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Case Report

Ampullary micropapillary adenocarcinoma widely metastatic to the lymph nodes: A case report

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ABSTRACT

First described in breast cancer, invasive micropapillary carcinoma is being increasingly recognized as an aggressive histological variant of cancers arising from many different organs such as the urinary bladder, the lung, the colorectum, the stomach, the pancreatobiliary system and the salivary glands. Only a few reports exist that focus on the ampullo-pancreatobiliary region.

We report a case of a 71 year old man who was admitted with symptoms of obstructive jaundice and slight epigastric pain and weight loss. Clinical and histological investigations revealed a small (< 2 cm) ampullary adenocarcinoma comprised of an invasive front showing a micropapillary pattern, mixed with a biliary type solid and cribriform component. Extensive lymphatic and lymph node involvement were present. The widespread lymph node metastases affected both staging categories N and M.

We believe micropapillary carcinoma should be considered in the WHO classification of Tumors of the Digestive System, in the ampullary chapter, as a separate histological subtype, in analogy with other organs to reflect its aggressive clinical behavior.

1. Introduction

The ampulla of Vater is a complex region composed of 3 histologically and physiologically distinct anatomic structures: the common bile duct (CBD), the pancreatic duct, and the duodenum, which adjoin the papilla of Vater [1].

Tumors of the ampulla of Vater may arise in the ampulla (intra-ampullary type), on the duodenal surface of the papilla (peri-ampullary type), or may involve both the intra-ampullary and peri-ampullary regions (mixed type). Thus, ampullary tumors may show biliary and/or intestinal features [2, 3]. A tumor is considered as a primary ampullary if its epicenter is located in the lumen or walls of the distal ends (intra-ampullary component) of the CBD and/or pancreatic duct, or at the "papilla of Vater", or the duodenal surface of the papilla [1].

The importance of defining the origin of a tumor is justified by the better prognosis of papillary adenocarcinomas when compared to pancreatic adenocarcinomas [4].

Micropapillary carcinoma arising from many different organs has been increasingly reported (urinary bladder, lung, breast, ovary, colorectum, stomach, organs constituting the ampullo-pancreatobiliary system and salivary glands) [5–21].

It has been associated with a propensity for lymphatic invasion, lymph node metastatic spread and poor prognosis, with the exception of ovarian tumors [16, 18].

Since this histological subtype clearly has shown distinct clinicopathological features, it is included as a separate entity in the WHO Classification series for tumors originating in the breast, lung, colorectal and urinary systems, where it has been most frequently reported.

In the gastro-intestinal tract, this variant was first reported in 2005.

Not one single case has been reported arising in the duodenum and only a few cases have been reported as arising in the stomach and the rest of the small bowel. In single series studies, the incidence of micropapillary differentiation has been reported in 4% of ampullo-pancreatobiliary carcinomas [9]. Whereas the reported incidence of colorectal cancer with micropapillary differentiation is higher, ranging from 9 to 19% [10, 22].

We describe a case of a small-sized ampullary adenocarcinoma (< 2 cm), with extensive lymphatic and lymph node involvement, which significantly affected staging categories.

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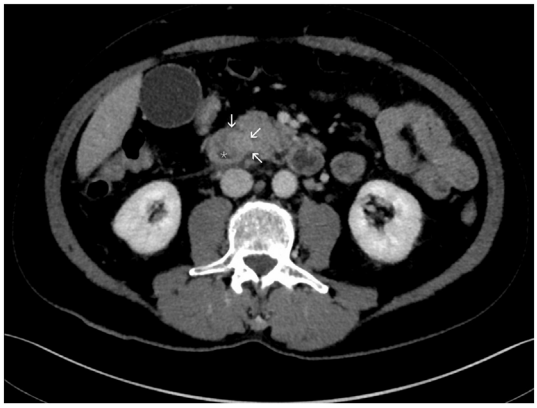

Fig. 1. The computed tomography scan shows an ampullary mass (arrows). The asterisk is in the duodenal lumen.

2. Case report

2.1. Clinical presentation

A 71 year old man was admitted with symptoms of obstructive jaundice, slight epigastric pain and weight loss (of 4 kg). A computed tomography scan of the abdomen was performed and showed a luminal expansion of the CBD due to an ampullary mass (Fig. 1), associated with a dilatation of both the biliary and pancreatic ducts.

A pylorus-preserving Whipple procedure took place the following week.

On follow-up, the patient died 11 months after surgery because of local recurrence of disease (retroperitoneal and peri-anastomotic) and systemic spread (pulmonary).

