Case Report

Periampullary Adenocarcinoma with Incidental Gastric Carcinoid Tumor: Report of an Extremely Rare Case

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Conflict of Interest: None
Received: 11.04.2021
Accepted: 14.09.2021
www.banglajol.info/index.php/JSSMC

Abstract:

Periampullary neoplasms are a heterogeneous group of tumors arising within 2 cm of the ampulla of Vater. Neuroendocrine tumors can originate throughout the entire body from neuroendocrine cells. These neoplasms exhibit deep differences, according to their origin and biological behavior. The synchronous presence of a primary tumor in patients affected by a neuroendocrine tumor is reported in the literature; incidence is variable and the most common site is the gastrointestinal tract. Here we report a case of 68 years old female presented with jaundice, anorexia and weight loss. She was mildly anaemic. Gall bladder was palpable. There was no organomegaly. Her CT scan and MRCP revealed a growth in periampullary region. So she underwent Whipple's procedure. Histopathological report showed synchronous presence of periampullary carcinoma and a carcinoid tumor in stomach. As the carcinoid tumor was nonfunctioning and very small in size, it was not identified in pre-operative work up.

Key Words:
Periampullary adenocarcinoma, Gastric carcinoid tumor

Key words: *Periamullary carcinoma, Carcinoid tumor, Synchronous*

[J Shaheed Suhrawardy Med Coll 2021; 13(2): 183-186]
DOI: https://doi.org/10.3329/jssmc.v13i2.65183

Introduction:

Periampullary neoplasms are a heterogeneous group of tumors arising within 2cm of the ampulla of Vater including ampullary adenocarcinoma, distal cholangiocarcinoma, duodenal adenocarcinoma and pancreatic ductal adenocarcinoma.[1] Neuroendocrine neoplasms can arise throughout the entire body from neuroendocrine cells; they are often found in the lungs and gastro-enteropancreatic (GEP) system, less frequently in the skin, breast and genitourinary tract .[2] Neuroendocrine tumors (NET) are sometimes associated with synchronous or metachronous primary malignancies.[3] A relationship exists between the presence of gastroenteropancreatic neuroendocrine neoplasms like carcinoid tumor and the simultaneous presence of primary neoplasms in other body regions. Pearson and Fitzgerald in 1944 reported, for the first time, a high incidence of carcinoid tumors with synchronous primary malignancy in an autopsy series.[4] Meanwhile, further reports demonstrated neuroendocrine neoplasms associated synchronous primary malignancy in up to 55% of cases.[5] In a meta-analysis, neuroendocrine tumor (NET) associated synchronous primary malignancy (SPM) averaged 17 % in 5280 patients.[6]

In order to explain the phenomenon of NET-associated SPM, the field-effect theory has been developed: a common carcinogenic effect that stimulates growth of neuroendocrine and SPM cancer cells simultaneously.[6] Moreover, NETs produce several neuropeptides/non-neuropeptides comparable to a growth factor and could promote the development of a synchronous primitive tumor within target organs. NETs produce and secrete various neuropeptides, while SPMs overexpress receptors for these compounds.[6] Since 1902, when the first neuropeptide secretin was discovered, about 60 such gut-brain peptides (neuropeptides) have been characterized.

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For an example, bombesin, a neuropeptide frequently produced and released by NET, stimulates in vitro growth of human breast cancer cells and is a potent autocrine growth factor in small cell lung cancers.[6]

In most of the cases the prognosis in NET associated SPM depends on SPMs.[7] It is observed that the growth of expression is almost doubled in SPMs than NET.[8] Moreover the clinical manifestation depends on the synchronous primary lesion rather than NETs; masking the presence of a neuroendocrine tumor.[8]

Here we present a case of a patient presented with periampullary carcinoma with synchronous carcinoid tumor in stomach.

Case Summary:

A 68 year old female presented to the surgery department of a private hospital with fluctuating jaundice for 2 months. She also gave history of melena for 2 times. Moreover, there was history of anorexia and weight loss for 1 month. On examination, she was deeply icteric and mildly anaemic.

