A case report on pancreatic plasmacytoma presenting as obstructive jaundice

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ABSTRACT

Background: Extramedullary solitary plasmacytomas (EMD) are an uncommon manifestation of plasma cell disorders. The median survival rate is less than 6 months. EMD rarely involves the digestive tract, the liver is the most common site with only a 2.3% incidence rate for pancreatic involvement.

Case Presentation: We report a case of a patient with a history of multiple myeloma who presented with painless obstructive jaundice and EUS findings of hypoechoic, solid irregular mass in the pancreatic head with surrounding peripancreatic lymphadenopathy. Fine needle aspirate (FNA) confirmed malignant infiltration of plasma cell neoplasm. The patient rapidly deteriorated and succumbed to his illness within 6 weeks of diagnosis.

Conclusion: Extramedullary plasmacytomas (EMD) involving the pancreas are uncommon and associated with a poor prognosis. Our case highlights the importance of considering extramedullary plasmacytoma when a patient with MM presents with a pancreatic mass, particularly one showing a rapid interval growth. This rate of growth is not seen with primary pancreatic parenchymal diseases such as cancer, cysts, and so on. This case highlights the importance of considering rare differentials while evaluating patients with pancreatic mass.

Keywords: Rare and lethal: a case report on the grim reality of EMD infiltrating the pancreas. Despite its rarity, timely recognition is crucial for improved outcomes in patients with multiple myeloma. #MedTwitter #Oncology #CaseReport"

Case report, extramedullary plasmacytoma, pancreatic plasmacytoma, multiple myeloma, pancreatic mass.

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Background

Extramedullary solitary plasmacytomas are an uncommon presentation of all plasma cell disorders [1]. It is associated with a poor prognosis with a median survival of fewer than 6 months [2]. Although rare to involve the digestive tract, the liver is the most common site [3]. Rarely, extramedullary pancreatic involvement is reported with autopsy studies demonstrating a 2.3% incidence rate [4].

Although pancreatic EMD is rare, it is important to maintain clinical suspicion in patients with plasma cell disorders as this may indicate disease relapse. We describe a patient with a history of multiple myeloma who relapsed with pancreatic extramedullary plasmacytoma.

Case Report

An 84-year-old male with a significant history of multiple myeloma and extramedullary cutaneous plasmacytomas presented with a 3-week onset of acholic stool, pruritus, and jaundice. Labs revealed a direct hyperbilirubinemia with a mixed hepatocellular and cholestatic pattern of liver injury. Tumor markers including CA-19-9, CA-125, CEA, and AFP were within normal range. Contrast-enhanced abdominopelvic computed tomography (CT) reported a 3.7 cm hypo-enhancing pancreatic head mass with biliary and pancreatic duct obstruction associated with celiac and superior mesenteric artery axis lymphadenopathy (Figure 1). Endoscopic ultrasound (EUS) revealed an irregular 33 mm × 37.4 mm hypoechoic, solid irregular mass in the pancreatic head with surrounding peripancreatic lymphadenopathy (Figure 2). Fine needle aspirate (FNA) revealed a CD138+ plasma cell-rich smear with positive Lamba *in-situ* hybridization stain (ISH) and negative Kappa ISH confirming a malignant plasma cell neoplasm (Figure 3). On endoscopic retrograde cholangiopancreatography (ERCP), the common bile duct was dilated to 15 mm in diameter with a 20 mm stricture within the intrapancreatic portion at the pancreatic head (Figure 2). Sphincterotomy was performed and a 10 mm × 8 cm covered metal stent

