Case Report

Signet ring carcinoma of ampulla of vater

Ardeshir Talebi, Fareshteh Mohammadizadeh, Mohsen Hani, Marzie Bagheri, Azam Bagheri

Department of Pathology, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran

Abstract

Signet ring carcinoma is a common type of adenocarcinoma of stomach but its occurrence in ampulla of Vater is extremely rare. There are only a few previous reported cases of signet ring carcinoma of ampulla of Vater. Here we reported a 61-year-old woman with obstructive jaundice. Ultrasonography and computed tomography (CT scan) examination showed intra- and extrahepatic bile duct dilatation. Endoscopic examination with biopsies revealed a small-size mass in ampulla of Vater with diagnosis of signet ring carcinoma. On consequent pancreatoduodenectomy the tumor was diagnosed as T2N0M0, stage IB. Because of the specific site of signet ring carcinoma of ampulla of Vater, the tumor seems to present itself at an early stage of disease. We review in the literature to suggest our idea.

Key Words: Ampulla of Vater, obstructive jaundice, signet ring carcinoma

Address for correspondence:
Dr. Mohsen Hani, Isfahan-Hakim Nezami Street, Khaghani Street, Gharakel Avenue, Nom 54, Isfahan, Iran. Phone number: 09131154653
E-mail: Mohsen_hani_t@yahoo.com
Received: 06.03.2013, **Accepted:** 15.06.2013

INTRODUCTION

Most Signet-ring cell carcinomas arise in stomach mucosa but it could appear in intestine and even in other organs than gastrointestinal tract. The World Health Organization (WHO) defines this tumor as a special type or a variant of gastrointestinal adenocarcinoma.[1] This tumor rarely has been happen in ampulla of Vater in pure form or in combination with usual type of adenocarcinoma. It seems that the prognosis of this tumor in ampulla of Vater is better than in stomach.[2] We reported our case of pure signet ring carcinoma of ampulla of Vater and review previously reported cases.

Access this article online	
Quick Response Code:	Website: www.advbiores.net
	DOI: 10.4103/2277-9175.124680

CASE REPORT

A 61-year-old woman was admitted to the Al Zahra hospital in Isfahan, Iran, in 2011 with occasional jaundice and generalized pruritic for 4 months; also, she complained of 7 kg weight loss during this period. She had no symptoms of nausea, vomiting, or abdominal pain. She had a past history of diabetes mellitus and blood hypertension. Physical examination was not contributory. In initial investigation, the laboratory test results demonstrated the rising level of glutamate-pyruvate transaminase, glutamic-oxaloacetic transaminase, alkaline phosphatase, γ-glutamyl transferase, and mild increase in total bilirubin and direct bilirubin. Ultrasonography indicted extra and intrahepatic biliary duct dilatation and increased common bile duct diameter. At endoscopic retrograde cholangiopancreatography (ERCP), the tumor of papilla of Vater was revealed and dilated intrahepatic and common bile duct with distal rat tailing was shown after dye injection [Figure 1]. Biopsies taken during ERCP were diagnosed as signet ring carcinoma. Subsequence CT scan indicated

How to cite this article: Talebi A, Mohammadizadeh F, Hani M, Bagheri M, Bagheri A. Signet ring carcinoma of ampulla of vater. Adv Biomed Res 2014;3:30.

intra- and extrahepatic ducts dilation, there were no obstructive mass or adenopathy. Liver, spleen, stomach, and pancreas were normal. At last the patient underwent a Whipple resection. On macroscopic examination there was polypoid mass at ampulla of Vater measured about 0.8 × 1 × 1 cm. pancreas and duodenum were normal. On microscopic examination the, malignant epithelial cells had intracytoplasmic mucin and individually infiltrating pattern that invaded to duodenal wall [Figure 2]. Regional lymph nodes were not involved. The ampullary carcinoma was diagnosed as T2N0M0, Stage IB according to the International Union Against cancer TNM classification (UICC).[3]

