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TITLE: A radiologically atypical case of an ampullary neuroendocrine tumor (ANET)

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ABSTRACT
Ampullary neuroendocrine tumors (NETs) are rare tumors, which shows characteristic CT features. We present a case of a 42 years old female, in which the CT features of an ampullary tumor were atypical for a neuroendocrine tumor, but was confirmed on histopathology and immune-histochemistry as an NET. The uniqueness of our case was that the lesion showed mild enhancement in the arterial phase which was persistent in the venous phase with wash out in the delayed phase

Keywords: Ampullary region, Computed Tomography (CT), Double Duct Sign, Neuro-endocrine tumor,
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INTRODUCTION

Neuroendocrine tumors are neoplasms that arise from cells of the endocrine and nervous systems. Many are benign, while some are malignant. These tumors are most commonly found in the small intestine, with the most common subtype being carcinoid tumors. Pancreas, lung etc. are other sites where these tumors are found [1].

The ultrasonography of the patient was done using a 6 Hz probe. The CT was done on a 64 slice multi-detector CT scanner. A triple phase CT was done by intravenous administration of 100 ml of contrast medium at a rate of 3 ml /s with a monophasic rate of injection. Arterial phase scanning was started after a delay of 23 s and portal venous scanning after a delay of 60s. The 1 & 5 mm thick sections were acquired with subsequent coronal reconstruction.

CASE REPORT

A 42 year old female patient, presented to the surgery OPD with complaints of weight loss with episodic pain in abdomen, yellowish discoloration of the eyes, generalized itching, vomiting since 4 months. On examination, mild tender hepatomegaly was noted and a clinical diagnosis of Obstructive Jaundice was made. For further evaluation, an Ultrasonography of the abdomen was advised, which revealed (Figure 1A):

• A small hypoechoic mass, in the peri-ampullary region with central vascularity.
• Mild central & peripheral intra-hepatic biliary radicle dilatation.
• Dilatation of the Common Bile Duct and Pancreatic Duct (“Double Duct Sign”)
• Mild hepatomegaly with a normal gall bladder.

For further evaluation and extensions of the lesion, a CT scan of the Abdomen was performed which revealed (Figure 1B, 1C & 1D):

• A well-defined 24 x 23 x 22 mm sized, hypodense mass lesion showing mild arterial phase enhancement which persisted on venous phase in the peri-ampullary region, projecting into the lumen of 2nd part of duodenum with no adjacent extension or invasion.
• Mild central & peripheral intra-hepatic biliary radicle dilatation.
• Dilatation of the Common Bile Duct and Pancreatic Duct (“Double Duct Sign”)
• Mild hepatomegaly with a normal gall bladder.

Radiologically, after USG and CT correlation, a diagnosis of a peri-ampullary neoplasm was made. Esophago-gastro-duodenoscopy guided biopsy was performed.

Esophago-gastro-duodenoscopy revealed presence of a small sessile, fleshy mass in the peri-ampullary region jutting into the 2nd portion of duodenum (Figure 1E). A tissue biopsy was taken.

Histo-pathological (Figure 1F) and Immuno-histochemistry report suggested the diagnosis of a Low Grade Neuro-Endocrine Tumor (G1 – WHO Classification) as the tumor cells expressed Cytokeratin, Synaptophysin and Chromogranin.

**DISCUSSION**

Neuroendocrine tumors (NETs) are a large group of neoplasms with a wide spectrum of clinical, imaging, and pathologic characteristics. Imaging features of NETs can be challenging, and a multitude of imaging modalities may be required for accurate diagnosis. Accurate interpretation of the imaging findings is important to facilitate diagnosis and contribute to patient management.

Pancreatic neuroendocrine tumors (also known as NETs) are rare tumors arising from a putative common precursor, the APUD cell (amine precursor uptake and decarboxylation). These cells are of common embryological origin from the ectodermal ridge. These neuroendocrine tumors are histologically closely related to certain other neuroendocrine tumors like pheochromocytoma and medullary carcinoma of the thyroid. These tumors may or may not synthesize and secrete hormones, out of which the functioning tumors are those in which hormone secretion by the tumor results in a clinical syndrome. Non-functioning tumors are those in which either there is hormone secretion with no recognizable clinical features or no hormone secretion at all. Functioning tumors usually present early, due to the clinical symptoms, and pose a challenge for the radiologist [2].
Ampullary neuro-endocrine tumor (ANET) are rare tumors, accounting for less than 1% of all gastrointestinal neuroendocrine tumors and less than 2% of all tumors of ampullary region [2] [3].

Histologically, although ANETs are similar to neuroendocrine tumors arising from other parts of gastrointestinal tract, they have distinct immunohistochemical features. Immunohistochemical staining is the main diagnostic method for these tumors, and ANET stain positively with chromogranin A and synaptophysin in majority of the cases [3] [4].

According to the International Classification of Diseases of Oncology, published by the World Health Organization (WHO), the ampulla has been designated a site specific code “241” and neuroendocrine tumors are given specific codes: neuroendocrine not otherwise specified (8246); small cell neuroendocrine carcinoma (8041); and large cell neuroendocrine carcinoma (8013) [4].

Radiologically, on Ultrasonography, the tumour appearance is of a well-defined round mass, which is homogeneously hypoechoic in relation to the pancreas, and vascular on Doppler imaging. There may be a hyperechoic lesion or focal distortion of the pancreatic parenchyma, which may help in detecting the lesion, particularly in younger patients where the tumors tend to be less conspicuous due to the generally lower echogenicity of the pancreas. The use of enhancement pattern of these tumors using Ultrasound contrast agents, is currently under investigation. [5]

Functioning tumors are generally small, with low inherent contrast between the tumor and surrounding pancreas. Majority of islet cell tumors are hypervascular and will be best seen after intravenous injection of contrast. However, the best phase for the demonstration of these hyperattenuating small lesions is unclear [6] [7] [8]. Narrow window settings may help to improve detection. In our case, the lesion was well detected in the venous phase.

Insulinomas can be hypovascular, cystic and hypodense to the surrounding pancreas. [9]. Cystic pancreatic endocrine tumors are usually benign and non-functioning. It is difficult to differentiate them from other cystic pancreatic neoplasms on imaging alone [9]. In patients with a suspected gastrinoma, the imaging emphasis should be given to the ‘gastrinoma triangle’
Large tumors are likely to be non-functioning with central necrosis and are more likely to be malignant. The features which are associated with malignancy include necrosis, large size, infiltration of the surrounding structures and calcification [10].

CONCLUSION

The uniqueness of our case was that the lesion showed mild enhancement in the arterial phase which was persistent in the venous phase with wash out in the delayed phase. These features were atypical for ANET, which was later confirmed on histopathology and immunohistochemistry.

CONFLICT OF INTEREST

NIL

REFERENCES


TABLES
NIL

FIGURE LEGEND
Figure 1: (A) USG (6 Hz probe) showing the ampullary mass with vascularity within (red arrow). (B) CT (axial – arterial phase) showing mild enhancement of the ampullary mass (red arrow). (C) CT (axial – venous phase) showing mild enhancement of the ampullary mass (red arrow). (D) CT (coronal – venous phase) Dilated Common Bile Duct (red arrow) & Dilated Pancreatic Duct (orange arrow): Double Duct Sign. (E). Esophago-gastro-duodenoscopy showing the ampullary mass. (F) Histopathological image of the mass lesion showing monomorphologic population of small round cells.
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(F) Histopathological image of the mass lesion showing monomorphic population of small round cells.