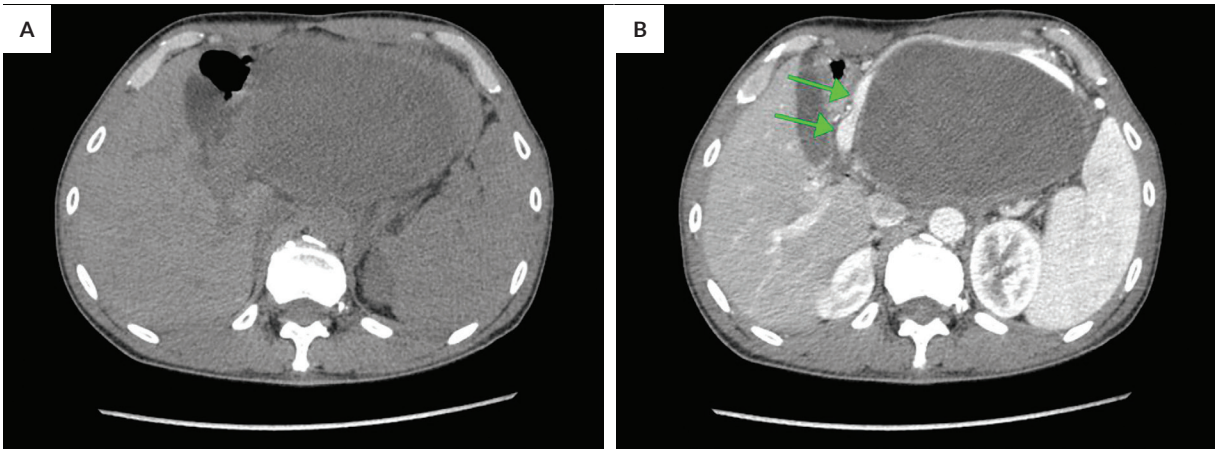

Figure 1. Plain **(A)** and contrast-enhanced **(B)** abdominal CT images showing a large cystic pancreatic mass showing compression of the portal vein *(green arrows)* with a poor plane of differentiation from the pancreatic mass.

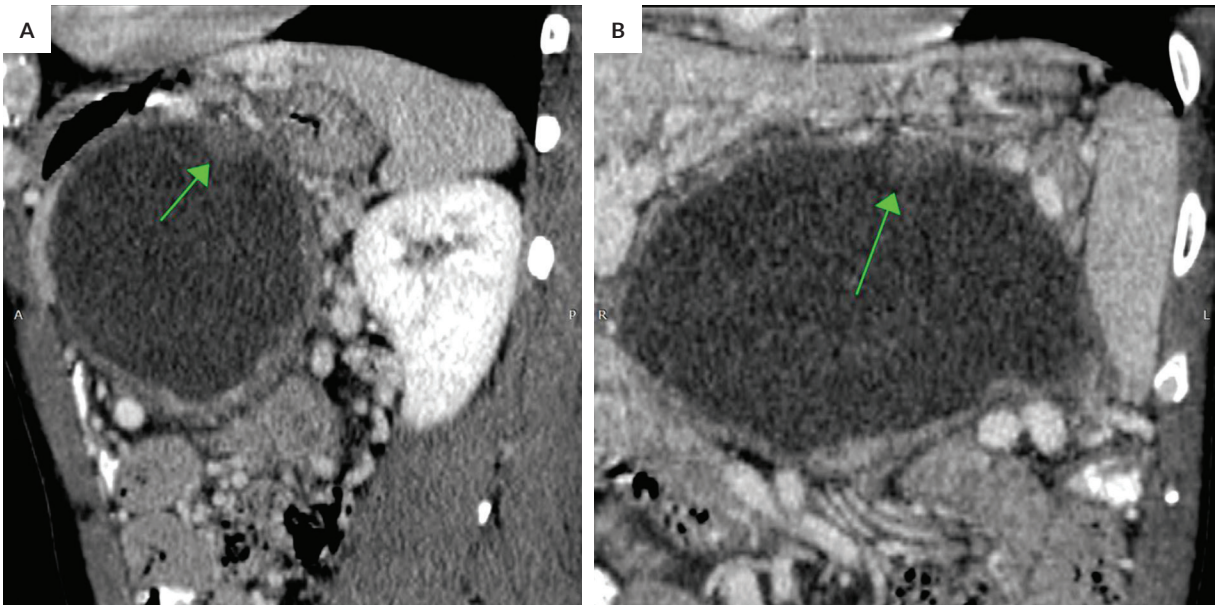

Figure 2. Sagittal **(A)** and coronal **(B)** views of the contrast-enhanced CT scan showing an enhancing peripheral solid component *(green arrows)* along the superior aspect of the pancreatic mass.

