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A Rare Cause Mimicking Pancreatic Cancer: Extramedullary Plasmacytoma

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ABSTRACT

Background: Secondary or metastatic tumors of the pancreas are exceedingly rare. This study aims to elucidate an uncommon etiology contributing to pancreatic masses: extramedullary plasmacytoma.

Case Report: A 77-year-old male patient, undergoing treatment for multiple myeloma, presented with jaundice and abdominal pain. Laboratory findings indicated cholestasis. Imaging techniques revealed a dilated biliary tree and a hypoechoic mass in the head of the pancreas. Endoscopic examination showed significant narrowing of the duodenal lumen with an infiltrative pattern in the duodenal wall. Biopsies were taken from the narrowed area, and percutaneous biliary drainage was performed in the radiology department. Histopathological analysis of the biopsies revealed features consistent with plasmacytoma, characterized by infiltration with atypical plasma cells and positivity for CD138 in immunohistochemistry.

Conclusion: In cases where a patient with multiple myeloma presents with a pancreatic mass, it is imperative to consider the possibility of a pancreatic plasmacytoma as a potential diagnosis.

Keywords: CD138, cholestasis, extramedullary plasmacytoma, jaundice, pancreatic mass.

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INTRODUCTION

When a patient is found to have a pancreatic mass, the usual assumption often favors a diagnosis of primary pancreatic cancer, as metastasis to the pancreas is relatively uncommon, comprising about 2% of all pancreatic tumors.[1] The primary focus in cases of primary pancreatic tumors is pancreatic adenocarcinoma, seen in about 90% of cases.[2] However, this case report describes an extremely unusual situation where a patient was diagnosed with extramedullary plasmacytoma located in the head of the pancreas. This condition presented with jaundice and invasion of the duodenal wall.

CASE REPORT

A 77-year-old male patient presented with a one-week history of jaundice and abdominal pain while receiving treatment with lenalidomide and dexamethasone for multiple myeloma. Clinical examination revealed jaundice affecting both the sclerae and integument. Laboratory analysis

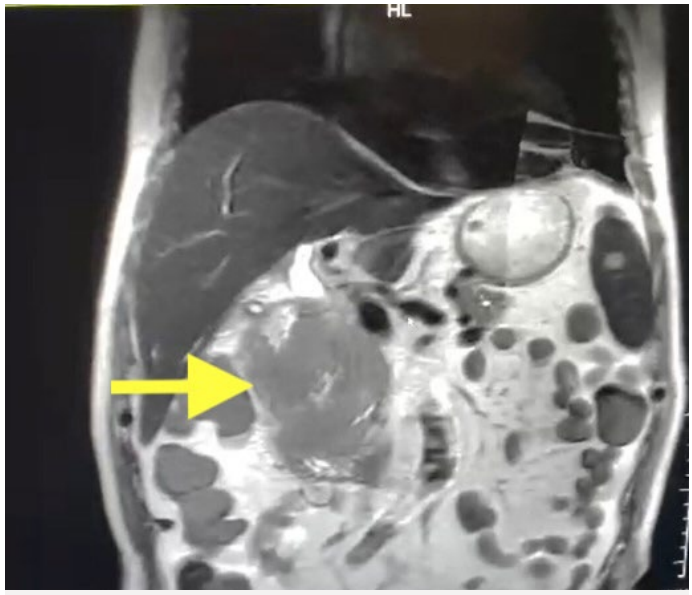

Figure 1. Pancreatic mass visible on the abdominal magnetic resonance imaging (MRI's) coronal plane, indicated by a yellow arrow.

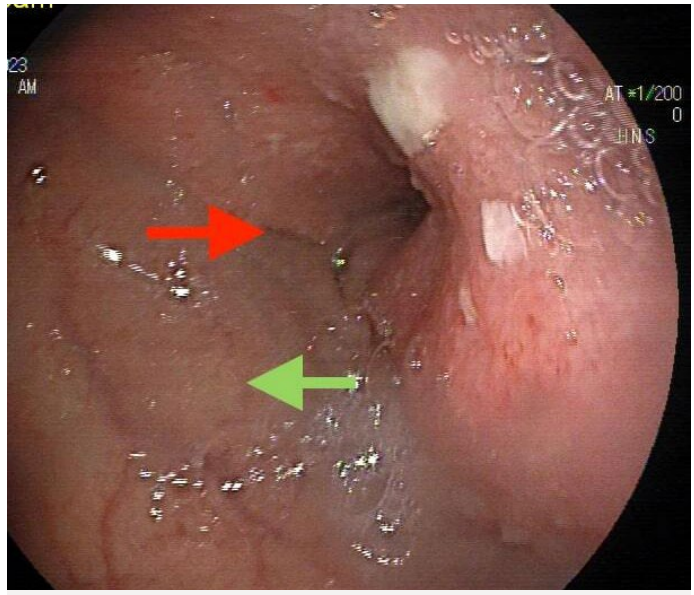

Figure 2. Normal duodenal mucosa on the left side, indicated by a yellow arrow; infiltrated irregular duodenal mucosa on the right side, indicated by a red arrow.

