

CASE REPORT

Signet Ring Cell Carcinoma of the Vater's Ampulla

Jose Manuel Ramia, Alfonso Mansilla, Jesús Villar, Karim Muffak, Daniel Garrote,
Jose Antonio Ferron

Hepatobiliary and Liver Transplantation Unit, Department of Surgery, Hospital Virgen de las Nieves.
Granada, Spain

ABSTRACT

Context Most tumors affecting Vater's Ampulla are adenocarcinomas and other histological variants are less frequent. A review of the literature revealed only seven previously reported cases of signet ring cell carcinoma of the ampulla of Vater. The presence of this kind of tumor has no clear histological explanation. Two possible theories have been proposed: the presence of gastric heterotopia in the ampulla of Vater or the existence of a perivaterian duodenal heterotopia of ulcerous etiology as the origin of a signet ring cell tumor which secondarily invades the ampulla of Vater.

Case report We performed a pancreatoduodenectomy in a 67-year-old woman with a T2N0M0 ampulla tumor. A histologic study revealed a signet ring cell neoplasm.

Conclusion Etiology and survival of signet ring cell carcinoma of Vater's ampulla is not well-defined in the literature due to the extreme rarity of this disease. Duodenopancreatectomy with pylorus preservation is the treatment of choice.

histological variants exist but are less frequent. Signet ring cell carcinoma is more commonly found in the stomach than at other sites in the digestive system. An intense search of the literature revealed only seven previously reported cases of signet ring cell carcinoma of the ampulla of Vater [1, 2, 3, 4]. We present a new case and discuss its possible etiology.

CASE REPORT

We present the case of a 67-year-old woman with a history of arterial hypertension, cholecystectomy, sigmoidectomy for sigmoid adenocarcinoma (T3N0M0) in 1996, and a second operation for anastomotic recurrence in 1997. She was referred to our center three months later with constitutional syndrome, asthenia, anorexia and 10 kg weight loss. She had had jaundice, choluria, and acholia without abdominal pain for the previous four weeks. Upon examination, she presented with good general health, mucocutaneous jaundice, and the abdomen was tender to palpation in the epigastrium. Liver laboratory tests showed total bilirubin 7.50 g/dL (reference range: 0.1-1.0 g/dL), AST: 109 IU/L (reference range: 0-45 IU/L); ALT 117 IU/L (reference range: 0-45 IU/L), alkaline phosphatase 835 IU/L (reference range: 0-125 IU/L) and GGT 1,569 IU/L (reference range: 0-32 IU/L): The remaining analyses and the coagulation study were normal. Ultrasonography and abdominal CT showed a protruding mass in the papilla

INTRODUCTION

Ampulla of Vater cancers are usually well-differentiated adenocarcinomas. Other

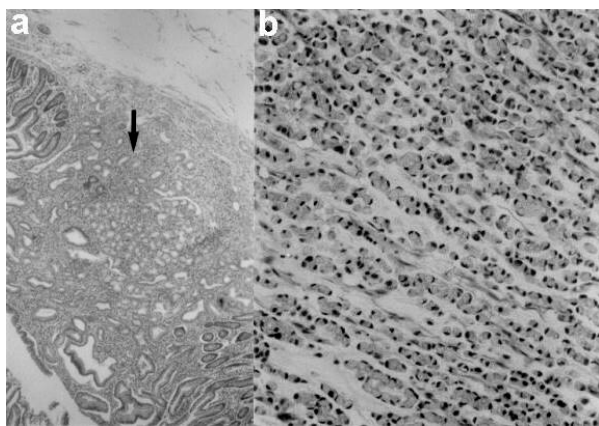


Figure 1. a. Gastric metaplasia (black arrow) in duodenal mucosa near the ampulla of Vater (H&E, x40). **b.** Signet ring cell ampullar cancer (H&E, x100)

compatible with a tumor in the ampulla and a dilatation of the extrahepatic bile duct; no locoregional lymph nodes were observed. ERCP confirmed the presence of a Vater's ampulla tumor. Biopsies were reported as signet ring cell carcinoma.

A cephalic duodenopancreatectomy was carried out. Postoperative delayed gastric emptying improved with erythromycin. The definitive histological study showed a neoplasm having a diameter of 1.8 cm with a congestive appearance. The histological sections showed small foci of gastric metaplasia in the duodenal mucosa near the ampulla (Figure 1a), which was infiltrated by a poorly differentiated signet ring cell adenocarcinoma (Figure 1b), with some foci of mucosecreting adenocarcinoma which infiltrated the ampullary wall, preserving the pancreatic parenchyma. None of the lymph nodes presented metastases (T2N0M0). Periodic acid-Schiff staining was positive and chromogranin was negative. The patient did not receive adjuvant therapy and remains alive and disease-free one year after surgery.

