Algorithm periampullary tumor diagnosis and rational use of imaging techniques

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Learning objectives

To review the diagnostic imaging algorithm used for periampullary tumors (PAT) and to compare it with our clinical practice.

To describe the radiological semiology of PAT.
Background

INTRODUCTION

The PAT are a heterogeneous group of tumors arising within a maximum distance of 2cm of the major duodenal papilla (MDP) which the ampulla of Vater, formed by the confluence of the bile duct and main pancreatic duct (MPD), leads to Fig 1. In 38% of cases a "double-barreled opening at the apex of the and in 2% of cases the two ducts open independently into the duodenum. Fig 2

The MDP is located either in the posteromedial aspect of the middle third of the descending duodenum or as anatomic variant in 25% of the general population, in the horizontal part.

The PAT have an annual incidence of approximately 30 cases per million population and arise in the ampulla of Vater, bile duct or duodenum. In the literature, the pancreatic head cancer is the main responsible for the PAT; we excluded it from our study for constituting a different group.

Radiologic-Pathologic correlation.

The importance of histological differentiation of PAT resides in the different behavior, with a distinct therapeutic approach and prognosis.

PAT originating from ampulla of Vater. Ampullary tumors . Fig 3

They are uncommon tumors including adenoma, intestinal and pancreatobiliary carcinoma, poorly differentiated adenocarcinoma, mucinous and papillary intestinal invasive carcinoma subtypes.

The tumors of ampulla of Vater have a higher incidence in patients with hereditary polyposis syndromes and the intestinal-like subgroup have a better prognosis and response to treatment.

Morphologically the ampullary tumors can be classified into nodular (intramural or exophytic), infiltrative (periductal thickening in MRI) or mixed.

Recently has been observed a relation between the histological type and the imaging findings by associating the ampuloma of pancreatobiliar origin with the infiltrative
pattern, and intestinal ampuloma with the nodular pattern Fig. 4. The last one appears as an oval filling defect at the distal end of the common bile duct (CBD) on MR cholangiopancreatography (MRCP).

More often we observe indirect signs of ampullary pathology:

a) Double-duct sign: Fig. 6

It consists of simultaneous dilatation of the CBD and MPD secondary to a stop in the ampullary region.

b) Expansion of the biliary intra/extrahepatic and/or abrupt dilatation of the pancreatic duct with irregular edges without evidences of other findings (fig.7)

The visualization of a dilated MPD associated with a lobulated multiloculated cystic lesion, suggests intraductal papillary mucinous tumor (IPMT). Although sometimes only appears a segmental dilatation of MPD. Fig. 8

**PAT originating from the duodenum** Fig. 9

**Duodenal Adenocarcinoma**

It appears as an annular and irregular narrowing with nodular or ulcerated pattern, and usually hypovascular.

**GIST**

They are mesenchymal tumors of the gastrointestinal tract that can be:

1.- Benign: small ovoid or rounded appearance, with endoluminal location and homogeneous enhancement

2.- Malignant: large extraluminal lesions with heterogeneous enhancement due to central necrosis and irregular or lobulated edges.

**Adenomas**

They are classified into three types: villosus type (fig.11) which is hypervascular and potentially malignant, Brunner glands and tubular type (both benign).

**PAT originating from common bile duct** (Fig 12)
They are adenocarcinomas in 95% of cases. Imaging findings are:

1. Infiltrative-narrowing with an irregular and thickened duct trajectory (Fig. 13 y 14)

2. Polypoid or papillary intraluminal, rare.

In MRI they are often hypointense on T1-weighted images and hyperintense on T2-weighted images, and also hypovascular in regard to the adjacent parenchyma and show increased enhancement in delayed phase.

Metastases as cause of PAT are rare (Fig. 15, Fig. 16)

Independently of its origin, dilatation of intra and extra-hepatic bile ducts along with thickening of the wall of the second portion of the duodenum suggests PAT.

**Diagnostic Imaging**

Knowledge of the indications and limitations of the various imaging tests in the diagnosis of PAT and use of a protocol, improve the diagnostic performance of the PAT and the assessment of tumor resectability.

The initial approach depends on its clinical debut, often obstructive jaundice in 80% of cases and/or epigastric pain in 37-60%.

**1-ABDOMINAL ULTRASOUND**

For its accessibility and low cost is usually the first exploration when we suspect obstructive biliopancreatic pathology. The most common finding is dilatation of the intra and extra-hepatic bile ducts that will face towards distal obstructive disease. It can also display a mass of periampullary location, but its sensitivity is low both in the diagnosis and staging, and does not avoid the realization of other imaging tests.