patchy areas of geographic necrosis (Figure 3B). On higher magnification, the neoplastic cells showed normochromatic to hyperchromatic and pleomorphic nuclei, inconspicuous nucleoli, with scant amphophilic cytoplasm. Mitoses could be readily seen (Figure 3C). Focal areas showing cartilaginous (not seen here) and rhabdoid differentiation, characterized by ample eosinophilic cytoplasm, were also evident (Figure 3D). A panel of immunohistochemistry stains showed that neoplastic cells were immunoreactive with S100 (Figure 3E) and myogenin (Figure 3F). Pancytokeratin, SMA, desmin, CD34, DOG1, and CD117 were all negative. The case was signed out as a malignant peripheral nerve sheath tumor with skeletal muscle and cartilaginous differentiation.

A multidisciplinary discussion among the hepatobiliary surgeon, medical oncologist, and radiation oncologist to discuss options for adjuvant therapy was done. Radiation therapy and systemic chemotherapy were not pursued due to lack of evidence for benefit in patients with retroperitoneal MPNSTs in the adjuvant setting. In addition, the potential toxicities of radiotherapy and its risk for adverse events given the retroperitoneal location were considered. Thus, the final consensus was to do close surveillance.

However, six months post-operatively, the patient reported recurrence of abdominal pain and new onset back pain. A repeat abdominal CT scan showed multiple, complex, heterogeneously enhancing masses in the gastrohepatic (7.8 x 6.9 cm), peripancreatic (4.4 x 4.0 cm and 5.2 x 6.0 cm), paraduodenal (2.1 x 2.6 cm), transverse mesocolon (1.9 x 2.6 cm), and left paraaortic regions (7.2 x 8.2 cm) (AP x W), for which the consideration were nodal-peritoneal metastases (Figures 4A and 4B). He was started on pain medications with temporary relief of symptoms, and oral nutritional support for upbuilding. He then underwent doxorubicin monotherapy at 60 mg/m^2 every three weeks for a total of five cycles (cumulative doxorubicin dose of 300 mg/m^2) which the patient tolerated well. During treatment, he developed anorexia, Common Terminology Criteria for Adverse Events (CTCAE) version 5.0 Grade 2, weight loss CTCAE Grade 2, and fatigue CTCAE Grade 1. There were no treatment interruptions and dose modifications done during treatment. Furthermore, cardiac status remained unremarkable with normal left ventricular dimension and good systolic function, and post-treatment ejection fraction (EF) of 68% from a pre-treatment EF of 66%. However, repeat imaging after chemotherapy (four months after the prior CT scans) showed progression of the nodal and peritoneal metastases, with notable increase in size of masses in the following regions: gastrohepatic (8.8 x 10 cm), peripancreatic between the portal vein and superior mesenteric artery (8.4 x 9.5 cm), and left peripancreatic region (7.0 x 8.1 cm) (Figures 4C and 4D). According to the Response Evaluation Criteria in Solid Tumors (RECIST) 1.1 criteria, the sum of the diameters of target lesions increased by 50.3% compared to baseline, is indicative of progressive disease. He was then scheduled for second-line chemotherapy with gemcitabine-docetaxel. Unfortunately, prior to the scheduled chemotherapy, the patient was hospitalized for decreased sensorium and dyspnea due to pulmonary embolism and subsequently expired.

DISCUSSION

Sarcomas are uncommon malignancies arising from the bone or soft tissue, with a worldwide incidence of approximately 1%.[3] A type of soft tissue sarcomas, malignant