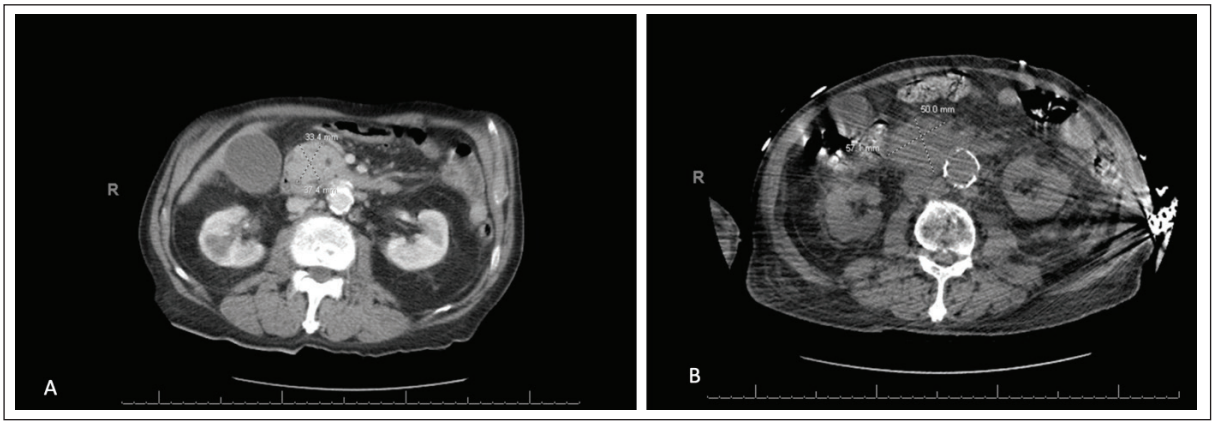

Figure 1. A - Contrast-enhanced abdominopelvic CT demonstrating a 33.4 mm × 37.4 mm hypo-enhancing pancreatic head mass. B - Contrast-enhanced abdominopelvic CT 2 weeks later demonstrating significant interval growth of hypo-enhancing pancreatic head mass to 50.0 mm × 57.1 mm.