Per abdominal examination revealed palpable gall bladder with no organomegaly. Her blood report showed decrease Hb level (9gm/dl), raised serum bilirubin (12.6mg/dl) and alkaline phosphatase (702U/ml). CA 19-9 was also elevated (>1000UI/ml). CT scan of abdomen showed dilated intra and extrahepatic biliary tree with distended gallbladder (Fig-1). MRCP also showed similar findings with double duct sign (Fig-2). Upper GI endoscopy revealed periampullary growth. After optimizing the patient by blood transfusion and nutritional supplements Whipple's procedure was carried out (Fig-3). There was no per or post-operative complications.

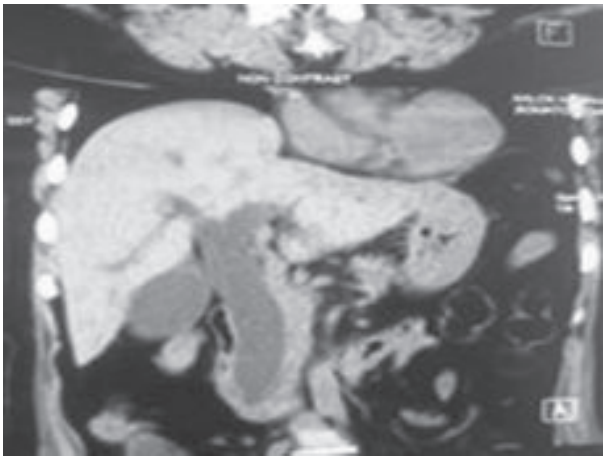

Fig.-1: *CT scan*

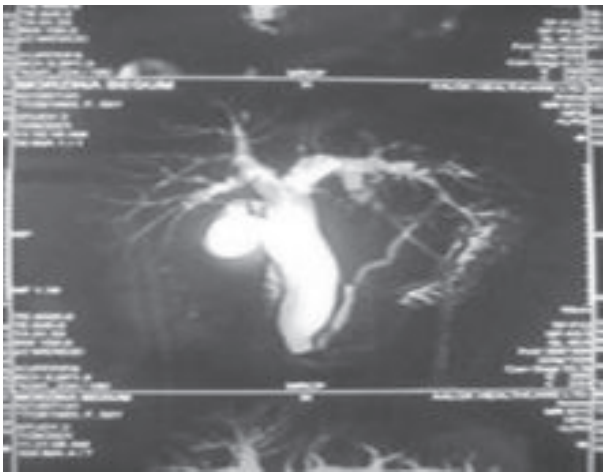

Fig.-2: *MRCP*

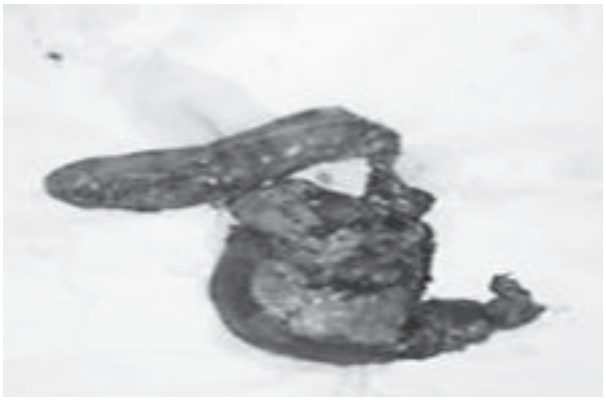

Fig 3: *Whipple's specimen*

Histopathology report showed well differentiated periampullary adenocarcinoma (pT2N0M0). But the section from stomach showed presence of carcinoid tumor indicating presence of both periampullary carcinoma and gastric carcinoid tumor synchronously (Fig-4).

Fig.-4: *Showing macroscopic appearance of periampullary carcinoma (red arrow) and gastric carcinoid tumor (blue arrow)*