showed total bilirubin at 9 g/dL, direct bilirubin at 8 g/dL, alkaline phosphatase at 458 U/L, gamma-glutamyl transferase at 656 U/L, alanine transaminase at 147 U/L, and aspartate transaminase at 90 U/L. Additional findings included a white blood cell count of 10^3/µL, hemoglobin at 10.2 g/dL, C-reactive protein at 100 mg/L, and sedimentation rate of 106 mm/h. Carbohydrate antigen (CA) 19–9 levels were within normal limits. Imaging studies, including abdominal ultrasound (US) and magnetic resonance imaging (MRI), revealed intrahepatic and proximal common bile duct dilatation, along with a 70 x 65 mm mass in the head of the pancreas causing obstruction of the common bile duct. A thickened duodenal wall with luminal constriction was also noted (Fig. 1).

In response to suspected cholangitis, empirical antibiotic therapy was initiated, and plans were made for endoscopic retrograde cholangiopancreatography (ERCP). However, endoscopic visualization showed severe narrowing of the duodenal lumen that precluded the passage of the endoscope. The mucosal layer appeared edematous, rigid, and erythematous, which led to biopsy acquisition (Fig. 2). The ERCP was unsuccessful due to the luminal stenosis, necessitating the insertion of a percutaneous drainage catheter under interventional radiology guidance. Following this intervention, laboratory parameters normalized. Histopathological examination of the biopsied specimens demonstrated infiltration by atypical plasma cells, confirmed by positive CD138 immunostaining, consistent with extramedullary plasmacytoma (Fig. 3).

The patient was subsequently transferred to the hematology department for radiotherapeutic intervention and optimization of his medical management.

DISCUSSION

Extramedullary plasmacytoma is an uncommon type of tumor found in less than 5% of plasma cell tumors. It is typically diagnosed after multiple myeloma of the bone marrow has been identified. The submucosal lymphoid tissue of the upper respiratory tract is the most frequent site for extramedullary lesions, although they can affect any tissue or organ. In the gastrointestinal tract, extramedullary plasmacytomas are found in just 10% of cases, with the liver, spleen, and stomach being the most common detection sites.[3] Pancreatic involvement is not commonly observed, with a prevalence of only 2.3% in autopsies.[4] Focal involvement is the most common manifestation, although diffuse infiltration can also occur. Obstructive jaundice and abdominal pain are the predominant clinical symptoms, often occurring because the head of the pancreas is the most commonly affected site.[5] Pancreatic plasmacytomas are more commonly observed in older patients, with a higher occurrence in males, who are three to five times more likely to be affected than females.[6] Our patient, similar to cases reported in the literature, was an older male who presented with symptoms of jaundice and abdominal pain. Upon examination, a plasmacytoma appeared as a focal mass within the head of the pancreas.

