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CASE REPORT

A First Report of Endoscopic Ultrasound for the Diagnosis of Pancreatic Amyloid Deposition in Immunoglobulin Light Chain (AL) Amyloidosis (Primary Amyloidosis)

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ABSTRACT

Context Pancreatic involvement in systemic light chain (AL)-amyloidosis is exceedingly rare. Prior reports of endoscopic ultrasound (EUS) for the diagnosis of amyloidosis are also limited. **Case report** We report the first description of EUS-guided fine needle aspiration (FNA) for the diagnosis of primary AL-amyloidosis involving the pancreas. **Conclusion** EUS-FNA can be effectively utilized for the characterization and cytologic diagnosis of pancreatic amyloidosis and potentially other accessible extraluminal amyloid deposits.

INTRODUCTION

Immunoglobulin light chain (AL) amyloidosis, or primary amyloidosis, is a plasma cell dyscrasia resulting in abnormal extracellular deposition of fibrillary protein which can occur in association with multiple myeloma [1]. Systemic AL-amyloidosis can involve multiple organs; however, pancreatic involvement is exceedingly rare. Prior reports of endoscopic ultrasound (EUS) for the diagnosis of amyloidosis are also limited. We report the first description of EUS-guided fine needle aspiration (EUS-FNA) for the diagnosis of primary AL-amyloidosis involving the pancreas.

CASE REPORT

A 50-year-old man undergoing staging evaluation for multiple myeloma was referred for EUS evaluation of a pancreatic head mass seen on abdominal CT scan (Figure 1). EUS confirmed an approximate 26.9x22.4 mm mostly hypoechoic solid mass process with smaller anechoic spaces suggestive of cystic component involving the pancreatic head and adjacent *porta hepatis* region (Figure 2). EUS-FNA was performed with a 25-gauge needle (Cook Medical, Bloomington, IN, USA). A total of four trans-duodenal passes were taken without complication (Figure 3). Antibiotic prophylaxis to prevent infection was provided due to possible cystic component. Cytopathology revealed abundant amorphous acellular waxy appearing proteinaceous deposits (Figure 4). Congo red staining of these deposits demonstrated apple-green birefringence under polarized light indicative of amyloid (Figure 5). The patient underwent subsequent autologous stem cell transplantation for treatment of myeloma with subsequent resolution of the pancreatic process.

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Key words Amyloidosis; Endoscopic Ultrasound-Guided Fine Needle Aspiration; Pancreas

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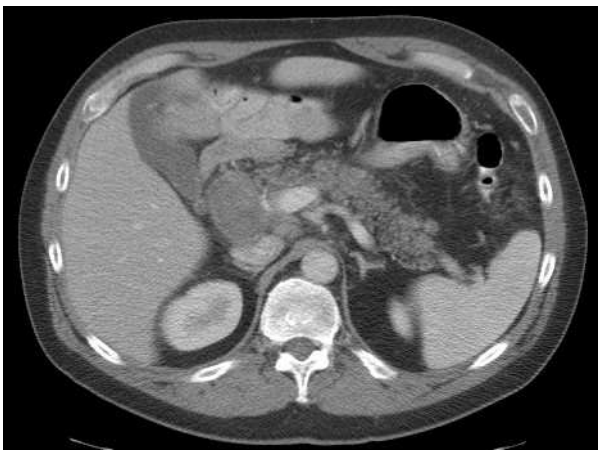

Figure 1. CT scan showing pancreas head process.

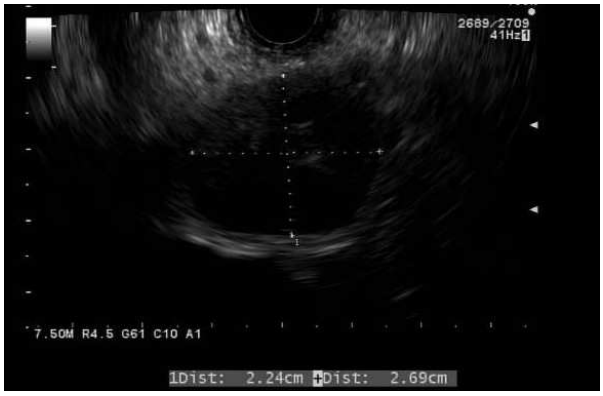

Figure 2. EUS image showing pancreatic head process (linear echoendoscope at 7.5MHz).

DISCUSSION

To our knowledge, this is the first description of EUS-FNA for the diagnosis of primary AL-amyloidosis involving the pancreas. Primary AL-amyloidosis is a plasma cell dyscrasia resulting in abnormal extracellular deposition of fibrillary protein which can occur in association with multiple myeloma [1]. Systemic AL-amyloidosis can involve multiple visceral organs including the mesentery and retroperitoneal space [1, 2]. Pancreatic involvement is nonetheless rare having been previously described only in isolated reports [3, 4]. Prior EUS descriptions of amyloidosis have been limited to a few reports of gastric and duodenal wall deposition [5, 6, 7, 8]. In these cases, EUS features of hypoechoic thickening of mucosal and submucosal layers with loss of normal gastric wall echo-layers were characterized. There are no EUS reports of isolated pancreatic or peripancreatic amyloid deposits to date. Pancreatic islet cell amyloidosis manifesting as a diffusely enlarged hypoechoic pancreas (mimicking pancreatitis) by trans-abdominal ultrasound has been reported [4]. The diagnosis of amyloid is typically made by core needle or excisional tissue biopsy with a subsequent tissue section stained with Congo red for the identification of amyloid. In this case, adequate cytologic material was obtained by