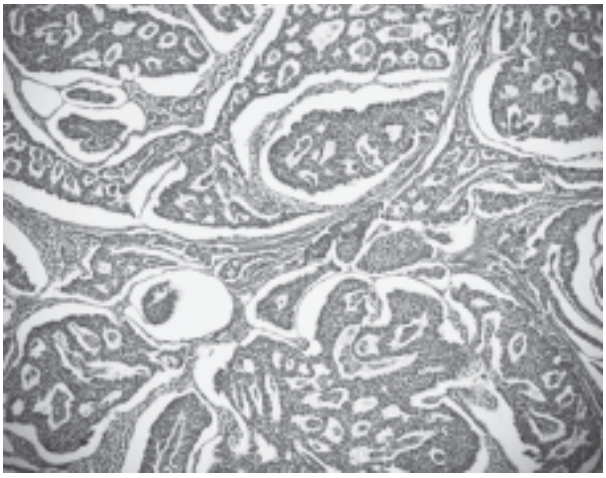

Fig.-5: *Showing microscopic appearance of gastric carcinoid tumor*

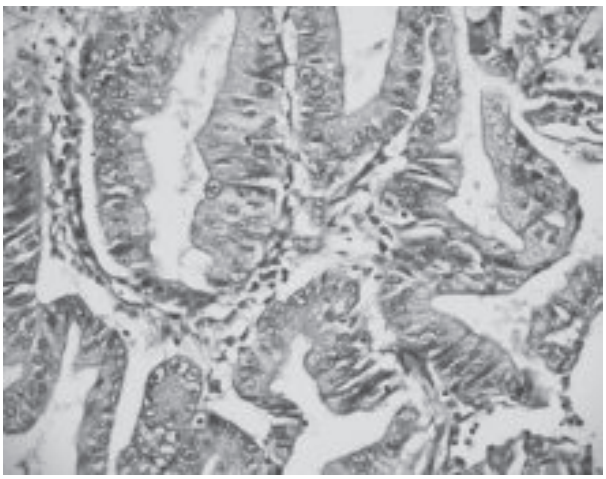

Fig.-6: *Microscopic view of periampulary carcinoma*

Discussion:

Periampullary adenocarcinoma is accounting for 2% of all malignant neoplasms of the gastrointestinal (GI) tract.[9] Its incidence is 2.9 new cases per million/year.[10] Diagnosis relies on several clinical, radiological and endoscopic investigations. Symptoms include nausea, anorexia, weight loss and abdominal pain in association with fluctuating jaundice due to failed bile drainage. An abdominal CT scan allows diagnosis and staging. MRI provides an anatomical representation of the biliary tree and of the main structures of the ampullary region. Surgical resection can be performed with pancreatico-duo-denectomy as the standard approach. In high-risk patients, local or endoscopic treatment can be offered. Five-year survival rates are about 10% in pancreatic adenocarcinoma and 40% in pariampullary adeno-carcinoma after adequate resection.[11]

Periampullary neoplasms and neuroendocrine tumors (NETs) exhibit significant differences, according to their origin and biological behavior. NETs like carcinoid tumor develop from neuroendocrine cells, which are located in several organs. NETs can be classified as "functioning" or "non-functioning." The latter are often asymptomatic and diagnosis is incidental unless mass effect and/or metastatic disease occurs, making surgical cure impossible.[12] In our case the carcinoid tumor of stomach was nonfunctioning and the findings were incidental.

The pathogenesis of synchronous presence of GI adenocarcinoma and NET like carcinoid tumor are yet to be discovered. But clinically most of the time the adenocarcinoma is expressed more than the carcinoid tumor because of it's slow progression. Most of the SPM cases, the NET component is found incidentally.[13] Our case showed similar findings.

The synchronous presence of a primary tumor in patients affected by a neuroendocrine tumor is reported in the literature but incidence is variable and the most common site is the gastrointestinal tract.[14] Diagnostic workup for periampullary neoplasms includes abdominal computed tomography (CT) scan, magnetic resonance imaging (MRI) , side view duodenoscopy and endoscopic ultrasound (EUS). In our case, these investigations revealed periampullary carcinoma. Later histopathology showed the presence of carcinoid tumor in the resected part of the stomach. As the tumor was small and non- functional, pre-operative work up could not detect the lesion.

Conclusion:

In this report, we present a rare case of periampullary carcinoma with asymptomatic synchronous carcinoid tumor incidentally detected by histopathology after the operation. Multiple primary tumors are seen synchronously and/or metachronously with neuroendocrine tumor, are an extremely rare phenomenon. These patients must be extensively evaluated for Synchronous primary malignancy during the workup and follow-up period. Further studies are required to clarify the mechanisms of carcinogenesis associated with neuroendocrine carcinomas and synchronous tumors.

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