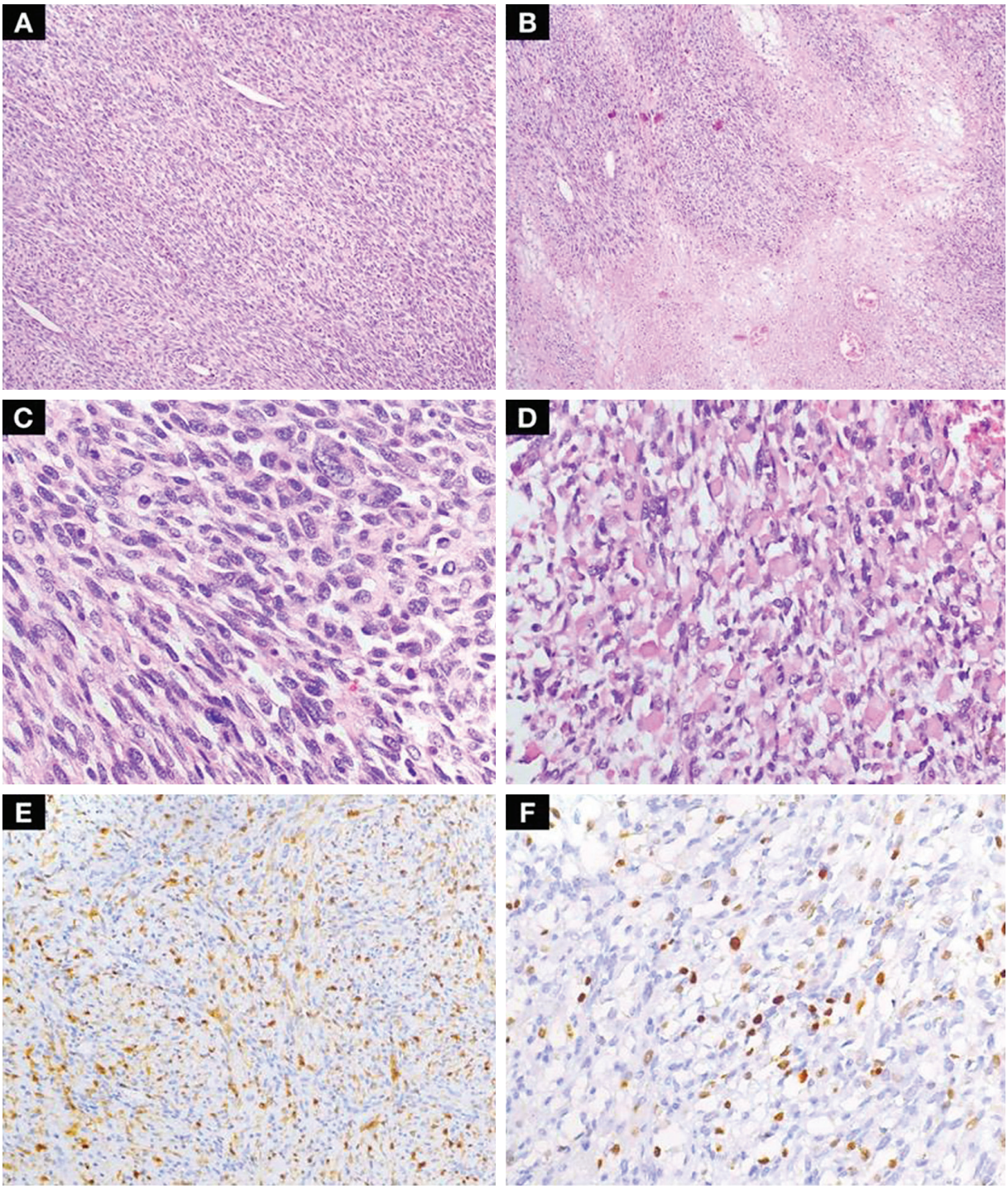

Figure 3. **(A)** Population of neoplastic spindle cells (hematoxylin and eosin, 100x) interspersed with **(B)** areas of necrosis (hematoxylin and eosin, 100x). **(C)** On higher magnification, significant atypia, pleomorphism, and brisk mitoses can be readily seen (hematoxylin and eosin, 400x). **(D)** Focally, cells showing rhabdoid differentiation was seen (hematoxylin and eosin, 400x). The neoplastic cells were immunoreactive for **(E)** S100 (200x) while the rhabdoid cells were positive for **(F)** myogenin (400x).

