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A Rare Case of Adenocarcinoma of Ampulla of Vater – A Case Report

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Authors' contributions

This work was carried out in collaboration between both authors. Author CSG wrote the paper and provided the technical details, author LMN detailed the history of the patient and reviewed the paper. Author CSG reviewed and approved the final. Both authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Introduction: Periampullary tumors account approximately 5% of all gastrointestinal cancers, and Vater's papilla. Vater's papilla tumors are the second most common entity of periampullary cancer.
Case Presentation: We present a 69-year-old female patient who presented to the emergency room with symptoms including: jaundice, hyperchromic urine and involuntary weight loss, which she havd developed over the preceding three weeks ago. The results of her laboratory tests showed hyperbilirubinemia with predominance of conjugated bilirubin and she had ultrasound scan of her abdomen and pelvis which demonstrated features adjudged to be suggestive of periampullary mass associated with dilated pancreatico-biliary tree. No abdominopelvic lymphadenopathy, no free peritoneal fluid on computed tomography (CT) scan was demonstrated. She underwent Endoscopic Retrograde Cholangio-Pancreatography (ERCP) which revealed a papillary apparatus with a budding, vegetative tumor with infiltration into her duodenum. She underwent exploratory laparotomy and Whipple's pancreaticoduodenectomy. Histopathology examination of the excised surgical specimen revealed Vater's papilla adenocarcinoma. She would be undergoing regular follow-up assessments after she has been discharged from hospital to ascertain her progress following her pancreato-duodenectomy which we had regarded as her treatment of curative intent pursuant to our multi-disciplinary team approach to her treatment.

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Conclusions: 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, it would be envisaged that the patient would have a good five year survival pursuant to her surgery and she would have regular careful follow-up assessments with inclusion of clinical assessments, laboratory test assessments and radiology imaging assessments.

Keywords: Periampullary cancer; adenocarcinoma; ERCP; pancreaticoduodenectomy; whipple procedure.

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*

1. INTRODUCTION

"Ampullary cancer of Vater's papilla is classified as periampullary cancer. Periampullary cancers account for 5% of all gastrointestinal cancers, whilst ampullary cancer is rare and does account for 0.2% of the cases" [1]. "Vater's papilla tumors are the second most common entity of periampullary cancers after pancreatic adenocarcinoma" [2]. "The ampullary 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 Pyrtek first reported malignant transformation of an papilla adenoma [4]. In more than 70% of the investigated ampullary carcinoma, tissue samples were found with severe dysplasia [5]. "Clinical presentation of ampullary cancer includes vague abdominal pain, jaundice, recurrent pancreatitis, liver enzyme elevation, or uncommon symptoms such as gastrointestinal bleeding or duodenal obstruction" [6].

"Diagnostic radiology imaging options include: ultrasound scan, computed tomography (CT) scan, magnetic resonance imaging (MRI) scan, endoscopic retrograde cholangiopancreato-graphy (ERCP), and endoscopic ultrasound (EUS)" [7]. "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" [8].

Aim of study: To report a case of a case of adenocarcinoma of Ampulla of Vater

2. CASE PRESENTATION

2.1 Clinical History

A 69-year-old overweight female patient (BMI: 27 kg/m2), who was known to have essential hypertension and hiatus hernia, presented to the emergency room with jaundice, hyperchromic urine and involuntary weight loss (5 kg in the last month), which she had developed over the preceding three weeks. She did not report having abdominal pain, nausea or vomiting. Initially her laboratory test results 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/µL), 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, cholecystolithiasis or choledocholithiasis, an abdominal ultrasound scan was performed which showed a solid lesion without a posterior shadow cone, without a Doppler signal. Intra and extrahepatic bile ducts were noted to be dilated. She subsequently underwent a computed tomography (CT) scan of her abdomen and pelvis which was reported to have demonstrated an elongated and increased gallbladder (approx. 13/4.5 cm), infundibular cudate cancer, without hyperdense stones. The

intra- and extrahepatic bile duct system was reported to be dilated and dilated choledochus was noted that measured up to 2 cm, up to the level of the duodenal papilla, where a spontaneously hypodense mass was seen, iodophilic, relatively well delimited (axial diameter of about 1 cm), which determined the upstream dilatation of her common bile duct (CBD) with a maximum diameter of 2 cm intrapancreatic and Wirsung duct (0.5 cm) - which raised the suspicion of a Vaterian ampulloma. No abdominopelvic lymphadenopathy, and no free peritoneal fluid was found. Radiological tumor staging was completed with a multi-detector CT scan of the thorax. No remote metastases were found. Her tumor marker levels were slightly elevated (CA 19-9; 42 U/ml), (CEA; 5 ng/ml). She had Endoscopic Retrograde Cholangio Pancreatography (ERCP) which revealed a papillary apparatus with a budding, vegetative tumor formation and the biopsies of the tumor mass were undertaken and pathology examination of the biopsy specimens revealed a papillary structure with features of adenomatous type, with high-grade dysplasia.

