Primary Pancreatic Lymphoma - CT Imaging Features and Differential Diagnosis

Poster No.: C-1644
Congress: ECR 2015
Type: Educational Exhibit
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Keywords: Contrast agent-oral, CT, Pancreas, Lymph nodes, Abdomen, Lymphoma
DOI: 10.1594/ecr2015/C-1644

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Learning objectives

To illustrate the CT imaging features of primary pancreatic lymphoma (PPL) and review the differential diagnostic criteria from acute pancreatitis and pancreatic adenocarcinoma, entities which differ greatly in terms of treatment options and prognosis.

Background

PPL is an uncommon, extranodal manifestation of any histopathologic subtype of B-cell Non-Hodgkin’s lymphoma, comprising less than 0.5% of all pancreatic malignancies, and less than 2% of all extranodal Non-Hodgkin's lymphomas [1].

The clinical presentation may be nonspecific, with symptoms ranging from abdominal pain, nausea and vomiting, to weight loss and small bowel obstruction [2].

The classic triad of Non-Hodgkin’s lymphoma symptoms (fever, chills, night sweats) is present in only 2% of cases [3].

Laboratory findings include elevation of amylase and lipase in pancreatitis, LDH elevated levels (although this can be observed in many lymphoproliferative disorders, including Non-Hodgkin’s lymphoma, but also in some cases of pancreatic adenocarcinoma) and normal levels of serum CA 19-9, possibly slightly elevated in case of biliary obstruction, (whereas 80% of patients with advanced pancreatic adenocarcinoma have high CA19-9 levels) [4]. Therefore, an increase in LDH levels combined with normal values of CA 19-9 is highly suggestive of PPL, although they do not represent strict diagnostic criteria.

In a case of PPL, non-operative evaluation and biopsy of pancreatic masses can avoid the need for invasive surgery. Radiological-guided percutaneous fine needle aspiration of the pancreas and endoscopic ultrasound are two techniques that have greatly improved the accuracy of diagnosis while avoiding laparotomy [2, 5].

Treatment of PPL involves both chemotherapy (CHOP regimen) and radiotherapy [6]- without surgical resection - with outcomes favourably to surgical series [7].

Findings and procedure details

In order to distinguish PPL from secondary involvement of the pancreas by Non-Hodgkin's lymphoma, Behrns’ clinical and diagnostic criteria [8] must be met. These include:
• a mass predominantly within the pancreas, with involvement of peripancreatic lymph nodes
• no distal lymph node involvement
• no palpable superficial lymphadenopathy
• no hepatic or splenic involvement
• no mediastinal nodal enlargement on chest radiograph
• normal white cell count

With PPL, most lesions are typically of uniform low attenuation and appear homogenous, but small heterogeneous areas within a tumor mass can be seen in isolated cases. Enhancement after the administration of IV contrast medium is usually poor, yet homogenous [1].

There are two recognised morphological patterns of pancreatic lymphoma [9]:

1. the localised, well circumscribed tumoral form (which occurs in 80% of cases in the pancreatic head, with a mean size of 8cm)
2. the diffuse form (general enlargement of the gland, infiltrating or replacing most of the pancreas)

Confirmed case of primary pancreatic lymphoma Fig. 1 on page 4, Fig. 2 on page 5

• In this case, the main differential diagnosis was made with acute necrotising pancreatitis, which represents the liquefactive necrosis of the pancreatic parenchyma, appearing as often multifocal lack of parenchymal enhancement. Fig. 3 on page 6, Fig. 4 on page 7

The infiltrative form may mimic the imaging findings of interstitial edematous pancreatitis Fig. 5 on page 8, Fig. 6 on page 9, Fig. 7 on page 10, Fig. 8 on page 11, which has:

• typically smooth parenchymal enhancement
• mild peripancreatic fat stranding
• minimal peripancreatic fluid
• no findings of necrosis

(according to the revised Atlanta Classification of Acute Pancreatitis)

On the other hand, the well circumscribed form can be easily misinterpreted as ductal adenocarcinoma Fig. 9 on page 12, Fig. 10 on page 13, Fig. 11 on page 14, Fig. 12 on page 15, especially in patients with dilatation or encasement of the pancreatic and common bile ducts [9].
Dilatation of Wirsung's duct can occur in PPL, pancreatic adenocarcinoma, as well as in acute pancreatitis. The ratio of duct diameter to the overall dimension of the gland may help differentiate between these pathologies:

- <0.5% - indicates PPL or pancreatitis [9]
- >0.5% - suggestive of adenocarcinoma (because of atrophy of the acinary elements associated with complete obstruction)[10]

Consequently, a bulky, cephalic pancreatic tumor which does not associate significant dilatation of the main pancreatic duct suggests the diagnosis of PPL, rather than that of adenocarcinoma.

Other strong indicators to the imaging diagnosis of PPL are[1]:

- enlarged lymph nodes below the level of the renal veins
- an invasive tumor growth pattern, which does not respect anatomic boundaries, infiltrating retroperitoneal or upper abdominal organs and the gastrointestinal tract

Furthermore, PPL can be decisively excluded if calcifications or necrosis are encountered [1].