2.2. Pathology

During the surgical procedure, a frozen section examination of the proximal duodenal margin and the pancreatic neck margin were requested. Intraoperative pathological examination resulted in a tumor free pancreatic neck margin, but lymphangitic carcinomatosis was discovered in the duodenal margin. This led to the additional resection of the distal portion of the stomach.

Macroscopically, a large sessile polyp (5 × 6 cm) surrounding the papilla (Fig. 2) was present in the duodenum. Bivalve slicing of the pancreatic head showed a solid 1,9 cm mass, extending into the CBD (Fig. 3).

Upon histological examination, the duodenal sessile lesion proved to be a duodenal tubulo-villous adenoma, with predominant low grade dysplasia and a high grade dysplastic area transitioning into an adenocarcinoma, situated in the duodenal surface of the papilla (Fig. 4). The adenocarcinoma showed a predominant (60%) micropapillary architecture (Fig. 5).

A second component was observed in the CBD, namely a biliary type solid and cribiform adenocarcinoma (Figs. 6 and 7), which invaded the pancreatic parenchyma and the peri-pancreatic fatty tissue (focally). The cribiform areas showed central comedo-type necrosis (Fig. 7).

We observed an extensive peritumoral lymphatic invasion, as well as perineural and venous invasion. All peri-pancreatic lymph nodes examined contained metastatic deposits, including two situated in the retroperitoneal margin, arriving in contact with the inked surface of the margin.

A further aortocaval lymph node was sent separately. It also proved to be metastatic, thus affecting the M category, due to its non-regional location.

The final pathological staging according to the 8th edition AJCC Cancer Staging Manual [2] was pT3b N2 M1 G3 L1 V1 Pn1 R1.

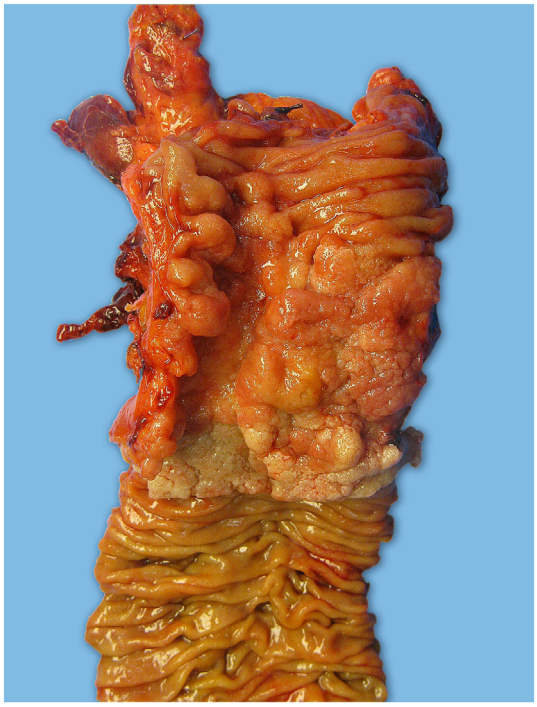

Fig. 2. Surgical specimen. Opening of the duodenum. A large periampullary sessile polyp is evident.

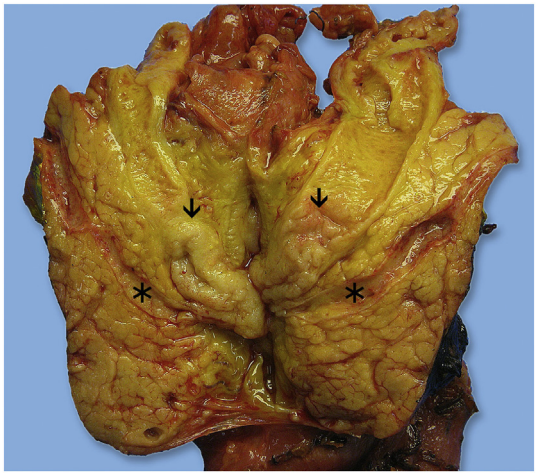

Fig. 3. Bivalve slicing of the pancreatic head shows a solid growth in the CBD (arrow). Asterisk is indicating the pancreatic duct.

Tests for mismatch repair deficiency were conducted by immunohistochemistry with the four antibodies MLH1, PMS2, MSH2 and MSH6. Tumor cells were positive, confirming no loss of those proteins.

For therapeutic reasons, we analyzed the expression of HER2 in the adenocarcinoma, as overexpression has been described in micropapillary tumors originating in the breast [23] and other organs, such as the urinary bladder [24],. The results were negative.

3. Discussion

Assessing the origin of a tumor involving the ampulla may be difficult, and occasionally impossible, especially in large tumors.

However, an effort to uncover said origin is of paramount importance, namely because of the different prognosis and different staging systems applied to duodenal, ampullary and pancreatic primary tumors [4].