DISCUSSION

Signet ring carcinoma is a type of adenocarcinoma that has unique appearance because of discohesive pattern of proliferation and intracytoplasmic accumulation of mucin.[4] Most of Signet ring carcinoma arise from stomach (90% of all signet ring carcinoma) and is about 15% to 30% of all gastric cancer, but in other gasterointestinal tract like colon its occurrence is fewer and is about 1% of all cancer.[1,5,6] In stomach, signet ring carcinoma arise from epithelial cell of mucous neck or foveola. It has a tendency to develop in a younger age group (<45 year) than the intestinal-type adenocarcinoma and is associated with a poor prognosis.[6] Signet ring carcinoma rarely has been seen in the ampulla of Vater. Sekoguchi *et al*. was first described signet ring carcinoma mixed with adenocarcinoma of ampulla of Vater in 1979.[2] The first reported case of pure signet ring carcinoma of ampulla of Vater was in 1990 by Gardner *et al*.[7]

We review the reported cases of signet ring carcinoma of ampulla of Vater and summaries the data in Table 1. We found 20 previously reported cases in the English language literature.

The mean age of patients is about 69 years. The mean age of 16 patients with pure signet ring carcinoma is about 59 years, and mean age of three patients with mixed adenocarcinoma with signet ring carcinoma is 65 years, and so the mean age of affliction are similar to median age for large bowel (59-63.5 year) and are 15 years younger than signet ring carcinoma of the stomach (<45 yar).[5,6]

Presented symptoms were jaundice, weight loss, fever, nausea, vomiting, fatigue, abdominal pain, and abdominal distention.[8-11]

In four cases there was metastasis to liver, thoracic vertebra or lymph node, the case with liver metastasis had not jaundice until abdominal exploration.[11,12]

As for diagnosis of signet ring carcinoma of ampulla of Vater, CT scan and ultrasound only showed a dilated CBD without an apparent mass lesion in the ampulla of Vater, therefore these techniques could not reveal malignant etiology but some authors note that contrast-enhanced ultrasound may have effective diagnostic benefits for ampullary stenosis by dividing tumorous from nonmalignant stenosis.[5]

Macroscopic appearance was often superficial protruding,[13] ulcerative, or diffuse infiltrative.

Histologically, the origin of signet-ring cells may be as the following: (1) ectopic gastric mucosa, (2) gastric-type metaplasia, and (3) differentiated from common adenocarcinoma. These conclusions are due to accompaniment situations of the disease.[7,13] Some previous immunohistochemical findings suggest that all of three hypotheses may be true;

