

Case report

A rare case of Vater's ampulla adenocarcinoma - case report

Abstract

Introduction: Periapillary tumors account approximately 5% of all gastrointestinal cancers, and Vater's papilla. Vater's papilla tumor are the second most common entity of periampillary cancer.

Case report: We present a 69-year-old female patient who presents to the emergency room for jaundice, hyperchromic urine and involuntary weight loss, symptoms debuted three weeks ago. Laboratory tests showed hyperbilirubinemia with predominance of conjugated bilirubin and ultrasonography is suggestive of periampillary mass associated with dilated pancreatico-biliary tree. No abdominopelvic lymphadenopathy, no free peritoneal fluid on CT scan. ERCP (Endoscopic Retrograde Cholangio Pancreatography) visualizes a papillary apparatus with a budding, vegetative tumor with infiltration into duodenum. Exploratory laparotomy proceeds Whipple's pancreaticoduodenectomy done. Histopathology revealed Vater's papilla adenocarcinoma.

Conclusion: Pancreaticoduodenectomy represent standard surgical approach in ampillary carcinomas.

Keywords: *periampillary cancer; adenocarcinoma; ERCP; pancreaticoduodenectomy; Whipple procedure;*

1. Introduction:

Ampillary cancer of Vater's papilla is classified as periampillary cancer. Periapillary cancers account for 5% of all gastrointestinal cancers, whilst ampulla's cancer is rare (0.2% of the cases)[1]. Vater's papilla tumor are the second most common entity of periampillary cancers after pancreatic adenocarcinoma[2]. The ampillary region is a histologically and physiologically complex region where three different structures meet: the common bile duct (CBD), pancreatic duct (PD) and duodenum[3]. Cattel and Pyrtok first reported malignant transformation of an papilla adenoma[4]. In more than 70% of the investigated apillary carcinoma, tissue samples were found with severe dysplasia[5]. Clinical presentation includes vague abdominal pain, jaundice, recurrent pancreatitis, liver enzyme elevation, or uncommon symptoms such as gastrointestinal bleeding or duodenal obstruction[3].

Diagnostic imaging options include sonography, computed tomography (CT), magnetic resonance imaging (MRI), endoscopic retrograde cholangiopancreatography (ERCP), and

endoscopic ultrasound (EUS)[6]. Definitive diagnosis is made histologically after sampling (ERCP) or resection. Pancreaticoduodenectomy (Whipple procedure) is regarded as the standard treatment for ampullary cancers whereas endoscopic ampullectomy is typically reserved for benign ampullary lesions[3].

2. Case report

2.1 Clinical history

A 69-year-old overweight female patient (BMI: 27 kg/m²), known to have essential hypertension and hiatal hernia, presents to emergency room for jaundice, hyperchromic urine and involuntary weight loss (5 kg in the last month), debuted three weeks ago. He doesn't present abdominal pain, nausea or vomiting. Initially laboratory tests showed hyperbilirubinemia (TB; 13 mg/dL) with a predominance of conjugated bilirubin (DB; 11 mg/dL); increased gamma-glutamyl transferase (GGT; 421 U/L), alkaline phosphatase (ALP; 192 U/L), mild hepatic cytolysis (ALT; 88 U/L - AST; 74 U/L), mild leukocytosis (WBC; 103/ μ L), hypochromic and microcytic anemia (Hb; 8.9 g/dL) and increased acute phase protein (CRP; 34 mg/dL), (Fibrinogen; 419 mg/dL).

To rule out cholecystitis, cholelithiasis or choledocholithiasis, an abdominal ultrasound was performed which showed a solid lesion without a posterior shadow cone, without a Doppler signal. Intra and extrahepatic bile ducts have been dilated. Abdominal ultrasound was completed by a CT abdomen and pelvis that described an elongated and increased gallbladder (approx. 13/4.5 cm), infundibular cudate, without hyperdense stones. The intra- and extrahepatic bile duct system was dilated: dilated choledochus up to 2 cm, up to the level of the duodenal papilla, where a spontaneously hypodense mass is seen, iodophilic, relatively well delimited (axial diameter of about 1 cm), which determines the upstream dilatation of CBD (maximum diameter of 2 cm intrapancreatic) and Wirsung duct (0.5 cm) - which raises the suspicion of a Vaterian ampulloma. No abdominopelvic lymphadenopathy, no free peritoneal fluid. Radiological tumor staging was completed with a multi-detector CT of the thorax. No remote metastases were found.

Tumor markers were slightly elevated (CA 19-9; 42 U/ml), (CEA; 5 ng/ml). The ERCP (Endoscopic Retrograde Cholangio Pancreatography) visualizes a papillary apparatus with a budding, vegetative tumor formation and the biopsies reveal the papillary structure of the adenomatous type, with high-grade dysplasia.

