Pancreatic Plasmacytoma Presenting as Jaundice in Patient with Relapsed Multiple Myeloma

Triston Berger, MD[1]; Jon Kandiah, MD[2]; Naveen Anand, MD[2]

1. Department of Internal Medicine, Norwalk Hospital, affiliated with Yale School of Medicine
2. Division of Gastroenterology and Hepatology, Norwalk Hospital, affiliated with Yale School of Medicine

Introduction

- Multiple Myeloma is a neoplastic proliferation of plasma cells producing a monoclonal immunoglobulin.
- Extramedullary Plasmacytomas (EP) are plasma cell tumors developing outside of the bone marrow.
- EPs are seen in 7% of multiple myeloma patients at diagnosis, and in an additional 6% with disease progression.
- Extramedullary disease (EMD) is difficult to treat and associated with adverse prognosis.
- EMD often effects the chest wall, liver, lymph nodes, skin/soft tissues, and paraspinal area.
- The pancreas is among the least common areas for EMD. It is often diagnosed postmortem due to the indolent nature and is generally asymptomatic.

Case

65 year old Female with a history of hypertension, anxiety, and hypothyroidism initially presented to emergency department with back pain. Labs reveled anemia, renal insufficiency, hypercalcemia. Further evaluation showed a free kappa/lambda ration of <0.01. A subsequent bone marrow biopsy demonstrated sheets of large, atypical plasma cells compatible with multiple myeloma.

The patient had 6 cycles of chemotherapy with repeat biopsy showing 5-10% residual plasma cells. An auto-stem cell transplant was performed with minimal residual neoplasm seen.

A few months later, she presented again with rib pain. Imaging showed extensive multifocal osseous disease. She was started on chemotherapy for refractory multiple myeloma with combination daratumumab, lenalidomide, bortezomib, and dexamethasone. She subsequently developed jaundice which was initially thought to be secondary to drug induced liver injury, however, imaging showed dilation of CBD to 1.5cm and she presented to hospital for further evaluation.

Physical
T: 36.8 °C **HR:** 74 bpm **RR:** 18 br/min **BP:** 102/80 mmHg **SpO2:** 97%
General: Elderly Caucasian female lying on a stretcher, in no acute distress
Head: NC/AT
Eyes: Scleral icterus
Mouth: Sublingual jaundice noted
Heart: RRR, no m/r/g
Lungs: CTABL
Abdomen: +BS x4, soft, nondistended, mild tenderness over RUQ
Neuro: no focal neurologic deficits appreciated
Psych: appropriate mood and affect
MSK: No spinal tenderness, mild tenderness over right lower rib cage
Skin: No Jaundice

Imaging:
MRCP Abdomen: 3.1 x 3.0 cm pancreatic head mass with focal compression and intrahepatic and extrahepatic biliary ductal dilation. Normal caliber main pancreatic duct. Common bile duct dilated up to 1.3 cm, with abrupt tapering at the level of the pancreatic head. Enhancing liver masses and widespread bone and soft tissue masses also noted.

Labs:

	8.1	
7.3		83
	24.7	

138	101	16	133
4.2	20	1.1	

ALP 270 U/L, GGT 504
AST 44U/L
ALT 64U/L
Total Bilirubin5.6 mg/dL (4.6 Direct)

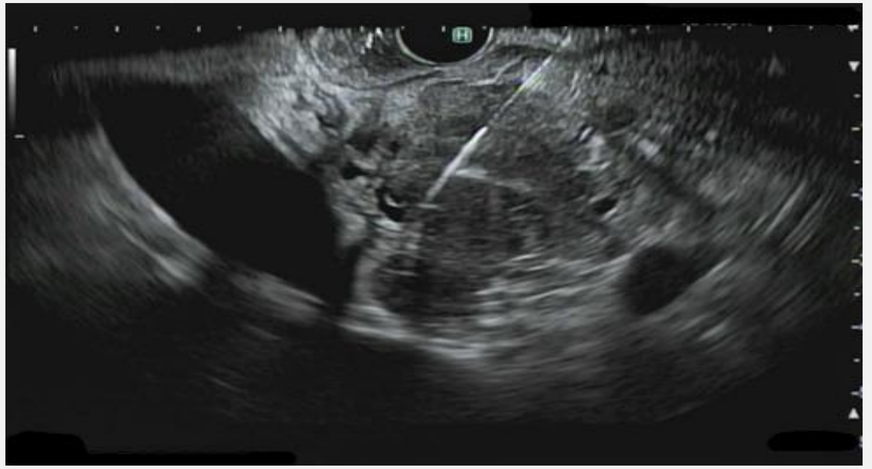

Figure 1. Endosonographic image of a fine-needle biopsy targeting a hypoechoic and well-defined lesion within the pancreatic head.

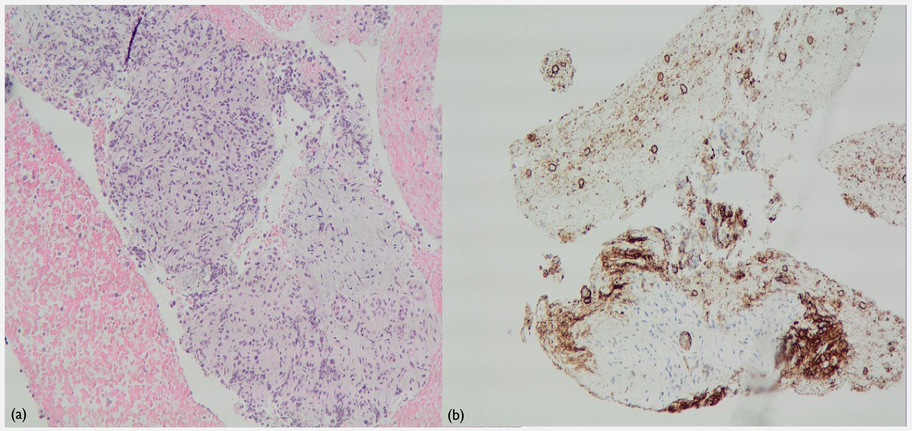

Figure 3. Histology of pancreatic biopsy under 100x magnification. (a) Hematoxylin and eosin stain demonstrating cells with eccentric nuclei, prominent nucleoli, and increased mitotic figures. (b) CD138 stain highly specific for plasma cells (blue) demonstrating increased mitotic figures.

