

# Primary squamous cell carcinoma of the ampulla of Vater - a rare entity

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## Abstract

A 28-year-old female presented with jaundice, abdominal pain and postprandial vomiting. UGI endoscopic examination demonstrated a friable polypoid ampullary mass. CECT of the abdomen revealed a dilated distal part of the common bile duct and dilated main pancreatic duct with a soft-tissue mass in the ampulla. The clinical presentation, radiographic and endoscopic investigation pronounced ampullary carcinoma. We performed Whipple's pancreaticoduodenectomy with curative intention. Histopathological examination revealed squamous cell carcinoma of the ampulla of Vater. To the best of our knowledge, it is the second such case with successful surgical treatment.

## INTRODUCTION

Squamous cell carcinoma is a very rare form of cancer located in the ampulla of Vater that is derived from ductal cells. Most adenomas and carcinomas of the small intestine and extrahepatic bile ducts arise in the region of Vater's papilla. Histologically, intestinal type adenocarcinoma, pancreatobiliary type adenocarcinoma, undifferentiated carcinomas and unusual types can be found. Histological and immunohistochemical findings support the concept of histogenetically different ampullary carcinomas which develop from intestinal or pancreatobiliary type of mucosa of Vater's papilla. Molecular alterations in ampullary carcinomas should be correlated closely with the different tumour biology and clinical outcome [1]. A wide variety of neoplastic lesions (mostly adenomas and adenocarcinomas) may involve the ampulla of Vater, but squamous cell carcinoma has been reported only in two publications, on extensive review of indexed literature. One of these was a metastatic squamous cell carcinoma [2]. Thus, primary squamous cell carcinoma, derived from ductal cells, is a very rare entity. Here, we report a case of a 28-year-old female with primary squamous cell carcinoma of the ampulla of Vater. To the best of our knowledge it is second case to be reported worldwide and the first case from India.

## CASE REPORT

A 28-year-old female presented with abdominal pain, jaundice and postprandial vomiting for one month. She also had associated history of weight loss and anorexia. Clinical

examination revealed pallor, icterus and hepatomegaly with no peripheral lymphadenopathy. The laboratory examination revealed a serum CA19.9 of 41U/ml, a total bilirubin level of 4.0 mg/dL, an aspartate aminotransferase level of 86 IU/L, an alanine aminotransferase level of 100 IU/L and an alkaline phosphatase of 170U/L. UGI endoscopic examination demonstrated a friable polypoid ampullary mass causing luminal obstruction in the second part of the duodenum. Biopsy revealed a well-differentiated squamous cell carcinoma. Metastatic work-up did not reveal any other primary focus of disease. CECT of the abdomen revealed a dilated distal portion of the common bile duct and a dilated main pancreatic duct with a soft-tissue mass in the ampulla.

We performed Whipple's pancreaticoduodenectomy. The gross specimen revealed a growth of 3x3cm in the ampulla of Vater (Fig. 1).

**Figure 1**

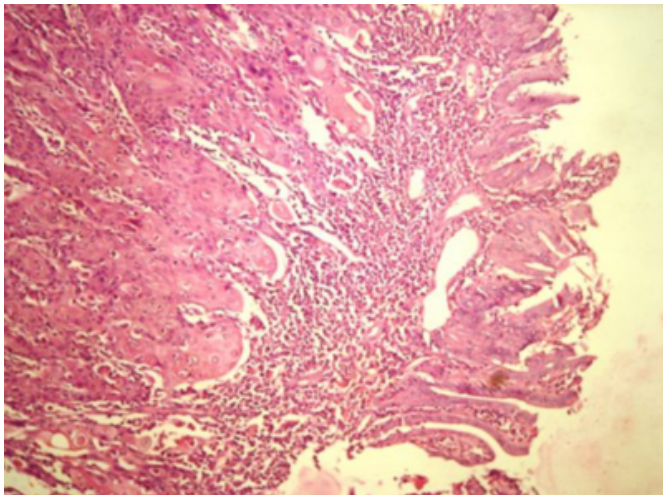
Fig. 1: Gross specimen showing growth in the ampulla of Vater



Histopathological examination revealed well-differentiated squamous cell carcinoma of the ampulla of Vater (Fig. 2).