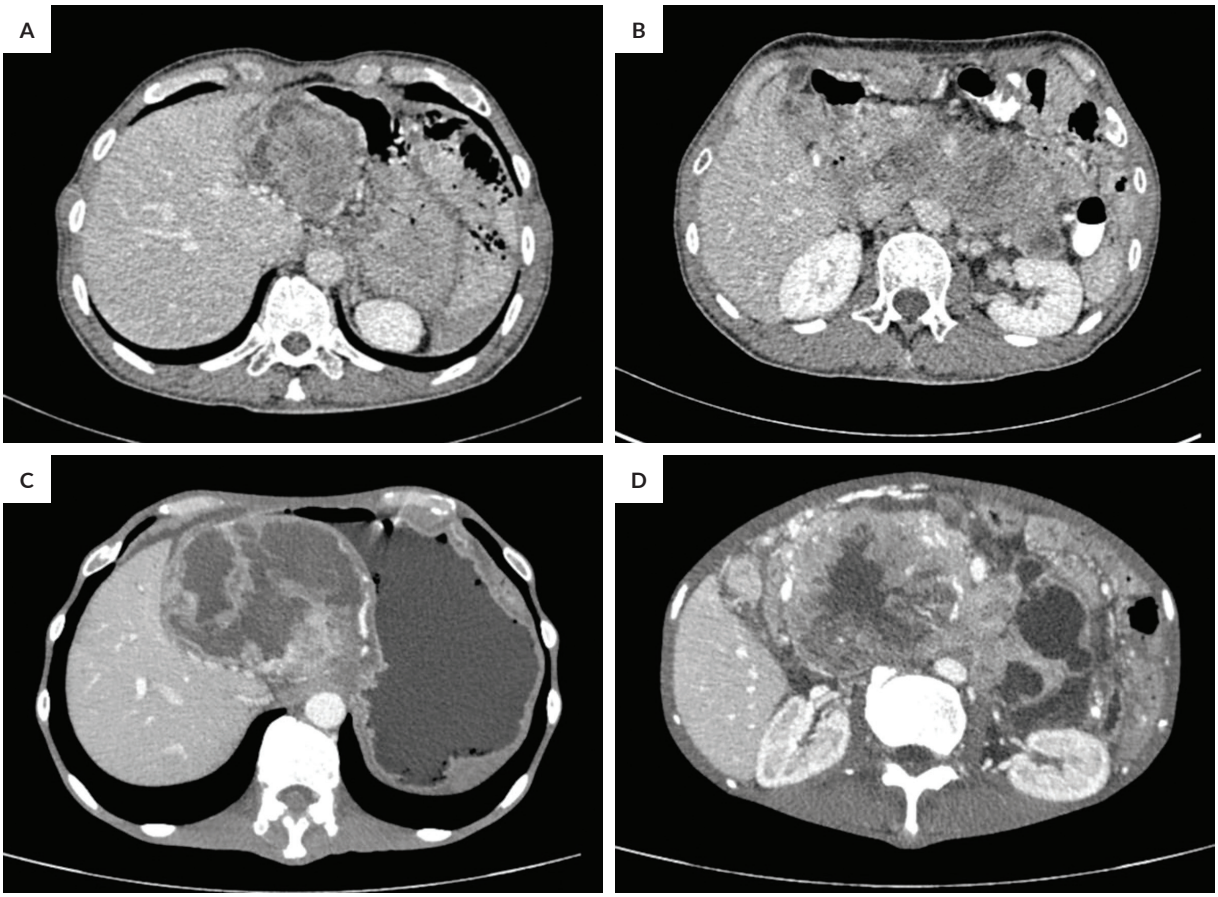

Figure 4. Contrast-enhanced abdominal CT images at the level of the gastrohepatic region and surgical bed showing: **(A,B)** Enlarged solid-cystic masses, consistent with tumor recurrence six months after surgery. **(C,D)** Increase in size of nodal-peritoneal metastases with an overall response +50.3% based on RECIST 1.1, after five cycles of doxorubicin chemotherapy, consistent with disease progression.

peripheral nerve sheath tumors (MPNSTs) belong to the subgroup of peripheral nerve sheath tumors, which are cancers that arise from the cells of the nerve sheaths that protect the peripheral nerves. MPNSTs in particular tend to metastasize, along with the other subtypes of malignant nerve sheath tumors such as melanotic malignant nerve sheath tumor, malignant granular cell tumor, and malignant perineurioma.[1,3] Although occasionally used interchangeably with the terms *malignant schwannoma* or *neurofibrosarcoma*, MPNST is the term being used according to the Fifth Edition of the World Health Organization (WHO)'s Classification of Tumors Soft Tissue and Bone Tumors, since these tumors may originate or differentiate more likely like any peripheral nerve sheath cell - not just Schwann cells. MPNSTs are extremely rare, having an incidence of only 0.001% in the general population, with increased predilection among patients diagnosed with neurofibromatosis type 1 (lifetime risk of 10%). Although the presence of MPNST is not required for the diagnosis of NF1.[1,4]

Clinically, patients may present with a history of a progressively enlarging mass, most commonly involving the head and neck, trunk and extremities. There may also be pain associated with the local effects of the mass, and occasionally, neurologic symptoms such as hypoesthesia or dysesthesia. Other symptoms are dependent on the disease site and local invasion, if any, on the surrounding structures. Imaging studies are crucial in the work up of MPNSTs to assess tumor characteristics and resectability. Some radiographic features include magnetic resonance imaging (MRI) findings of peritumoral edema, irregular and/or locally invasive margins as well as tumoral heterogeneity.[1]

Although there are radiographic features usually associated with MPNSTs, histologic confirmation is still needed for definitive diagnosis. Due to the rarity of the condition, referral to a pathologist specializing in sarcomas or slide review done in a center with a high volume of sarcoma cases is crucial. Resection is preferred where possible, but a core-needle biopsy may suffice in cases of unresectable or locally advanced diseases to identify if the tumor is malignant. Additional immunohistochemistry stains may include S100, Ki67, TP53, CD34, and p16 to differentiate the histology of the tumor more accurately.[1] In terms of

oncologic staging, the TNM (tumor, nodes, metastasis) Classification of Malignant Tumors system of the American Joint Committee on Cancer (AJCC) is usually recommended for soft tissue tumors. However, MPNST is among the malignant soft tissue tumors where pathologic staging using the AJCC system is not considered to be clinically relevant. Furthermore, the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) grading was also not used because of its unclear prognostic significance for MPNST. However, for this patient, his tumor was deemed 'high-grade' due to the presence of cells with skeletal muscle differentiation, which according to WHO is particularly aggressive compared to those without.