Images for this section:
Fig. 1: Pancreas of globally increased dimensions, with multiple hypodense, nodular lesions, with a rim pattern of enhancement, disseminated throughout the gland and associated with local enlarged lymph nodes. Arterial phase contrast enhanced CT examination.
Fig. 2: Pancreas of globally increased dimensions, with multiple hypodense, nodular lesions, with a rim pattern of enhancement, disseminated throughout the gland and associated with local enlarged lymph nodes. Portal venous phase CT examination.
**Fig. 3:** Acute pancreatitis with focal necrosis. Pancreas of increased dimensions, with a heterogeneous structure in the cephalo-corporeal segments, with low attenuation areas which present no enhancement after IV administration of the medium contrast. Bilateral thickening of the perirenal anterior fascia. Edema along the margins of the pancreas and liver, extending into the lesser sac and the left paracolic space. Strandng of the peripancreatic fat and of the root of the mesentery. Arterial phase contrast enhanced CT examination
**Fig. 4:** Acute pancreatitis with focal necrosis. Pancreas of increased dimensions, with a heterogenous structure in the cephalo-corporeal segments, with low attenuation areas which present no enhancement after IV administration of the medium contrast. Bilateral thickening of the perirenal anterior fascia. Edema along the margins of the pancreas and liver, extending into the lesser sac and the left paracolic space. Stranding of the peripancreatic fat and of the root of the mesentery. Portal venous phase CT examination
**Fig. 5:** Acute edematous pancreatitis. Diffuse enlargement of the pancreas, with visible acinar structure. Stranding of the peripancreatic fat, extended to the left anterior perirenal fascia. Edema spread through the root of the mesentery and into the lesser sac. Arterial phase contrast enhanced CT examination
**Fig. 6:** Acute edematous pancreatitis. Diffuse enlargement of the pancreas, with visible acinar structure. Stranding of the peripancreatic fat, extended to the left anterior perirenal fascia. Edema spread through the root of the mesentery and into the lesser sac. Portal venous phase CT examination
Fig. 7: Acute edematous pancreatitis with fluid collections. General increase in size of the pancreas, with effacement of the contours. Peri-hepatic and peri-pancreatic fluid collections, extending into the lesser sac and laterally into the right paracolic space, distending posteriorly the left anterior perirenal fascia. Arterial phase contrast enhanced CT examination.
Fig. 8: Acute edematous pancreatitis with fluid collections General increase in size of the pancreas, with effacement of the contours. Peri-hepatic and peri-pancreatic fluid collections, extending into the lesser sac and laterally into the right paracolic space, distending posteriorly the left anterior perirenal fascia. Portal venous phase CT examination.
**Fig. 9:** Adenocarcinoma of the pancreas. Poorly defined pancreatic mass located in the body of the pancreas, with extension into the cephalic and caudal portions, with intensely irregular contours and heterogeneous structure, with gas inclusions. The mass has low, predominantly peripheral, inhomogenous enhancement, and determines the encasement of the splenic vein, superior mesenteric artery and vein. The lesion comes in contact with the second and third segments of the duodenum, apparently invading the wall of the later one. There are noticeable collateral perigastric and mesenteric vessels and peripancreatic and mesenteric lymph node involvement. Arterial phase contrast enhanced CT examination
Fig. 10: Adenocarcinoma of the pancreas. Poorly defined pancreatic mass located in the body of the pancreas, with extension into the cephalic and caudal portions, with intensely irregular contours and heterogeneous structure, with gas inclusions. The mass has low, predominantly peripheral, inhomogenous enhancement, and determines the encasement of the splenic vein, superior mesenteric artery and vein. The lesion comes in contact with the second and third segments of the duodenum, apparently invading the wall of the latter one. There are noticeable collateral perigastric and mesenteric vessels and peripancreatic and mesenteric lymph node involvement. Portal venous phase CT examination
Fig. 11: Pancreatic adenocarcinoma. Extensive low-attenuatin lesion, occupying the body and tail of the pancreas, hypoenhancing, with central necrosis. The mass invades the lesser gastric curvature, comes into contact with the left anterior perirenal fascia and effaces the contour of a jejunal loop. It encases the splenic artery and vein and determines partial thrombosis of the superior and inferior mesenteric veins. There are also evident enlarged perigastric, peripancreatic and mesenteric lymph nodes. Arterial phase contrast enhanced CT examination.
**Fig. 12:** Pancreatic adenocarcinoma. Extensive low-attenuating lesion, occupying the body and tail of the pancreas, hypoenhancing, with central necrosis. The mass invades the lesser gastric curvature, comes into contact with the left anterior perirenal fascia and effaces the contour of a jejunal loop. It encases the splenic artery and vein and determines partial thrombosis of the superior and inferior mesenteric veins. There are also evident enlarged perigastric, peripancreatic and mesenteric lymph nodes. Portal venous phase CT examination.
Conclusion

• Primary pancreatic lymphoma is a rare, potentially curable pancreatic tumour, whose symptoms, imaging and biological markers can be misleading, often being difficult to differentiate from those of pancreatic adenocarcinoma or acute pancreatitis.
• CT is by far, the most common imaging technique used in the evaluation of pancreatic masses, but the definitive diagnosis is made histopathologically.
• Confronted with such a mass, it is vital to know when to indicate a non-surgical biopsy, since the treatment for pancreatitis is largely supportive and PPL does not require surgical staging or a palliative Whipple's procedure before chemotherapy or radiation therapy.

Personal information

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