**Figure 2**

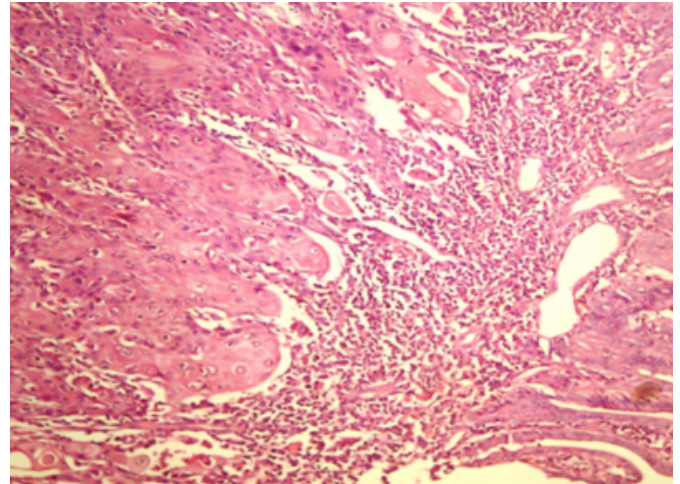
Fig. 2: Microscopic examination showing well differentiated squamous cell carcinoma of the ampulla of Vater (magnification 10X10x, H&E stain)



Sections from the common bile duct showed squamous metaplasia (Fig. 3).

**Figure 3**

Fig. 3: High-power view of CBD showing squamous metaplasia (10X40x, H&E stain)



On serial sectioning of the tumor specimen, no adenomatous component was found. There was no lymphovascular invasion and none of the twelve lymph nodes removed had metastases.

The postoperative course was uneventful and the patient was discharged on the ninth post-operative day.

## DISCUSSION

Carcinoma of the ampulla of Vater is defined as a malignant tumor arising in the last centimeter of the common bile duct as it passes through the wall of the duodenum and ampullary papilla. The pancreatic duct and common bile duct merge and exit by way of the ampulla into the duodenum. The ductal epithelium in these areas is columnar and resembles that of the lower common bile duct. Hence, 90% of ampullary tumors are adenocarcinomas. Neuroendocrine tumors, cystadenomas, and adenomas represent additional, but uncommon, histologic types. Squamous cell carcinoma is a very rare form of cancer located in the ampulla of Vater that is derived from ductal cells. Histological and immunohistochemical findings support the concept of histogenetically different ampullary carcinomas which develop from intestinal or pancreaticobiliary type of mucosa of Vater's papilla. Molecular alterations in ampullary carcinomas should be correlated closely with the different tumour biology and clinical outcome. As we know, a wide variety of neoplastic lesions may involve the ampulla of Vater, but primary squamous cell carcinoma has been reported only in one publication from Poland. [3]

In the group of patients with periampullary carcinomas treated by pancreaticoduodenectomy, those with ampullary

location are most likely to survive long term. As might be expected, patient outcome has in general been associated with tumour site, stage and type of surgical resection, but some authors have questioned the role of surgical resection for periampullary tumours, concluding that poor outcomes for patients after resection do not justify surgical intervention [4–8]. However, we agree that the surgical resection remains the cornerstone in the treatment of periampullary carcinomas [9].

In our patient the tumor was in the ampulla of Vater with negative margins of resection and all the twelve lymph nodes resected were negative. Microsections did not reveal any adenomatous component and the metastatic work-up did not show any other primary focus of disease. This primary squamous cell carcinoma might have arisen because of the malignant transformation of the pre-existing squamous metaplasia of the epithelium.

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