2.2 Surgical Treatment

Following the medical investigations and the specialized diagnosis, the undertaking of operation was decided and this operation was subsequently undertaken, Duodenopancreatectomy (Whipple procedure) was therefore undertaken.

Pathology examination of the pancreato-duodenectomy specimen demonstrated at the ampullary level, malignant tumor proliferation with well-differentiated ADK appearance with tubular pattern, that was associated chronic inflammation affecting the duodenal wall and the tumor was staged pT2-N0-M0-G1 which included stage Ib neoplasia according to ampullary tumors classification. The patient postoperative recovered slowly with no untoward effects, with resumption of her digestive tolerance and intestinal transit on the fourth day postoperatively.

3. DISCUSSION

Vater's ampulla cancers are one of the periampullary tumors with more favorable prognosis than others [9]. It has been pointed out that higher resectability rate of the tumor is associated with the better prognosis [10]. The prognostic factors for Vaters ampulla cancers following surgery had been reported to include: jaundice, depth of tumor infiltration, pancreatic invasion lymph node metastasis, perineural invasion tumor and residual tumor status [11]. "The undertaking of preoperative biliary drainage in jaundiced patients was stated to be controversial" [12]. "Patients with total bilirubin level higher than 5 mg/dl just before surgery, demonstrated poorer survival in a Korean study (Choi et al.) and patients without pancreatic invasion had significantly longer survival than those with pancreatic invasion" [10]. Although the etiology of ampullary carcinomas was unknown in the majority of cases that had been reported, several conditions had been documented to be associated with this malignancy, mostly in case reports or small series [13]. "Familial adenomatous polyposis (FAP) is an important risk factor for the development of ampullary carcinoma" [14].

"Seventy-five percent of all ampullary neoplasms are adenocarcinomas, 20% are benign adenomas, and 5% are neuroendocrine tumors" [15]. ""Adenocarcinomas account for 90% of ampullary malignancies; the rest the tumors include unusual types, such as mucinous, signet-ring cell, and undifferentiated carcinomas" [16]. Histopathologically, 90% of ampullary adenocarcinoma can be classified into pancreaticobiliary or intestinal types [17]. Immunohistochemical analysis has shown high expression of CEA and CA 19-9 in the tumor [13]. Elevated serum concentrations of CEA and CA 19-9 had been detected in 11% to 29% and 41% to 63% of patients with ampullary carcinomas [18]. Elevations of these serum tumor markers had 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-and the recommended surgical resection should entail pancreaticoduodenectomy (Whipple procedure) [3].

Outcomes are good in the absence of lymph node metastases, with 5-year survival rates of 59% to 78% [19]. Considering the fact that our reported patient had absence of lymphatic metastasis and absence of metastasis anywhere else in the body and the tumor was staged pT2-N0-M0-G1, this enabled us to undertake a pancreaticoduodenectomy with the expectation that the patient would have a good prognosis.

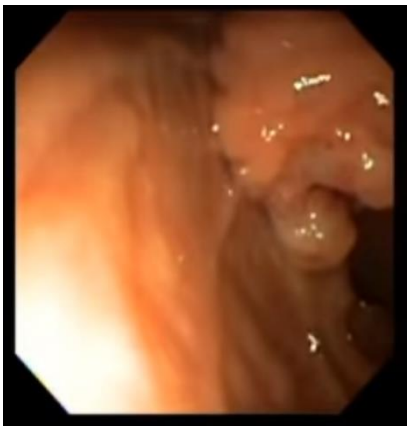

Fig. 1. ERCP – Periampullary tumor

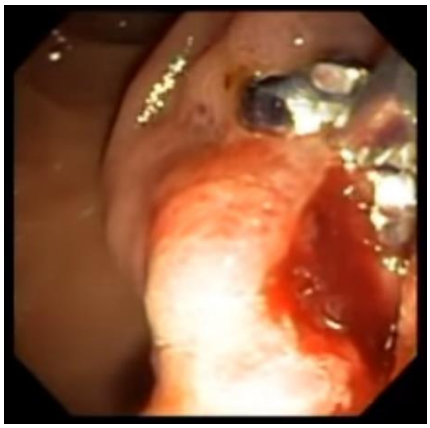

Fig. 2. ERCP – Biopsy of tumor

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, prognostic are good at five year after surgery.

CONSENT

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

ETHICAL APPROVAL

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

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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