Ampullary Carcinoid: A Case Report

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Ampullary Carcinoid: A Case Report

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BACKGROUND

- Carcinoid tumors of the Ampulla of Vater are rare low-grade, well differentiated tumors.
- They account for less than 1% of all gastrointestinal neuroendocrine tumors (NETs) and less than 2% of all tumors in the ampullary region.[1]
- The most common presenting symptoms are jaundice and non-specific abdominal discomfort.
- The recognition of these tumors is important, as historically patients with ampullary NETs have been more likely to develop metastases at smaller sizes and higher mortality than patients with duodenal NETs.[2]
- We present a case of ampullary carcinoid found in a patient with non-specific abdominal pain.

CASE PRESENTATION

- A 52 year old female presented with two years of epigastric pain and weight loss. The pain was predominantly nocturnal and associated with cramping. It would wake her from sleep, and cause nausea and vomiting. A trial of PPI was ineffective.
- **CT Scan Abdomen and Pelvis:** Abnormal round enhancing lesion in the ampullary region measuring 16mm with dilation of the extrahepatic biliary system.
- **EGD:** Confirmed a subepithelial appearing ampullary mass. Biopsy was negative for adenoma.
- **EUS:** Given the clinical suspicion for malignancy she underwent EUS. Showed a subepithelial ampullary mass measuring 18mm in diameter without bile duct obstruction. The mass was hypoechoic, with a thick rimmed submucosal border. Fine needle biopsies using 19 and 22-guage shark core needles were obtained. Cytology was diagnostic for ampullary carcinoid.
- **Cytology:** Showed the tumor was AE1/AE3 positive, CD45 negative, synaptophysin positive, chromogranin negative, CD56 positive, and Ki-67 involving <5% of tumor cells confirming a well-differentiated neuroendocrine tumor (typical carcinoid tumor).
- She underwent duodenotomy with resection of ampullary tumor, and biliary sphicteroplasty by surgical oncology.
- **Pathology:** Neuroendocrine tumor, well differentiated (carcinoid tumor) 1.5 x 1.4 x 1.0 cm greatest dimensions, ampulla of Vater. Tumor predominantly involves duodenal wall, duodenal submucosa and focally duodenal mucosa. Tumor focally involves deep margin of resection and focally involves lateral margin of resection. TNM tumor stage: pT2, pNX.
- A repeat EUS was performed four months after surgery with no evidence of residual lesion, and biopsies of the involved site were normal.

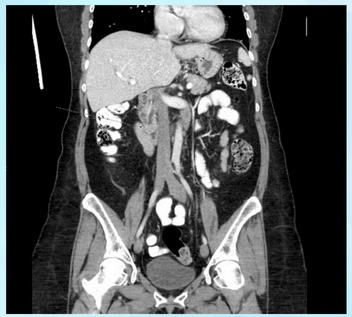

Figure 1: CT of the Abdomen and Pelvis showing an abnormal round enhancing lesion in the ampullary region possibly measuring up to 16 mm.

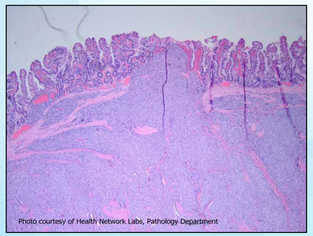

Figure 2

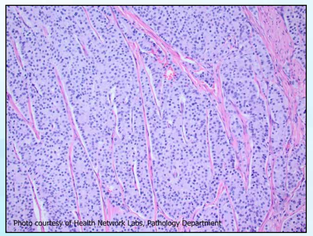

Figure 3

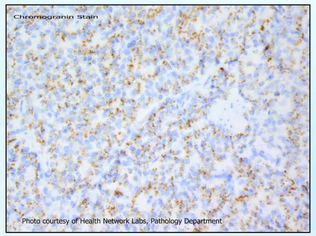

Figure 4

CONCLUSION

- NETs are low-grade endocrine cell tumors of the GI tract which rarely occur in the ampulla of Vater.
- The small number of documented cases limits the knowledge on their presentation and behavior.
- The most common method of diagnosis is EGD with biopsies, which can be followed by EUS to assess local invasion.[3]
- Pathologic findings consist of medium size cells with eosinophilc cytoplasm and central round nuclei with mild atypia and immunohistochemistry may show expression of chromogranin, synaptophysin, gastrin, somatostatin and/or neuron specific enolase.[4]
- Treatment is generally surgical resection of tumor and involved lymph nodes.
- Radical surgical resection with pancreaticoduodenectomy appears to be the appropriate treatment of ampullary carcinoids regardless of size.[4]
- It is important to keep this clinical entity in working differential diagnoses of abdominal pain and jaundice.
- As seen in our case EGD biopsies may be unrevealing necessitating further evaluation with EUS and core biopsies for definitive diagnosis.

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