The management of MPNST entails a multimodality, multidisciplinary approach consisting of surgeons, medical oncologists, radiation oncologists, and pathologists, mostly due to the rarity and complexity of this disease entity. However, its treatment is mostly surgical in nature due to the tumor's poor response to chemotherapy. Wide resection of MPNST to achieve adequate margins (R0 resection) is associated with improved outcomes. MPNST from the extremities have better outcomes compared to other sites of involvement primarily due to the capacity to perform adequate resection in these areas. Surgical resection may be harder to achieve in cases of retroperitoneal MPNST due to the potential direct invasion of adjacent organs and difficulty in achieving adequate margins.[1]

Radiotherapy may be offered to patients as well, especially those with large and high-grade tumors or those with R1 or R2 resection.[1] Neoadjuvant radiotherapy among high-grade, large MPNSTs is sometimes considered as it can decrease radiation fields and doses, resulting in lower toxicities, as opposed to adjuvant radiotherapy.[4] Some studies suggest improved local recurrence rates, but the role of radiotherapy in improving overall survival has yet to be established.[4,5] For this patient, there was no preoperative histopathologic diagnosis of MPNST hence neoadjuvant radiotherapy was not considered, and outright resection was pursued. Meanwhile, adjuvant radiotherapy was not done as well due to the lack of definitive evidence for benefit and potential for toxicities. Hence, close surveillance was recommended.

In terms of chemotherapeutic options for MPNST, there is a paucity of Phase III randomized controlled trials due to the rarity of the condition. However, some patients with MPNST are included in larger sarcoma trials where possible response to specific chemotherapeutic agents may be extrapolated based on the results of the overall population.[6-8] However, in the absence of clearer evidence on the effects of certain systemic chemotherapies on MPNST specifically, it may be difficult to formulate definitive conclusions from these trials. Some retrospective cohort studies looked at the chemotherapeutic agents given for patients with MPNST, in the neoadjuvant, adjuvant and metastatic setting.[9-11] In general, systemic chemotherapy is offered in patients with locally advanced and unresectable tumors (more than 5 cm) or metastatic disease. Regimens varied across the literature, including doxorubicin, dacarbazine, ifosfamide or etoposide containing regimens. However, the response to these agents is still poor.[6-8,10-12] Retrospective data from a sarcoma referral center in Poland showed a median progression free survival of 3.9 months (95% CI 2.5-5.4) among patients given chemotherapy in the first-line (metastatic) setting, with reported lower risk of progression when given doxorubicin chemotherapy in combination with ifosfamide (HR 0.30; 95% CI 0.09 to 0.96, p=0.042).[12] An exploratory analysis of the same study showed gemcitabine based-regimens to have a significantly worse prognosis compared to doxorubicin-based regimens (HR 1.74, 95% CI 1.14-2.66, p=0.01). However, this study only had 11.3% of its patients with primary tumors at the retroperitoneal area. There was no data provided on the specific treatment interventions and response in patients with retroperitoneal (or pancreatic) MPNSTs.[12] Upregulation of both deoxyribonucleic acid (DNA) repair mechanisms and drug efflux transporters have been postulated to be contributors to the chemo-resistant nature of MPNSTs. Molecular profiling identified mutations in the (mitogen-activated protein kinase) MAPK pathway, loss of tumor suppressor genes (*TP53* and cyclin-dependent kinase inhibitor 2A or *CDKN2A*), and alterations in polycomb repressive complex 2 PRC2. However, the role of targeted therapies in MPNST has yet to be established.[13]

The prognosis of MPNSTs in general remains to be poor, with 10-year disease specific survival at 31.6%, and 7.5% for patients with metastases. Poor prognostic factors include large tumors (≥10 cm) at diagnosis, high-grade lesions, incompletely resected tumors, and presence of distant metastases.[14,15] Unfortunately for the case presented, he had a large, high-grade tumor that was incompletely resected, which eventually metastasized, indicating a poorer prognosis.

A review of literature through April 2024 was done to identify publications reporting adult patients with MPNST arising from the pancreas. A comprehensive search was performed using PubMed, Medline and Google Scholar using the terms "malignant peripheral nerve sheath tumor," "pancreas," and "pancreatic schwannoma." Two authors performed independent literature search and reviews, and compiled a comprehensive list of available studies. Reference lists of included studies and other relevant publications were also hand-searched for additional studies that may be included. Only studies in the English language were included.