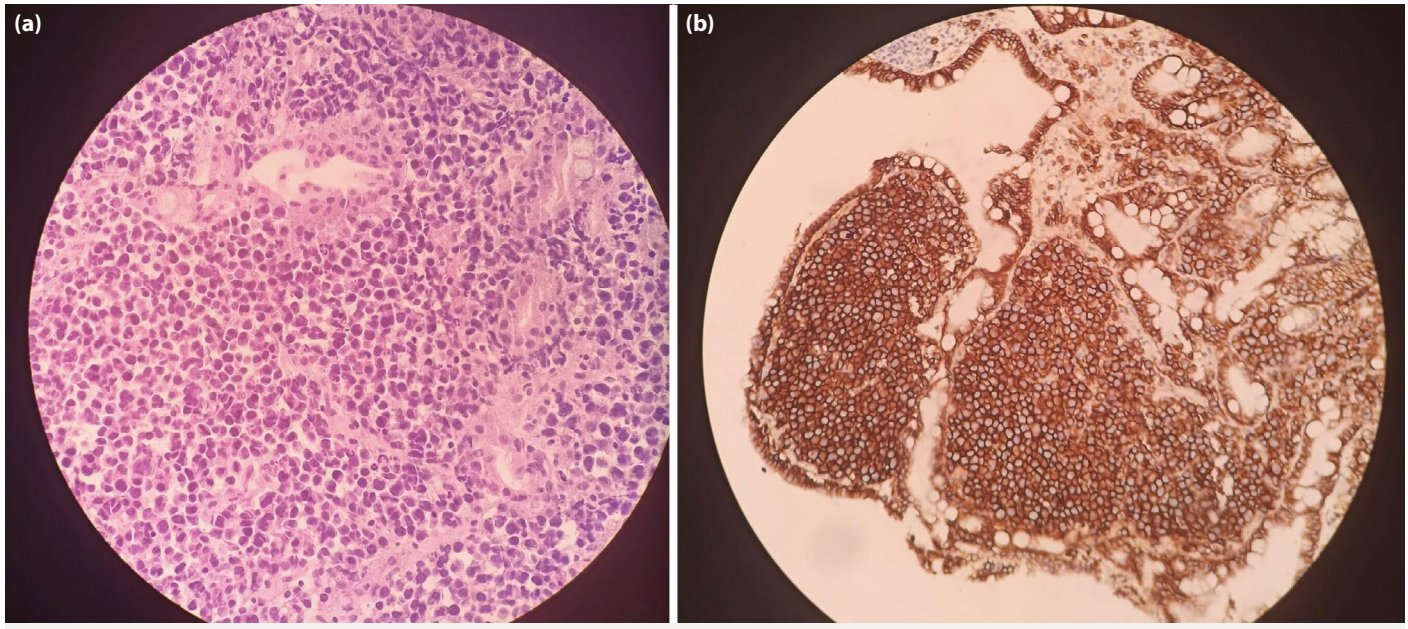

Figure 3. (a) Plasmacytoid cells with abundant eosinophilic cytoplasm and eccentrically placed nuclei, stained with Hematoxylin and Eosin (H&E), magnification 40x. **(b)** Diffusely and strongly positive immunostaining for the plasma cell marker CD138, magnification 20x.

Various imaging techniques such as ultrasound, computed tomography, magnetic resonance imaging, and endoscopic ultrasound (EUS) are used to diagnose the disease. However, the imaging results are often non-specific and can mimic findings associated with other pancreatic neoplasms such as adenocarcinoma, neuroendocrine tumors, lymphoma, and metastases.[5] We consulted with radiologists during our patient interactions.

Characterized by its hypoechoic appearance on US and hypointense signal on MRI, the medical team considered the possibility that the lesion could be a primary pancreatic tumor, specifically adenocarcinoma, especially in a patient who had prior exposure to chemotherapy agents.

A biopsy is the recommended procedure for diagnosing pancreatic plasmacytoma. Various methods for performing a biopsy include percutaneous, endoscopic, or surgical approaches. Endoscopic ultrasound fine-needle aspiration (EUS-FNA) is a highly efficient technique, with a diagnostic accuracy rate of 70–90%.[6] Among 63 patients with pancreatic plasmacytoma, 14 were diagnosed via endoscopic ultrasound-guided fine needle aspiration, 9 through computed tomography-guided fine needle aspiration, 11 by surgical biopsy, 7 through endoscopic biopsy, and 3 postmortem via autopsy.[7] Occasionally, the duodenal wall may experience infiltration, and a diagnosis can be obtained by performing direct endoscopic biopsies.[8]

Extramedullary plasmacytoma (EMP) of the pancreas may be misidentified as neuroendocrine (islet cell) tumors due to its rarity and the cytomorphological similarities between endocrine and plasma cells.

Immunohistochemical analysis on the biopsy specimen or flow cytometry on aspirated material is essential to confirm the monoclonality and establish a definitive diagnosis of a plasma cell neoplasm. The plasma cells demonstrate robust expression of CD38, CD138, and exhibit light-chain restriction.[9]

Endoscopy in our patient revealed direct infiltration of the duodenal wall. The use of biopsy instruments identified a diffuse infiltration of neoplastic plasmacytoid cells into the lamina propria. Cytomorphology showed an abundance of eosinophilic cytoplasm with nuclei arranged eccentrically. Strong and diffuse positive immunostaining for the plasma cell marker CD138 was observed by immunochemistry. Following the diagnosis of extramedullary plasmacytoma, the patient was transferred to the department of hematology.

CONCLUSION

When a patient with multiple myeloma presents with a pancreatic mass, it is crucial to consider pancreatic plasmacytoma as a potential diagnosis.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Author Contributions: Concept – IK, SU, ZA; Design – IK, SU, ZA; Supervision – IK, SU, ZA; Resource – IK, SU, ZA; Materials – IK, SU, ZA; Data Collection and/or Processing – IK, SU, ZA; Analysis and/or Interpretation – IK, SU, ZA; Literature Search – IK, SU, ZA; Writing – IK, SU, ZA; Critical Reviews – IK, SU, ZA.

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