**2.-MDCT**

It is currently the technique of choice for the diagnosis of PAT. Contraindications are due to ionizing radiation and the use of iodinated contrast media.

The biphasic acquisition technique, pancreatic (40-50 sec) and portal phase (60-70 sec) and performing 2D, 3D and vascular reconstructions, allow a better assessment of the periampullary region and vascular infiltration, making MDCT the best technique in the diagnosis, staging and assessment of tumor resectability.
The treatment and prognosis of PAT are determined by the degree of tumor infiltration, presence of lymphadenopathy, distant metastases and especially by histological type and local vascular infiltration, defining the same criteria for unresectability as for pancreatic head tumors. (Fig. 17 and Fig 18)

With the incorporation of MDCT and vascular reconstruction techniques, the sensitivity and specificity for assessment of tumor resectability are 90-100% and 94-98% respectively, with a PPV for resectability of 91-98% and a NPV of 99%.

One limitation is the possible confusion of vascular invasion with fibrosis after chemotherapy administration.

3-ENDOSCOPIC ULTRASONOGRAPHIC (EUS)

The greatest contribution of EUS to the diagnosis of PAT is the possibility of taking samples of the lesions and provide guided endoscopic treatment (endoscopic papillectomy, reserved to the ampullary superficial adenomas).

It is the technique with greater NPV for detection of PAT. Its specificity to define vascular invasion is high, being a routine indication when there are doubts about vascular invasion of portal and splenic veins in CT, but with more limitations for SMA and SMV.

It is an invasive test and requires highly trained personnel for implementation, so its availability is limited.

4-MR

Its contraindications include the inherent ones in the achievement of the test, and its limitations are the lower spatial resolution with regard to the CT and the lack of visualization of the papillary region, which can occur without fluid signal because of functionally frequent contractions of the Oddi.

For this we should make dynamic MRCP of the sphincter of Oddi, which is performed with radial slices repeating for 5 minutes every 5 seconds.

The indication is the evaluation of the biliary obstruction, with a sensitivity and specificity similar to CT in the evaluation of the tumor mass and in the study of vascular invasion. It plays an important role in the filiation of dubious hepatic lesions on MDCT.

The MRI scan includes upgraded sequences in T1 in phase and out of phase, upgraded sequences in T2, SSFSE and SPGRE 3D T1 gradient sequences with fat saturation before and after administration of gadolinium. The dynamic study should be triphasic
including an arterial-pancreatographic (35-45 s), a portal phase (70 s) and a retarded phase (180-300 s).

SSFSE cholangiographic sequences of thick slices are performed, obtaining high spatial resolution.

Its sensitivity and specificity is similar to that of the MDCT, so it is an alternative to this.

ERCP is an invasive procedure not without complications, which is included within the therapeutic techniques. Although sampling the lesion allows diagnosis, we don’t include it in the imaging diagnostic algorithm.

Our experience

In our hospital, between 2007 and 2011, 26 cases of PAT were histologically diagnosed after surgical resection (15 males and 11 females) of whom 10 were ampullary, 10 cholangiocarcinomas and 6 duodenal tumors (GIST 1, 2 villous adenomas, 2 duodenum adenocarcinomas and 1 duodenal metastasis of sarcoma).

21 ultrasound were performed as initial imaging test in patients who started with obstructive jaundice and 19 cases presented dilated intrahepatic and / or extrahepatic bile ducts. One of them showed dilated bile ducts and a space-occupying mass lesion in ampulla suggesting ampuloma, and in one case no sonographic findings were observed.

From the total of 24 cases studied by CT, in 21 were previously examined using ultrasound and in 3 of them the CT was the initial study. PAT was detected as most likely diagnosis in 16 cases, and 7 of them were also studied by MRI, without providing further details. In two of them EUS was performed being diagnostic after FNA study.

In the 10 cases, 2 without CT study and 8 with negative CT study, the diagnosis was made by ERCP in 9 of them and by MRI in 1 case.
Fig. 1

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Fig. 2

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Fig. 3

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Fig. 4: Male, 71, jaundice, pruritus. b, c Ampullary mass a, c Dilation of the extrahepatic bile duct. AP: Intestinal-type ampullary adenocarcinoma with lower histologic grade.

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Fig. 5: Female, 49, postprandial epigastric pain and pruritus since 1 month. a. nodular intraluminal protrusion of the papilla. b. CholangioMR. Choledochal dilatation with distal oval filling defect. c,d. Dilation of intra and extrahepatic biliary tract. AP- Ampullary adenocarcinoma moderately differentiated.