Data on the clinicopathological features (including age, gender, symptoms, tumor location, tumor size, radiographic characteristics and extent of tumor involvement, nodal status, distant metastases), treatment rendered (surgery, chemotherapy, radiotherapy, immunotherapy), and outcomes (postoperative complications, local or distant recurrence, overall survival) were collected.

Nine case reports of MPNST arising from the pancreas were retrieved after literature search.[16-24] The mean age of documented cases is 56 years old (SD 16), with an equal

number of males and females. The most common presenting symptom is abdominal pain, which was present in all documented cases. Most of the tumors were located at the body of the pancreas, with a mean tumor size (largest dimension) of 11.9 cm (SD 4.9 cm). Half of the cases were reported to have tumors that directly invaded the surrounding structures such as the duodenum, stomach and colon.[18,19,22,23] Nodal involvement was reported in only one other case, and only one other case report showed distant metastases (cutaneous metastases at the right posterior thigh) after initial resection of the tumor.[22,24] All cases underwent resection of the primary pancreatic tumor with only two other cases undergoing systematic lymph node dissection.[16-24] One patient was reported to have undergone palliative radiotherapy on the cutaneous metastases of the thigh, followed by nivolumab. However, after initial response of the tumor, there were noted new cutaneous lesions outside the RT field and the patient eventually expired.[24] There were no published cases of pancreatic MPNST that underwent systemic chemotherapy. The clinicopathologic features, treatment modalities done, and outcomes of reported cases are summarized in the Appendix.

CONCLUSION

Accurate histopathologic confirmation by a pathologist specializing in sarcomas is crucial in the treatment planning and prognostication of patients with MPNST. Surgery with adequate margins remains a key treatment modality that is associated with improved survival rates. To date, the role of chemotherapy and radiotherapy in improving patient outcomes has not yet been established.

Despite being a rare occurrence, improving knowledge on MPNSTs is important, particularly in their inclusion among the differential diagnoses for pancreatic tumors. In addition, more studies are needed to establish effective treatments in unresectable or metastatic disease.

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Statement of Authorship

All authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

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REFERENCES

1. Knight SWE, Knight TE, Santiago T, Murphy AJ, Abdelhafeez AH. Malignant peripheral nerve sheath tumors-a comprehensive review of pathophysiology, diagnosis, and multidisciplinary management. Children (Basel). 2022 Jan;9(1):38. doi: 10.3390/children9010038. PMID: 35053663. PMCID: PMC8774267.
2. Yao C, Zhou H, Dong Y, Alhaskawi A, Ezzi S, Wang Z, et al. Malignant peripheral nerve sheath tumors: latest concepts in disease pathogenesis and clinical management. Cancers (Basel). 2023 Feb; 15(4):1077. doi: 10.3390/cancers15041077. PMID: 36831419. PMCID: PMC9954030.
3. Sbaraglia M, Bellan E, Dei Tos AP. The 2020 WHO Classification of Soft Tissue Tumours: news and perspectives. Pathologica. 2021 Apr;113(2):70-84. doi: 10.32074/1591-951X-213. PMID: 33179614. PMCID: PMC8167394.
4. Martin E, Coert JH, Flucke UE, Sloof WBM, Yo VKY, van der Graaf WT, et al. A nationwide cohort study on treatment and survival in patients with malignant peripheral nerve sheath tumours. Eur J Cancer. 2020 Jan;124:77-87. doi: 10.1016/j.ejca.2019.10.014. PMID: 31760312.
5. Kahn J, Gillespie A, Tsokos M, Ondos J, Dombi E, Camphausen K, et al. Radiation therapy in management of sporadic and neurofibromatosis type 1-associated malignant peripheral nerve sheath tumors. Front Oncol. 2014 Nov;4:324. doi: 10.3389/fonc.2014.00324. PMID: 25452937. PMCID: PMC4233912.
6. Spunt SL, Million L, Chi YY, Anderson J, Tian J, Hibbitts E, et al. A risk-based treatment strategy for non-rhabdomyosarcoma soft-tissue sarcomas in patients younger than 30 years (ARST0332): a Children's Oncology Group prospective study. Lancet Oncol. 2020 Jan;21(1): 145-61. doi: 10.1016/S1470-2045(19)30672-2. PMID: 31786124. PMCID: PMC6946838.
7. Gronchi A, Ferrari S, Quagliuolo V, Broto JM, Pousa AL, Grignani G, et al. Histotype-tailored neoadjuvant chemotherapy versus standard chemotherapy in patients with high-risk soft-tissue sarcomas (ISG-STS 1001): an international, open-label, randomised, controlled, phase 3, multicentre trial [published correction appears in Lancet Oncol. 2017 Jun;18(6):e301. doi: 10.1016/S1470-2045(17)30334-0. PMID: 28499583.
8. Gronchi A, Stacchiotti S, Verderio P, Ferrari S, Broto JM, Pousa AL, et al. Short, full-dose adjuvant chemotherapy (CT) in high-risk adult soft tissue sarcomas (STS): long-term follow-up of a randomized clinical trial from the Italian Sarcoma Group and the Spanish Sarcoma Group. Ann Oncol. 2016 Dec;27(12):2283-8. doi: 10.1093/annonc/mdw430. PMID: 27733375.
9. Chou D, Bilsky MH, Luzzati A, Fisher CG, Gokaslan ZL, Rhines LD, et al. Malignant peripheral nerve sheath tumors of the spine: results of surgical management from a multicenter study. J Neurosurg Spine. 2017 Mar;26(3):291-8. doi: 10.3171/2016.8.SPINE151548. PMID: 27834629.
10. Hirbe AC, Cosper PF, Dahiya S, Van Tine BA. Neoadjuvant ifosfamide and epirubicin in the treatment of malignant peripheral nerve sheath tumors. Sarcoma. 2017 May;2017:3761292. doi: 10.1155/2017/3761292. PMID: 28546782. PMCID: PMC5435903.
11. Kroep JR, Ouali M, Gelderblom H, Le Cesne A, Dekker TJA, Glabbeke MV, et al. First-line chemotherapy for malignant peripheral nerve sheath tumor (MPNST) versus other histological soft tissue sarcoma subtypes and as a prognostic factor for MPNST: an EORTC soft tissue and bone sarcoma group study. Ann Oncol. 2011 Jan;22(1):207-14. doi: 10.1093/annonc/mdq338. PMID: 20656792. PMCID: PMC3003614.
12. Sobczuk P, Teterycz P, Czarnecka AM, Świtaj T, Kosela-Paterczyk H, Kozak K, et al. Systemic treatment for advanced and metastatic malignant peripheral nerve sheath tumors-a sarcoma reference center experience. J Clin Med. 2020 Sep;9(10):3157. doi: 10.3390/jcm9103157. PMID: 33003503. PMCID: PMC7601777.