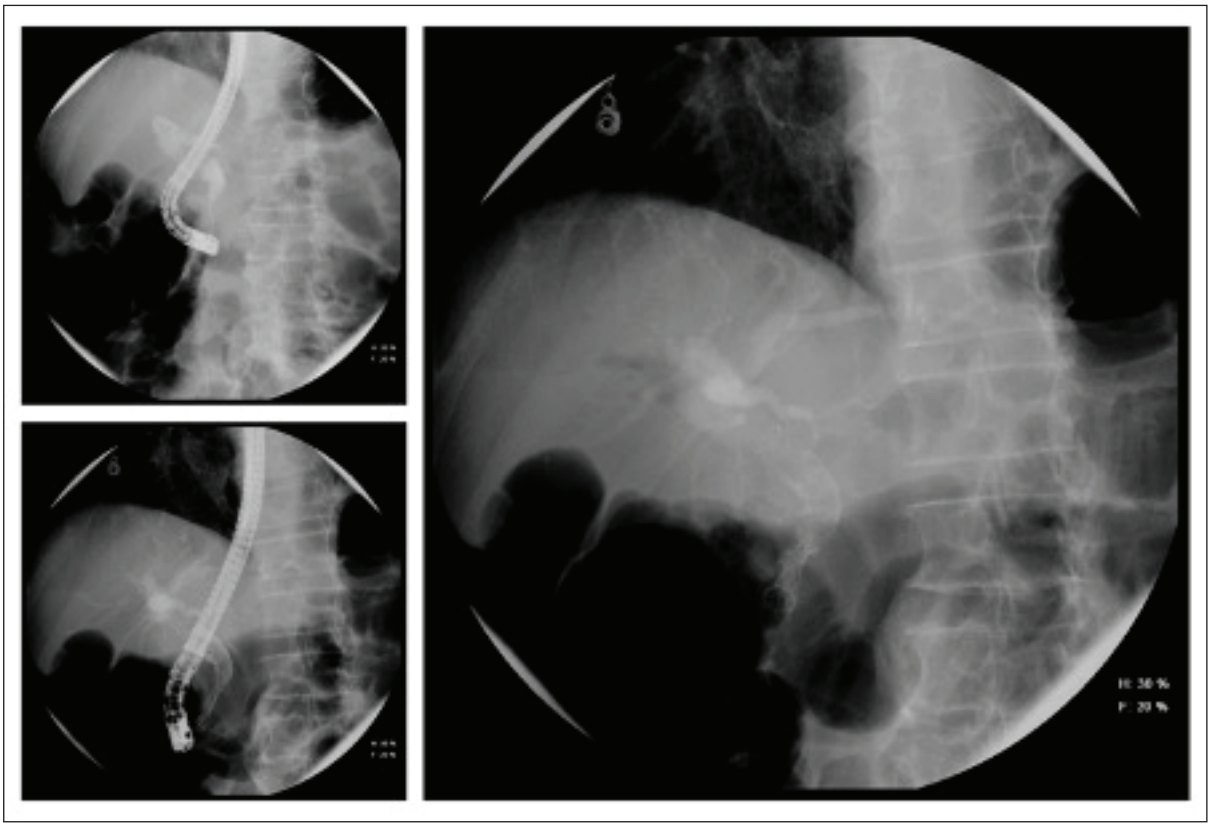

Figure 2. *ERCP revealed. (A) a dilated common bile duct to 15 mm in diameter with (B) diffuse intrahepatic biliary duct dilatation. (C) A 20 mm stricture within the intrapancreatic portion at the pancreatic head (yellow arrow). Sphincterotomy was performed and a 10 mm × 8 cm covered metal stent was placed.*

was placed. The jaundice and pruritus resolved. A repeat CT in 4 weeks shows a significant increase in the size of pancreatic head mass to 50 mm × 57 mm. The patient rapidly deteriorated and succumbed to his illness within 6 weeks of diagnosis.

Discussion

In 1947, Hefferman described the first report of pancreatic plasmacytoma presenting as acute intestinal obstruction [5]. Pancreatic involvement may present as an isolated plasmacytoma or in association with underlying multiple myeloma, which can either precede a diagnosis of multiple myeloma or as a manifestation of disease recurrence [6].

Pancreatic plasmacytomas have a 3–5-fold greater male preponderance with a mean age of onset of 55–58.5 years [7]. Typically, focal involvement of the head of the pancreas is noted although cases of diffuse involvement have been reported [8]. Patients commonly present with

