CASE REPORT

An unusual presentation of a benign pancreatic lesion containing amyloid

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SUMMARY

We present a unique case of a benign pancreatic lesion which was positive for amyloid in a 55-year-old female patient without systemic amyloidosis. Further testing revealed islet-type amyloid polypeptide (or amylin), a protein found in various diseases such as diabetes, insulinoma and pancreatic adenocarcinoma—none of which was seen in our patient.

BACKGROUND

Pancreatic lesions have a very broad differential but encountering amyloid can be surprising and unexpected as it is a very rare disease. This case report highlights the workup necessary when a patient is found to have amyloid deposits in the pancreas.

CASE PRESENTATION

A 55-year-old woman with history of non-small cell lung carcinoma status post chemoradiation therapy currently in remission presented to the emergency department with several days of nausea, upper back pain and light-headedness. She reported several episodes of blurry vision and dizziness over the past few years. Vital signs on presentation were within normal limits. Abdominal examination was benign with no evidence of tenderness, organomegaly or ascites. She did not have a history of smoking, alcohol use or illicit drug use.

INVESTIGATIONS

A transabdominal ultrasound showed an 8 mm hypoechoic lesion in the body of the pancreas (figure 1). Subsequent 3 T MRI confirmed a hypointense lesion on T1-weighted images (figure 2). An endoscopic ultrasound (EUS) with

Figure 2 T1-weighted 3 T MRI findings confirming a hypointense lesion.

fine-needle aspiration (FNA) of the lesion was performed (figure 3). Pathology revealed positive Congo red deposits in an islet-cell distribution (figure 4). Mass spectrometry detected a peptide profile consistent with islet-type amyloid polypeptide deposition (IAPP). Glucose levels were within normal limits. Insulin levels were not assessed as the patient was never hypoglycaemic. Plasma cell dyscrasias were ruled out with normal serum protein electrophoresis and negative serum free light chains. Other laboratory parameters including carbohydrate antigen 19-9 (CA 19-9), carcinoembryonic antigen (CEA), amylase and lipase were within normal reference ranges.

DIFFERENTIAL DIAGNOSIS

IAPP-type amyloid can be seen in pancreatic islets of patients with diabetes or insulinomas.¹ High levels of amylin are also seen in pancreatic adenocarcinoma in conjunction with diabetes.² Other disease entities to consider are multiple myeloma

Figure 1 Transabdominal ultrasound showing a hypoechoic lesion in the body of the pancreas.

Figure 3 Endoscopic ultrasound showing the pancreatic lesion.

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Figure 4 Amyloid deposits are amorphous, eosinophilic extracellular substance on the H&E stain (A) and has a glassy pink tinctorial quality on Congo red stain (B) (20× magnification).

and Waldenström's macroglobulinaemia, as they are associated with systemic amyloid light-chain (AL) amyloidosis.

TREATMENT

Surgical oncology was consulted but, given the benign pathology findings, surgical resection of the lesion was not advised.

OUTCOME AND FOLLOW-UP

The patient continues to experience intermittent non-specific symptoms and has been reassured that the pancreatic lesion is not malignant. It was emphasised to her that the lesion was not the cause of her symptoms. After discussion with multiple specialists, she has decided to follow-up with repeat MRI in 6 months with the option to pursue a repeat EUS if the size of the lesion has changed.

DISCUSSION

IAPP, or amylin, is a protein that is normally cosecreted with insulin in beta cells.³ It is involved in the regulation of glucose

Learning points

- ▶ Islet amyloid polypeptide (IAPP) is a hormone that is involved in the regulation of glucose, and hence, it plays an important role in the pathogenesis of glucose intolerance and type 2 diabetes.
- ▶ A pancreatic lesion containing IAPP should prompt a workup to rule out diabetes, insulinoma, pancreatic adenocarcinoma and multiple myeloma.
- ▶ Pancreatic involvement in systemic amyloidosis is rare. The mere presence of amyloid deposits in the pancreas is not diagnostic of systemic amyloidosis; other causes should be ruled out.
- ▶ When IAPP in pancreas is seen without any known cause (such as in our case), close follow-up surveillance imaging is advised.

by modulating gastric emptying and affecting satiety.⁴ Although IAPP plays an important role as a hormone, it is also thought to be involved in the pathogenesis of type 2 diabetes via aggregation in pancreatic islets causing cytotoxic properties and fibrosis, leading to beta-cell dysfunction and destruction.⁵

IAPP has also been seen in many neuroendocrine tumours, specifically insulinomas.⁶ Suspicion for an insulinoma was initially high in this patient as her history was strongly suggestive of hypoglycaemic events in the setting of an obvious pancreatic lesion. However, multiple fasting glucose measurements without hypoglycaemia militated against this. Plasma IAPP has also been found to be elevated in patients with pancreatic adenocarcinoma and diabetes.² There are data suggesting a causal association between type 2 diabetes mellitus and pancreatic adenocarcinoma,⁷ but it is unclear if increased levels of IAPP in the setting of pancreatic adenocarcinoma could be contributing to this association. Our patient's FNA of the lesion was benign, further making the aetiology of this patient's amyloid deposits unclear.

Cases of pancreatic amyloidosis are very rare. Onur *et al* published a case report where a patient presented with abdominal pain, glucose intolerance and a large pancreas (rather than a focal lesion) with a biopsy revealing amyloid deposits specifically in the islet cells.⁸ The patient in their case, however, had glucose intolerance which is not surprising given the known association of islet amyloidosis and diabetes.

This patient's presentation was unusual in that she had IAPP-type amyloid deposits in her pancreas despite the absence of any of the known associated comorbidities such as diabetes, pancreatic adenocarcinoma and multiple myeloma. Amyloid deposits in the pancreas can also be seen in systemic AL amyloidosis, but this is exceedingly rare and there are only few reports in the literature of this occurring.⁹ However, the amyloid deposits in these cases of systemic amyloidosis were not specifically located in pancreatic islets but in acinar tissues as well, suggesting that the mechanism of deposition in the two disease processes is different.¹⁰

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