2.2 Surgical treatment

Following the medical investigations and the specialized diagnosis, the operation is decided and practiced with the informed consent of the patient: Duodenopancreatectomy (Whipple procedure).

The surgical piece presents at the ampullary level, malignant tumor proliferation with well-differentiated ADK appearance with tubular pattern, associated chronic inflammation affecting the duodenal wall (pT2-N0-M0-G1) which includes stage IB neoplasia according to ampullary

tumors classification. Postoperative evolution is slowly favorable, with resumption of digestive tolerance and intestinal transit on the fourth day postoperative.

3. Discussion

Vater's ampulla cancers are one of the periampullary tumors with more favorable prognosis than others[7]. Higher resectability rate is associated to the better prognosis[8]. The prognostic factors for Vater's ampulla cancers following surgery have been reported to be jaundice, depth of tumor infiltration, pancreatic invasion lymph node metastasis, perineural invasion tumor and residual tumor status[3]. Preoperative biliary drainage in jaundiced patients was controversial. Patients with total bilirubin level higher than 5mg/dl just before surgery, demonstrated poorer survival in Korean study (Choi et al.) and patients without pancreatic invasion had significantly longer survival than those with pancreatic invasion[8]. Although the etiology of ampullary carcinomas is unknown in the majority of cases, several conditions have been associated with this malignancy, mostly in case reports or small series[9]. Familial adenomatous polyposis (FAP) is an important risk factor for the development of ampullary carcinoma[9].

Seventy-five percent of all ampullary neoplasms are adenocarcinomas, 20% are benign adenomas, and 5% are neuroendocrine tumors[9]. Adenocarcinomas account for 90% of ampullary malignancies; the rest include unusual types, such as mucinous, signet-ring cell, and undifferentiated carcinomas[9]. Histopathologically, 90% of ampullary adenocarcinoma can be classified into pancreaticobiliary or intestinal types[10]. Immunohistochemical analysis has shown high expression of CEA and CA 19-9 in the tumor[9]. Elevated serum concentrations of CEA and CA 19-9 have been detected in 11% to 29% and 41% to 63% of patients with ampullary carcinomas[11]. Elevations of these serum tumor markers have been associated with tumor recurrence and lower rates of disease-free survival in univariate but not multivariate analyses.

In most cases without infiltration or metastases, surgical resection is recommended-pancreaticoduodenectomy (Whipple procedure)[3].

Outcomes are good in the absence of lymph node metastases, with 5-year survival rates of 59% to 78%[12]. At this patient, considering the absence of lymphatic or other organs metastases (pT2-N0-M0-G1), it allowed to perform a pancreaticoduodenectomy with relative good prognostic.



Fig.1 ERCP – Periampullary tumor

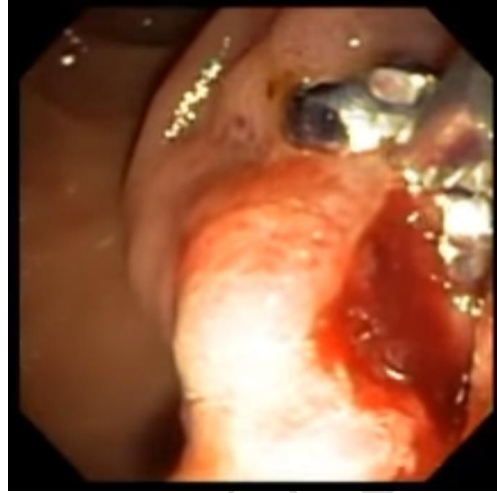


Fig.2 ERCP – Biopsy of tumor

4. Conclusion

Surgical resection is the only curative treatment for ampullary carcinoma and the standard surgical approach is pancreaticoduodenectomy. In absence of lymph node metastasis and any organ metastasis, prognosis is good at five years after surgery.

Abbreviations:

CT – Computer tomography
ERCP – Endoscopic retrograde cholangiopancreatography
CBD – Common bile duct
PD – Pancreatic duct
MRI – Magnetic resonance imaging
EUS – Endoscopic ultrasound
BMI – Body mass index
TB – Total bilirubin
DB – Direct bilirubin
GGT – Gamma-glutamyl transferase
ALT – Alanine aminotransferase
AST – Aspartate aminotransferase
CRP – C-Reactive Protein
CEA – Carcinoembryonic antigen
CA 19-9 – Carbohydrate antigen 19-9

Statements:

Consent for publication: As the corresponding author, I confirm that the manuscript has been read and approved for submission by all co-authors.

Informed Consent: The informed consent was obtained from the patient for publication and any accompanying images.

Statement of Ethics: The accompanying manuscript does not contain any studies carried out by the authors on humans or animals.

Ethical Approval: The treatment strategy/study protocol was approved by a local tumor board/ethics committee.

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