DISCUSSION

We present herein the eighth report in the literature of a signet ring cell carcinoma of the ampulla of Vater [1, 2, 3, 4]. Out of the seven patients previously reported, six were male [4]. The mean age of the published cases was 60.3 years (range: 25-72 years). Jaundice was

the most common symptom (5 out of 7 patients); the macroscopic appearance was superficial-protruding (4 cases), ulcerative (2 cases) or diffuse infiltrative (1 case) [4]. The treatment was duodenopancreatectomy in six patients and local excision in one high-risk surgical patient [1, 2, 3, 4]. None of the resected patients had lymph node involvement. According to the TNM classification, they were divided between T3N0M0 (5 cases), T2N0M0 (1 case), and TxNxM0 (1 case). The present patient had the typical characteristics of other cases in terms of age, jaundice, macroscopic appearance, and treatment approach, but differed in being female with a T2 tumor. Gardner *et al.* described a double-secreting amphicrine tumor with a large population of neuroendocrine cells [2]. In contrast, Arnal *et al.* reported a pure tumor solely formed by signet ring cells. In the present case there were only two populations, colloid cells and signet ring cells. The latter were not isolated cells, as in some colloid carcinomas, but constituted instead a large proportion of the tumor [1].

The presence of a signet ring cell tumor in the ampulla of Vater has no clear histological explanation. Two possible theories have been proposed to explain the histological variability:

1. The presence of gastric heterotopia in the ampulla of Vater is very rare [5]. Signet ring cell cancer - a common tumor in the gastric epithelium - can develop on the ampullary or periampullary ectopic gastric mucosa. The scant presence of ampullary gastric heterotopia accounts for the low number of ampullary signet cell tumors. The patient reported by Gardner *et al.* (2) and our patient both presented with areas of peritumoral ectopic gastric mucosa.

2. Areas of gastric-type epithelium have been observed in the duodenal bulb of patients with peptic ulcers. These acquired metaplastic areas lack glandular epithelium and are macroscopically normal, and it has been postulated that they are due to a protective response to elevated intraluminal acidity [6]. The presence of a perivaterian

duodenal heterotopia of ulcerous etiology may be the origin of a signet ring cell tumor which secondarily invades the ampulla of Vater.

Signet ring cell tumors localized in other digestive organs have a poor prognosis. The scant number of cases reported in the ampulla precludes any conclusions about survival associated with this histological variant [1, 2, 3, 4]. Interestingly, none of these cases have shown lymph node involvement, which is a determinant prognostic factor in cancer of the ampulla of Vater [4]. Duodeno-pancreatectomy with pylorus preservation is the treatment of choice in ampullar cancers [4]. Adjuvant therapy has not showed survival benefit in patients without lymph node infiltration.

Received August 19th, 2004 - Accepted September 14th, 2004

Keywords Ampulla of Vater; Carcinoma, Signet Ring Cell; Neoplasms; Pancreas; Surgery

Correspondence

Jose Manuel Ramia
C/Buensuceso 6, 4D

Granada 18014

Spain

Phone: +34-958.536.859

Fax: +34-958.020.680

E-mail address: jose_ramia@hotmail.com

References

1. Arnal Monreal FM, Lorenzo Patino MJ, Sacristan F, Ghanime Saide G. Carcinoma de células en anillo de sello de la ampolla de Vater. Rev Esp Enferm Dig 1994; 85:391-3. [PMID 8049111]
 2. Gardner HA, Matthews J, Ciano PS. A signet-ring cell carcinoma of the ampulla of Vater. Arch Pathol Lab Med 1990; 114:1071-2. [PMID 2171451]
 3. Casella R, Rittmann WW, Meier R, Wegmann W, Widmer MK, Hunger T. Signet ring cell carcinoma of Vater's papilla: a very rare malignancy. Helv Chir Acta 1994; 60:987-90. [PMID 7876027]
 4. Hara T, Kawashima H, Ishigooka M, Kashiyama M, Takanashi S, Hosokawa Y. Signet-ring cell carcinoma of the ampulla of Vater: a case report. Hepatogastroenterology 2002; 49:561-3. [PMID 11995497]
 5. Blundell CR, Kanun CS, Earnest DL. Biliary obstruction by heterotopic gastric mucosa at the ampulla of Vater. Am J Gastroenterol 1982; 77:111-4. [PMID 7072678]
 6. Hoedemaeker PJ. Heterotopic gastric mucosa in the duodenum. Digestion 1970; 3:165-73. [PMID 4915587]
-