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**Fig. 6:** Male, 46, pain in right hypochondrio and weight loss since 2 month. a, b.- Dilation of the biliary tract and Wirsung. AP: Ampullary ductal adenocarcinoma. DOUBLE DUCT SIGN c-CholangioMR. Choledochal and Wirsung dilatation and the two both separated in the ampullary zone.

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Fig. 7: Male, 66, jaundice. a, b, Dilatation of intra and extrahepatic bile duct. AP: villous ductal papilloma

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Fig. 8: Female, 64. Postprandial epigastric pain. a, b, c, Segmental cystic dilatation of Wirsung in pancreatic body and tail. AP: intraductal papillary mucinous neoplasm.

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Fig. 9

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Fig. 10: Female, 65. Anemia and duodenal ulcer. She presents duodenal tumor adjacent to pancreatic head. a, b, c, d. Nodular lesion dependent of duodenal mucous. e- slightly hyper intense in T2 f-SPGR3D T1 (LAVA) CORONAL desiccate of duodenal mucosa by mass. AP. Low-grade GIST

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Fig. 11: Hyperattenuated nodule with duodenal filling defect. AP-Duodenal villous adenoma

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Fig. 12

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Fig. 13: Male, 72. jaundice for 10 days evolution, epigastric pain and pruritus. a, e. Intrahepatic duct dilatation b,c. Enhancing of the distal bile duct wall. d,e. Solid nodule slightly hyperattenuated AP. Well differentiated Cholangiocarcinoma

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Fig. 14: Female, 65. obstructive jaundice. a- increased circumferential enhancement in distal common bile duct. b,c, hyperattenuated nodule. d-Distal bile duct obstruction. AP Ulceroinfiltrating Cholangiocarcinoma

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Fig. 15

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Fig. 16: Female, 27. Family history of alveolar sarcoma of the tigh and lung. Metastases confirmed by biopsy. Debuts with postprandial epigastralgia, intense asthenia and melena. a. TC without contrast. Mass who rejects pancreatic head. b-SSFSE, c- LAVA dynamic with contrast, d-T1 without contrast, e-croronal LAVA dynamic, f- DWI B 500 intraduodenal mass with central necrosis suggestive of metastases confirmed in biopsy.

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Imaging findings OR Procedure details

Diagnostic Imaging

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<table>
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<th>Grade</th>
<th>Raptopoulos</th>
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<td>Unresectable</td>
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<td>4</td>
<td>Vascular occlusion</td>
<td>Circunferential involvement &gt;75%</td>
<td>Unresectable</td>
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**CT criteria of vascular involvement of neoplasia of head of the pancreas and periampullary tumors**

Modified from S. Gusmini et al. Vascular involvement in periampullary tumors: MDCT, EUS, and CDU. Abdominal imaging 2009; 34;519

**Fig. 17**

© S. Gusmini et al. Vascular involvement in periampullary tumors: MDCT, EUS, and CDU. Abdominal imaging 2009; 34;519
CT criteria for unresectability of pancreatic and periampullary tumors

1- Vascular peripancreatic invasion
   - Vascular occlusion
   - Vessel encasement
   - Circumferential involvement > 50%

2- Metastases to lymph nodes beyond regional pancreatic nodes

3- Hepatic metastases

4- Peritoneal implants and malignant ascitis


Fig. 18

Conclusion

The most profitable algorithm in the diagnosis and staging of the PAT is performing MDCT followed by endoscopic ultrasound tumor in the cases with dubious resectability. Fig. 19. The MRI has a sensitivity, specificity and diagnostic accuracy similar to MDCT, used as an alternative to this, for visualization of biliary ducts or to classify the dubious lesions during the staging process.

Endoscopic retrograde cholangiopancreatography is considered a therapeutic technique, being MDCT the technique of choice for staging and diagnosis of vascular invasion.

The role of ultrasound is limited to the initial assessment of biliary ducts in the study of obstructive jaundice, although put on the track of the existence of periampullary pathology.

In our clinical practice the lack of availability of EUS increased the number of MR studies performed after MDCT, without improving diagnostic accuracy, which corroborates the profitability of implementation of a diagnostic imaging algorithm for PAT. Fig. 20
Conclusions

- The difficulty of interpreting the imaging findings with therapeutic implications in various PT, necessitate combination of techniques that improve the diagnostic accuracy.

- The most profitable algorithm in diagnostic imaging and stanging of the PT is MDCT, followed by the EUS.

- MR is indicated as an alternative to MDCT in the evaluation of the biliary tract, or if in doubt about filiation of injuries in staging.

- In our study, the lack of availability of EUS increased the number of diagnostic tests performed without improving diagnostic accuracy.

Fig. 19

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Fig. 20

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