Diagnostics

- Endoscopic ultrasound and endoscopic retrograde cholangiopancreatography performed to investigate ductal dilation
  - EUS: Hypoechoic, heterogenous mass measuring 32x25mm with well defined borders and an intact interface between surrounding vasculature suggesting lack of invasion. The mass was biopsied using fine needle aspiration (Figure 1). A round mass was also seen in the left lobe of the liver measuring 30x28mm. There was no lymphadenopathy or pancreatic ductal abnormality.
  - ERCP: Intrahepatic and extrahepatic ductal dilation (16mm) with smooth tapering distally along 20mm long area of stenosis (Figure 2). No filling defects were identified. A 10mmx6cm covered metal stent was placed with good bile flow.
- Biopsies demonstrated sheets of highly malignant cells with plasmablastic features, consistent with pancreatic plasmacytoma (Figure 3).

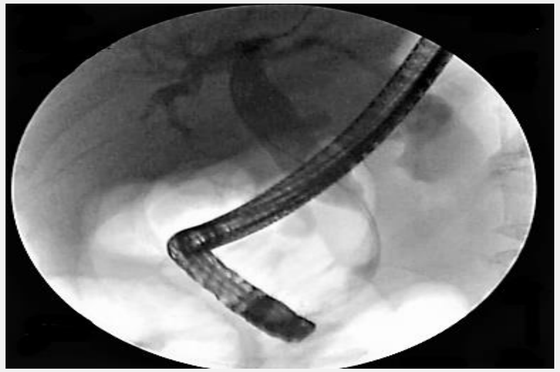

Figure 2. Fluoroscopic image taken during endoscopic retrograde cholangiopancreatography demonstrating a biliary stricture with upstream biliary ductal dilatation secondary to extrinsic compression from pancreatic mass.

Discussion

- The development of EMD in multiple myeloma is uncommon and associated with poor prognosis.
- The patient in our case developed multiple EPs including paraspinal, dermal, hepatic, and pancreatic.
- The pancreas is one of the least common areas of EMD development. It typically does not present until later in the disease course due it's indolent nature, and is often diagnosed postmortem. When it does present, obstructive jaundice is common.
- Computerized tomography showed common bile duct dilation but did not reveal a pancreatic abnormality prior to hospital admission. MRCP should be obtained for thorough evaluation.
- Plasmacytoma can appear similar to other primary diseases on imaging, such as adenocarcinoma, therefore investigation with EUS-FNA plays a vital role in diagnosis.
- The patient was subsequently started on therapy for debulking. She continues to receive therapy for multiple myeloma and has not had any further hepato-biliary complications.

Contact

Triston Berger, MD
Nuvance Health
Email: triston.berger@nuvancehealth.org

References

1. Hirata, S., Yamaguchi, K., Bandai, S., Izumo, A., Chijiiwa, K., & Tanaka, M. (2002). Secondary extramedullary plasmacytoma involving the pancreas. Journal of hepato-biliary-pancreatic surgery, 9(1), 111–115. https://doi.org/10.1007/s005340200012
2. Wang, Xin MDa; Xie, Huaiya MDb; Zhang, Lu MDc,*. Multiple myeloma with onset of pancreas involvement: A case report. Medicine: July 2019 - Volume 98 - Issue 30 - p e16567 doi: 10.1097/MD.0000000000016567
3. Rawstron, A. C., Davies, F. E., DasGupta, R., Ashcroft, A. J., Patmore, R., Drayson, M. T., Owen, R. G., Jack, A. S., Child, J. A., & Morgan, G. J. (2002). Flow cytometric disease monitoring in multiple myeloma: the relationship between normal and neoplastic plasma cells predicts outcome after transplantation. Blood, 100(9), 3095–3100. https://doi-org.ezproxy.uvm.edu/10.1182/blood-2001-12-0297
4. Abu-Hammour, A. M., Venu, R. P., Etzkorn, K. P., Showel, J. L., Zaytsev, P. M., & Brown, R. D. (1996). Common bile duct obstruction caused by multiple myeloma of the pancreas. Gastrointestinal Endoscopy, 44(5), 606–608. https://doi.org/10.1016/s0016-5107(96)70018-4
5. Castellani, L., Burgésser, M. V., Guanchiale, L., Benavídez, A., de Diller, A. B., Basquiera, A. L., & Balderramo, D. (2014). Pancreatic plasmacytoma with biliary obstruction as a manifestation of multiple myeloma relapse. Gastroenterología y Hepatología, 37(6), 357–359. https://doi.org/10.1016/j.gastrohep.2013.10.004
6. Lu, T., Pu, H., & Zhao, G. (2017). Primary pancreatic plasmacytoma: a rare case report. BMC gastroenterology, 17(1), 167. https://doi.org/10.1186/s12876-017-0729-z
7. Paydas, S. (2019). Pancreatic plasmacytoma: A rare but important entity for gastroenterologists, oncologists and hematologists. Journal of Oncological Sciences, 5(3), 109–111. https://doi.org/10.1016/j.jons.2019.10.002