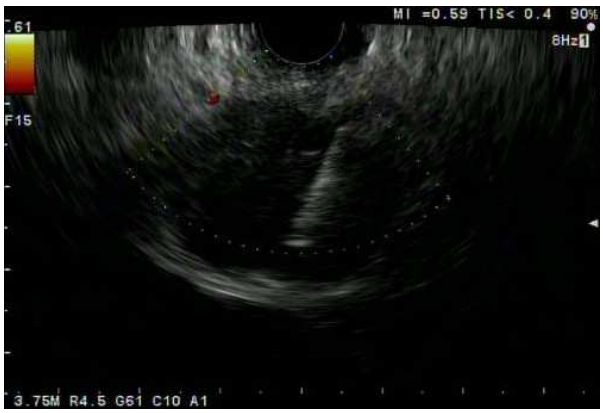

Figure 3. EUS image showing fine needle aspiration of pancreatic head process.

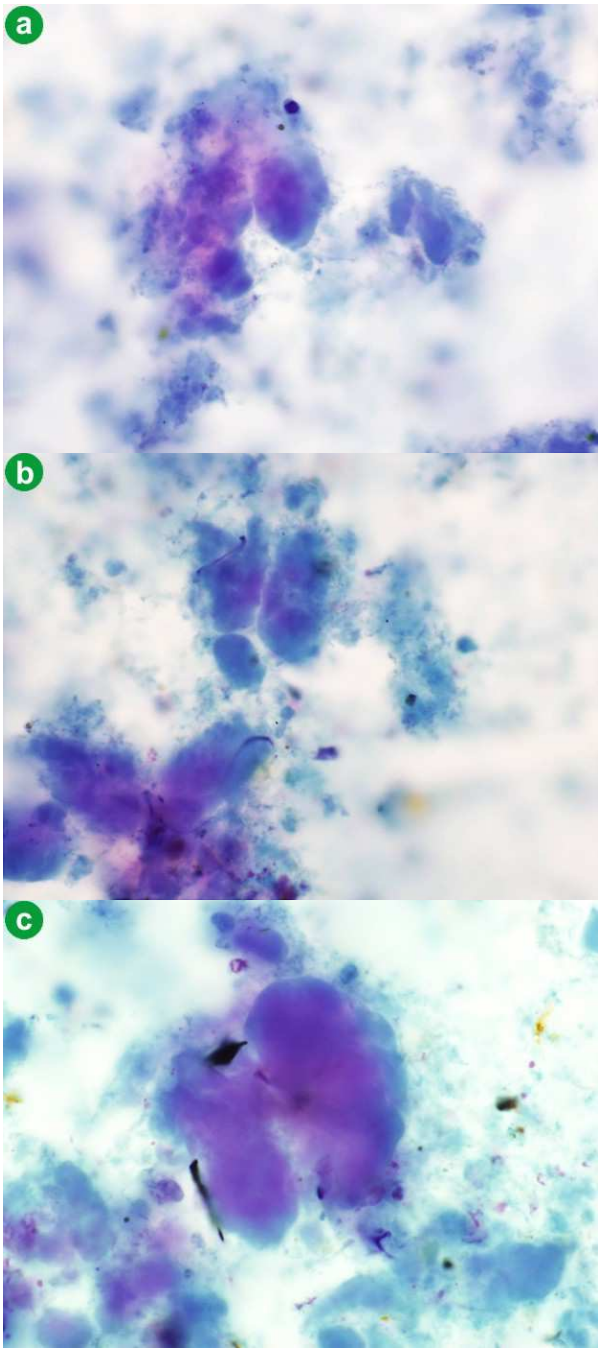

Figure 4. Select images of cytopathology slides from FNA of pancreatic process revealing abundant amorphous acellular waxy appearing proteinaceous deposits (Papanicoloau stain - high power).

FNA for direct smear and cytospin preparations which permitted confirmatory Congo red staining. This case illustrates that the recognition of characteristic cytologic features of amyloid, in particular the presence of amorphous waxy-like proteinaceous deposits, can provide clues to the diagnosis of amyloidosis and prompt Congo red staining for confirmation even in the absence of other clinical evidence for amyloidosis or myeloma [9, 10, 11]. In summary, EUS-FNA can be effectively utilized for the characterization and cytologic diagnosis of pancreatic

amyloidosis and potentially other accessible extraluminal amyloid deposits in select rare cases when indicated; excisional tissue biopsy from periumbilical fat or rectal biopsy are the usual method of choice for the diagnosis of amyloidosis.

Conflict of interest The authors have no potential conflict of interest

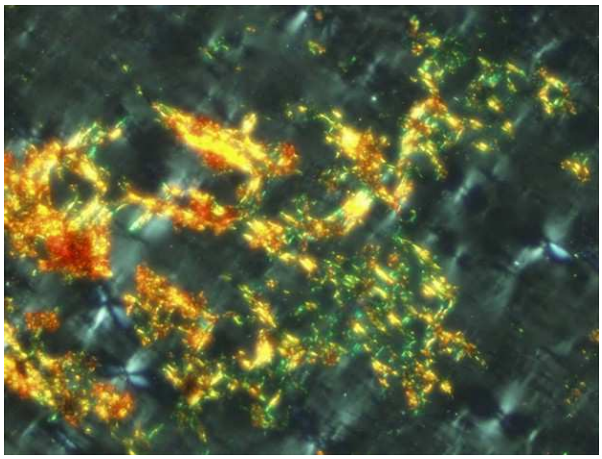

Figure 5. Direct smear from FNA of pancreatic head process stained with Congo red demonstrating apple-green birefringence under polarized light indicative of amyloid.

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