13. Peacock JD, Cherba D, Kampfschulte K, Smith MK, Monks NR, Webb CP, et al. Molecular-guided therapy predictions reveal drug resistance phenotypes and treatment alternatives in malignant peripheral nerve sheath tumors. J Transl Med. 2013 Sep;11:213. doi: 10.1186/1479-5876-11-213. PMID: 24040940. PMCID: PMC3848568.
14. Zou C, Smith KD, Liu J, Lahat G, Myers S, Wang W, et al. Clinical, pathological, and molecular variables predictive of malignant peripheral nerve sheath tumor outcome. Ann Surg. 2009 Jun;249(6): 1014-22. doi: 10.1097/SLA.0b013e3181a77e9a. PMID 19474676.
15. Valentin T, Le Cesne A, Ray-Coquard I, Italiano A, Decanter G, Bompas E, et al. Management and prognosis of malignant peripheral nerve sheath tumors: The experience of the French Sarcoma Group (GSF-GETO). Eur J Cancer. 2016 Mar;56:77-84. doi: 10.1016/j.ejca.2015.12.015. PMID: 26824706.
16. Móller Pedersen V, Hede A, Graem N. A solitary malignant schwannoma mimicking a pancreatic pseudocyst. A case report. Acta Chir Scand. 1982;148(8):697-8. PMID: 7170905.
17. Eggermont A, Vuzevski V, Huisman M, De Jong K, Jeekel J. Solitary malignant schwannoma of the pancreas: report of a case and ultrastructural examination. J Surg Oncol. 1987 Sep;36(1):21-5. doi: 10.1002/jso.2930360106. PMID: 3626558.
18. Walsh MM, Brandspigel K. Gastrointestinal bleeding due to pancreatic schwannoma complicating von Recklinghausen's disease. Gastroenterology. 1989 Dec;97(6):1550-1. doi: 10.1016/0016-5085(89)90402-2. PMID: 2511055.
19. Coombs RJ. Case of the season. Malignant neurogenic tumor of duodenum and pancreas. Semin Roentgenol. 1990 Apr;25(2):127-9. doi: 10.1016/0037-198x(90)90045-6. PMID: 2112269.
20. Hirose T, Maeda T, Furuya K, Kiyasu Y, Kawasaki H. Malignant peripheral nerve sheath tumor of the pancreas with perineurial cell differentiation. Ultrastruct Pathol. 1998 May;22(3):227-31. doi: 10.3109/01913129809033473. PMID: 9792302.
21. Titus AS, Pinckard NB, Cook WJ, Vickers SM. Solitary malignant peripheral nerve sheath tumor of the pancreas. Hepatogastroenterology. 1999 Jan;46(25):514-7. PMID: 10228853.
22. Stojanovic MP, Radojkovic M, Jeremic LM, Zlatic AV, Stanojevic GZ, Jovanovic MA, et al. Malignant schwannoma of the pancreas involving transversal colon treated with en-bloc resection. World J Gastroenterol. 2010 Jan;16(1):119-22. doi: 10.3748/wjg.v16.i1.119. PMID: 20039458. PMCID: PMC2799907.
23. Balineni P, Arcot R, Devygounder K, Rahaman K, Narayansamy B, Prabhu M, et al. Malignant peripheral nerve sheath tumor of the pancreas-A case report. Int J Surg Case Rep. 2019 Feb;55: 239-42. doi: 10.1016/j.ijscr.2019.02.011. PMID: 30785006. PMCID: PMC6383168.
24. Yanofsky VR, Ng E, Ratner D. Cutaneous metastasis of malignant peripheral nerve sheath tumor arising from the pancreas: a rarity. Dermatol Surg. 2019 Aug;45(8):1107-9. doi: 10.1097/DSS.0000000000001812. PMID: 30720497.

