Multiple myeloma presenting as plasmacytoma causing obstructive jaundice

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ABSTRACT

Jaundice, weight loss, and anorexia are common symptoms that raise concern for pancreatic malignancy. Although the most common form of pancreatic malignancy is pancreatic adenocarcinoma, not all pancreatic malignancies are exocrine in origin. Pancreatic plasmacytomas are plasma cell tumors that can lead to similar presentations. Plasmacytomas are classified as either intra- or extramedullary, depending on their location; intramedullary plasmacytomas are more common than extramedullary plasmacytomas. Here, we present a case of pancreatic plasmacytoma in association with advanced multiple myeloma diagnosed using endoscopic ultrasonography-guided fine-needle aspiration.

KEYWORDS Esophageal ultrasound; multiple myeloma; plasmacytoma; pancreas

Extramedullary plasmacytomas represent a rare type of plasma cell tumor that can present outside of the bone marrow. They can occur as a sequelae of advanced multiple myeloma with extramedullary involvement or as a solitary plasma cell tumor without bone marrow involvement. Although extremely uncommon, they should be considered in the differential diagnosis among those presenting with suspected pancreatic mass and obstructive jaundice. Endoscopic ultrasonography-guided fine-needle aspiration (EUS-FNA), which has an excellent diagnostic accuracy and safety profile, is the modality of choice for establishing tissue diagnosis. Here, we present a case of obstructive jaundice caused by extramedullary plasmacytoma mimicking a pancreatic adenocarcinoma, diagnosed by EUS-FNA.

CASE DESCRIPTION

A 72-year-old white man presented to our institution with complaints of abdominal discomfort and painless jaundice. On admission, his lab work was significant for abnormal liver biochemistries with alanine transaminase 417 IU/L, aspartate aminotransferase 414 IU/L, alkaline phosphatase 1309 IU/L, total bilirubin 15.9 mg/dL, and direct bilirubin 13.4 mg/dL, with normal lipase, amylase, and

white blood cell count. Physical exam revealed diffuse jaundice; however, abdominal exam did not reveal any focal findings. Computed tomography of the abdomen and pelvis was significant for a large mass adjacent to the pancreatic head along with multiple retroperitoneal and intra-abdominal nodules (*Figures 1 and 2*). Subsequently, the patient underwent EUS-FNA of the pancreatic lesion. Tissue biopsy was positive for plasmacytoma. Histology showed sheet-like proliferation of predominantly immature atypical plasma cells with prominent nucleoli, with scattered mature plasma cells in the background.

The patient was subsequently scheduled for therapeutic endoscopic retrograde cholangio pancreatography (ERCP), but this was unsuccessful as the common bile duct could not be cannulated due to bulging papilla. As such, interventional radiology-guided biliary drainage was placed with successful decompression of the biliary tree and corresponding improvement in liver biochemistries. Further workup showed evidence of widespread plasmacytomas including involvement of heart and bone. No M protein was detected in serum or urine protein electrophoresis. Free kappa and lambda light chain levels in urine were 36.7 (reference range 1.35–24.19) and 3.4 (0.24–6.66), with a kappa/lambda ratio of 21.29 (2.04–10.37). X-rays demonstrated lytic lesions of the right humeral head and proximal humeral neck, and lesions

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Figure 1. Contrast-enhanced axial CT image demonstrates a large homogeneously enhancing mass of the pancreatic head and neck causing compression of the distal common bile duct.

Figure 2. Contrast-enhanced coronal CT image demonstrates an ill-defined mass in the head and body of the pancreas causing common bile duct obstruction.

of the right and left ilium. He subsequently underwent chemotherapy with significant improvement observed on the most recent positron emission tomography scan.

DISCUSSION

Multiple myeloma is defined as clonal proliferation of plasma cells. Plasmacytomas are considered tumors consisting of abnormal plasma cells that grow within the soft tissue or bony skeleton. They are classified as either being intra- or extramedullary, depending on their location, although intramedullary plasmacytomas are more common.¹

Pancreatic plasmacytomas remain a rare entity, with only 5% of all plasma cell neoplasms occurring outside of the bone marrow. They primarily affect men, with the mean age

at presentation being approximately 55 years.² Although the majority of extramedullary plasmacytomas have been reported in the upper respiratory tract, 10% of extramedullary plasmacytomas occur in the gastrointestinal tract. Gastrointestinal plasmacytomas are usually limited to the liver and spleen, with only a limited number of cases describing pancreatic involvement.^{3,4}

The typical presentation of pancreatic plasmacytoma involves abdominal pain and jaundice as a result of biliary tree obstruction. Here, we describe visceral involvement of multiple myeloma, which was brought to clinical attention through atypical involvement of the pancreatic head and body. Most similar reported cases describe microscopic plasma cell infiltration of the pancreas, making this presentation of a well-formed mass unusual.

Radiologic features of plasmacytomas can be described as either a focal or solid mass with homogenous intravenous contrast enhancement, making it rather nonspecific, as other pancreatic tumors share the same descriptive features.⁵ Plasma cell tumors are considered to be sensitive to radiation and chemotherapy; therefore, preferred treatment involves a combination of surgery, radiation, and chemotherapy.^{5–7} In most cases of biliary obstruction caused by the tumor, ERCP with stent placement is performed for symptomatic relief of obstructive jaundice. However, ERCP was unsuccessful in this case due to bulging of the papilla, requiring biliary drain placement by interventional radiology, with subsequent resolution of jaundice.

This case highlights the importance of keeping a broad differential when suspecting a pancreatic malignancy in patients with obstructive jaundice and those with a history of plasma cell disorders. Extramedullary plasmacytomas should be considered in multiple myeloma patients presenting with a pancreatic mass.

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