A rare case of primary pancreatic lymphoma

Clinical History:

A 47-year-old man presented with a week history of epigastric pain. He had no sign of obstructive jaundice. On examination, a vague mass was felt at the epigastric region. Blood investigation showed raised serum amylase.

Imaging Findings:

Computed Tomography (CT) shows a large hypodense pancreatic head mass with little enhancement. The mass encases the gastroduodenal artery and surrounding peripancreatic vessels without causing occlusion or thrombosis. The common bile duct is encased but there is no pancreatic or biliary ductal dilatation. Minimal peripancreatic lymph nodes were noted. No other associated lymphadenopathy was found in the rest of the abdomen or pelvis. Overall findings are suspicious of pancreatic lymphoma.

As this was a potentially resectable disease if it was a case of pancreatic head adenocarcinoma, the likelihood of primary pancreatic lymphoma had been conveyed to the hepatobiliary surgeon. The decision of biopsy was made after a multidisciplinary team discussion.

Endoscopic ultrasound (EUS) guided biopsy was then performed and histopathological examination (HPE) revealed the diagnosis of a diffuse large B-cell lymphoma of the pancreas. The patient was thus started with chemotherapy.

Discussion:

BACKGROUND

Primary pancreatic lymphoma (PPL) is an extremely rare entity. It accounts for less than 0.5% of all pancreatic tumours [1,2,3]. PPL is usually of B-cell subtype of Non-Hodgkin's lymphoma as seen in our case. Lymphomatous involvement of the pancreas can be divided into primary or secondary, with secondary involvement being more common. Chemotherapy is the mainstay treatment for PPL and surgical intervention is only reserved for cases where histopathological diagnosis cannot be obtained.

CLINICAL PERSPECTIVE

Clinical presentations are very non-specific. Commonly, the patient will present with abdominal pain, weight loss, jaundice, nausea and vomiting. It can mimic acute pancreatitis or other pancreatic tumours clinically. Fever, chills, and night sweats which are specific to Non-Hodgkin lymphoma only present in 2% of the patients [1].

IMAGING PERSPECTIVE

CT examination is the most common imaging modality to characterise pancreatic lesions. There are two different
patterns of PPL [3]:
1) Diffuse involvement of the pancreas - the pancreas will appear diffusely enlarged;
2) Focal involvement of the pancreas, commonly the pancreatic head will be involved.

The focal involvement of the PPL is the one that will usually cause diagnostic and management dilemma as the imaging findings can mimic the more common pancreatic adenocarcinoma which will require surgical intervention. However, there are certain points in a CT examination that can favour the diagnosis of PPL, which are present in our case:
- Encasement of the surrounding vessels without causing thrombosis or occlusion;
- Biliary and pancreatic ducts are usually not dilated even though the tumour is extremely large;
- Homogenous tumour with little or no enhancement.

OUTCOME
Patient can undergo endoscopic ultrasound (EUS) guided biopsy or CT guided biopsy to confirm the diagnosis. The mainstay of treatment is chemotherapy.

There was a management dilemma in our case as this patient had a potentially resectable disease if it was a case of adenocarcinoma. CT/EUS guided biopsy might upstage the disease due to the risk of peritoneal seeding. However, in view of the radiological findings of likely PPL, EUS guided biopsy was decided upon after multidisciplinary team discussion.

TEACHING POINTS
Albeit rare, primary pancreatic lymphoma is an important entity to be considered when a patient presents with a pancreatic mass, it is likely that patients will have good response to chemotherapy if diagnosed early and surgery can be avoided. Radiologists need to be aware of its imaging findings and to raise the possibility of pancreatic lymphoma whenever appropriate.

Differential Diagnosis List: Primary pancreatic lymphoma - diffuse large B-cell lymphoma., Pancreatic adenocarcinoma, Secondary lymphoma

Final Diagnosis: Primary pancreatic lymphoma - diffuse large B-cell lymphoma.

References:
**Description:** Large pancreatic head mass with little enhancement (red arrow). Note the vessels are encased by this mass but there is no thrombosis or vessel occlusion. **Origin:** Department of Radiology, Sarawak General Hospital, Malaysia.
Figure 2

Description: Enlarged lymph node (red arrow). Note that the intrahepatic ducts are not dilated. Origin: Department of Radiology, Sarawak General Hospital
Figure 3

Description: Large pancreatic head mass (red arrow). Origin: Department of Radiology, Sarawak General Hospital