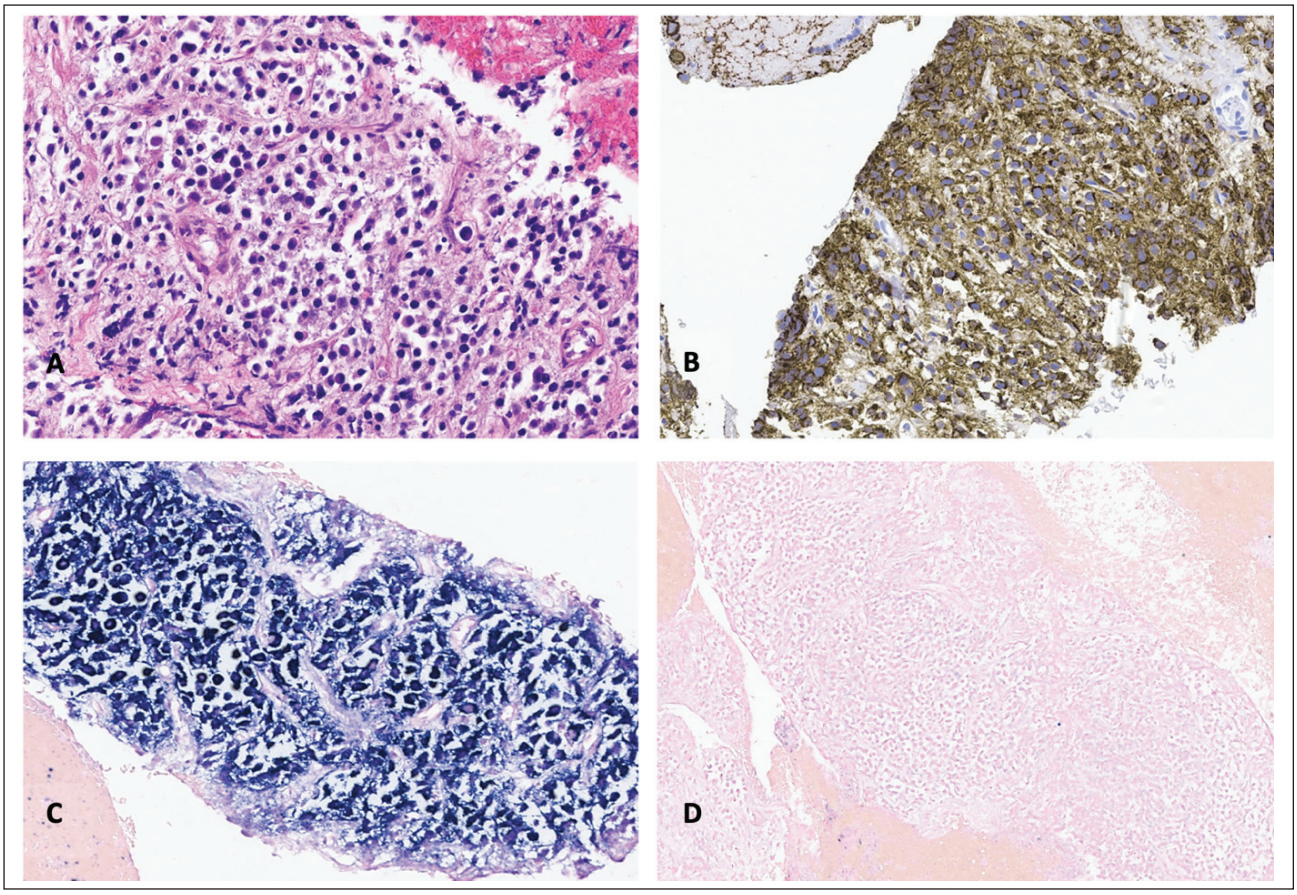

Figure 3. *EUS guided FNA from pancreatic mass. (A). Aspirate smear demonstrating scattered lymphoid cells with a predominance of plasma cells in nests. Plasma cells are recognized by eccentrically placed nuclei and the granular appearance of the nucleus. (B) CD138 positivity confirming the presence of plasma cells which may be secondary to inflammation or a neoplastic population. (C) Lamba* In-situ *hybridization positive stain consistent with lamba chain expression. (D) Kappa* In-situ *hybridization negative stain. The presence of lambda expression and the absence of kappa expression signifies a monoclonal or malignant population of plasma cells supporting the diagnosis of a plasma cell neoplasm.*

jaundice and abdominal pain [9,10]. Abdominal pain may be attributed to acute pancreatitis or from rapid tumor growth [11]. Other symptoms include weight loss, anorexia, and upper gastrointestinal bleeding [12].

Radiological features are non-specific. CT findings typically describe a discrete, well-circumscribed, homogenous, and hypoattenuating lesion which is often multilobulated [8,13]. Positive emission tomography scan demonstrates increased uptake similar to other pancreatic neoplasms. The differential diagnosis included pancreatic adenocarcinoma, lymphoma, and metastatic disease [14]. Laboratory findings are non-specific and tumor markers including CA 19-9, CEA, CA-125, and AFP are usually normal [10], which aids in suggesting a hemopoietic etiology. Definitive diagnosis is best achieved with EUS-guided biopsy. Immunostaining for CD138, CD79a and Lamba/Kappa light chain *in-situ* hybridization can aid in the diagnosis [15]. As seen in our patient pancreatic plasmacytomas can increase rapidly in size and thus unsurprisingly can present as obstructive jaundice.

Management is guided by the presence or absence of diffuse multiple myeloma, prior treatment regimens, cytogenetic features, and gene expression profiles [2]. Treatment options include chemotherapy, immunotherapy, or radiation; however, the prognosis remains poor [2].

Conclusion

Pancreatic plasmacytomas are rare and associated with a poor prognosis. They often present as pancreatic head tumors and require tissue diagnosis. It may serve as the initial manifestation of relapsing multiple myeloma warranting prompt treatment. Pancreatic plasmacytomas can be rapidly progressive and often lethal. Bile duct stenting may be performed for symptomatic relief in patients with obstructive jaundice. This case serves as a reminder to the astute gastroenterologist to consider extramedullary plasmacytoma when a patient with multiple myeloma presents with a pancreatic mass, particularly when encountered with rapid interval growth as seen in our patient.

List of Abbreviations

CT	Computed tomography
ERCP	Endoscopic Retrograde Cholangiopancreatography
EUS	Endoscopic ultrasound

EMD Extramedullary plasmacytomas
FNA Fine needle aspirate
ISH In-situ hybridization stain

Conflict of interests
The authors declare that there is no conflict of interest regarding the publication of this article.

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Consent for publication
Consent obtained from family for submission of case report.

Ethical approval
Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	84 years, male
2	**Final diagnosis**	Pancreatic plasmacytoma
3	**Symptoms**	Acholic stool, pruritus, and jaundice
4	**Medications**	NA
5	**Clinical procedure**	EUS, ERCP, FNA
6	**Specialty**	Gioncology