APPENDIX

Table 1. Summary of documented cases of pancreatic MPNST, including the presented case, with their clinicopathologic features and outcomes

Author (Year)	Age/ Sex	Symptoms	Tumor				Nodal Involvement	Presence of distant metastases	Treatment given	Treatment Outcome	Follow up Period (Months)
			Location	Size (cm)	Radiologic characteristics	Extent of tumor/ Local invasion					
Móller (1982)	60M	Abdominal pain Weight loss	Body + Tail	20	NR	NR	NR	NR	Surgery	No recurrence	4
Eggermont (1987)	40F	Abdominal pain Jaundice Acholic stools Weight loss	Head	10	Large solid tumor with central necrosis	No invasion	None	None	Surgery (PD)	Uneventful postoperative course No recurrence	9
Walsh (1989)	35F	Abdominal pain Melena Anemia	Head	NR	NR	Invasion of duodenum	NR	None	Surgery (PD)	Uneventful postoperative course No recurrence	24
Coombs (1990)	74F	Melena Anemia	Head	7	Solid with necrotic center	Invasion of duodenum	NR	None	Surgery (PD)	NR	NR
Hirose (1998)	76F	Abdominal pain Abdominal mass	Body	4.5	Well-circumscribed, enhancing mass	No invasion	None	None	Surgery (Distal pancreatectomy, splenectomy, LND)	No recurrence	13
Titus (1999)	72M	Weight loss Anorexia	Body	12	Mid-pancreatic mass	NR	NR	NR	Surgery	NR	NR

Table 1. Summary of documented cases of pancreatic MPNST, including the presented case, with their clinicopathologic features and outcomes *(continued)*

Author (Year)	Age/ Sex	Symptoms	Tumor				Nodal Involvement	Presence of distant metastases	Treatment given	Treatment Outcome	Follow up Period (Months)
			Location	Size (cm)	Radiologic characteristics	Extent of tumor/ Local invasion					
Stojanovic (2010)	24F	Abdominal pain Dyspepsia Weight loss Abdominal mass	Body + Tail	18	Well-defined round, hypodense mass	Invasion of stomach and transverse colon	Present	None	Surgery (hemi-pancreatectomy, splenectomy, omentectomy and transverse colon resection, LND)	Uneventful postoperative course	28
Balineni (2019)	62M	Abdominal pain Anorexia Vomiting	Body + Tail	10	Heterogeneously enhancing mass with internal necrosis and calcifications	Abutting the stomach, transverse colon, left adrenal and splenic hilum	None	None	Surgery (distal pancreatectomy and splenectomy)	Uneventful postoperative course	0.5
Yanofsky (2019)	60M	Erythematous nodule, right posterior thigh	(Resected)	N/A	N/A	N/A	None	Present (cutaneous)	Palliative radiotherapy, followed by nivolumab	Initial shrinkage of tumor on right thigh but noted disease recurrence outside the RT field and eventual mortality	NR
Case Report (2024)	59M	Abdominal pain Weight loss Anorexia	Head + Body	13.8	Large cystic mass with enhancing peripheral solid component	Invasion of the portal vein	Present[1]	Peritoneal	Surgery (distal pancreatectomy, splenectomy, LND) followed by chemotherapy[2]	Refractory to first-line chemotherapy and eventual mortality	12

1 – Harvested nodes during surgery were initially negative for tumor, but eventually noted nodal involvement upon disease progression

2 – Doxorubicin monotherapy

LND – Lymph Node Dissection, NR – Not reported, PD – Pancreaticoduodenectomy, RT – Radiotherapy