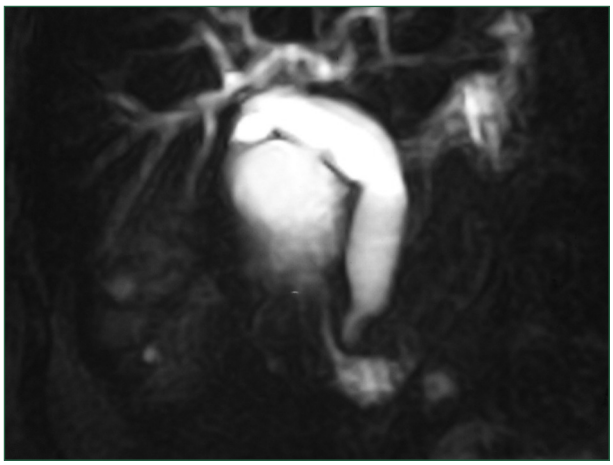

Figure 1: Dilated intrahepatic and common bile duct with distal rat tailing

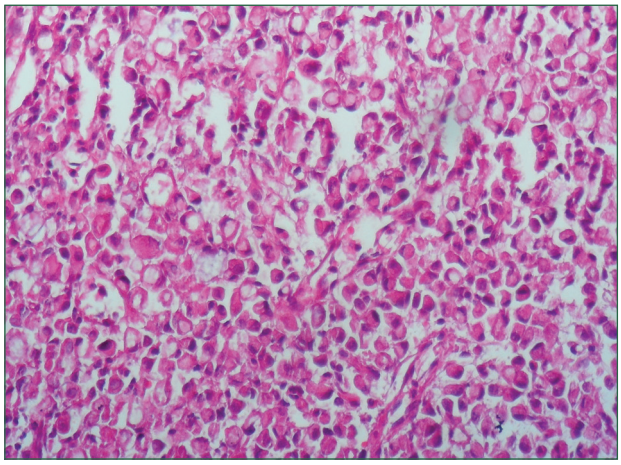

Figure 2: Signet ring cells with intracytoplasmic mucin

Table 1: Reported cases of signet ring carcinoma of ampulla of Vater

case	Age/sex	histopathology	TNM stage	procedure	Out come	Sign and symptoms
Gardner *et al.*[7]	69/F	Pure SRCC	T3N0M0 Stage IIA	Pancreaticoduodenectomy	Unknown	Unknown
Tseng *et al.*[22]	47/M	Pure SRCC	T3N0M0 Stage IIA	Pancreaticoduodenectomy	Alive 6 months	Unknown
Hara *et al.*[8]	68/M	Pure SRCC	T2N0M0 Stage IB	Pancreaticoduodenectomy	Alive 10 months	postprandial abdominal pain and nausea
Nabeshima *et al.*[12]	49/M	Pure SRCC	T3NxM1 Stage IV	NONE	Dead after 12 months	unknown M1= bone marow
Eriguchi *et al.*[9]	83/M	Pure SRCC	T3N0M0 Stage IIA	Pancreaticoduodenectomy	Alive 18 months	fatigue, fever, and obstructive jaundice
Ramia *et al.*[13]	67/F	SRCC with ADC	T2N0M0 Stage IB	Pancreaticoduodenectomy	Alive 12 months	asthenia, anorexia and 10 kg weight loss, jaundice, abdominal tenderness
Fang *et al.*[15]	53/M	Pure SRCC	T2N0M0 Stage IB	Pancreaticoduodenectomy	Alive 25 months	obstructive jaundice
Li *et al.*[6]	56/F	SRCC with ADC	T2N1M0 Stage IIB	Pancreaticoduodenectomy	Alive 12 months	pruritus, weight loss, and obstructive jaundice
Purohit *et al.*[11]	32/F	Pure SRCC in biopsy	TxNxM1 Stage IV	NONE	Unknown	vomiting, abdominal pain and distention, m1=liver
Bloomstone *et al.*[16]	58/F	Pure SRCC	T2N0M0 Stage IB	Pancreaticoduodenectom	Alive 134 months	painless
Akatsu *et al.*[17]	43/F	Pure SRCC	T2N0M0 Stage IB	Pancreaticoduodenectomy	Alive 90 months	pruritus and jaundice
Gao *et al.*[5]	38/F	Pure SRCC	T3N0M0 Stage IIA	Pancreaticoduodenectomy	Alive 6 months	pruritus, jaundice, nausea, 3 kg lost weight loss
Ishibashi *et al.*[1]	59/m	Pure SRCC	T3N0M0 Stage IIA	Pancreaticoduodenectomy	Dead 18 months	upper abdominal pain and icterus
Sekoguchi *et al.*[2]	Unknown	SRCC with ADSC	Unknown	Unknown	Unknown	Unknown
Maekawa *et al* [4]	72/M	SRCC with ADC	T3N0M0 stage IIA	Pancreaticoduodenectomy	Alive 6 months	jaundice
Arnal Monreal *et al.*[10]	Unknown	SRCC with endocrine cells	Unknown	Whipple's resection	disease free 24 months after	obstructive jaundice
Casella *et al.*[18]	70/M	Pure SRCC	Unknown	local transduodenal excision	asymptomatic 1year after	jaundice
Valeri *et al.*[19]	66/M	Pure SRCC	Unknown	pancreaticoduodenectomy	Unknown	Unknown
Tas *et al.*[20]		Pure SRCC	Unknown	Unknown	Unknown	Unknown
Garcia *et al.*[21]	73/M 74/M	2case of SRCC	T2N1M0 T3N0Mx	Pancreaticoduodenectomy pancreatectomy	disease free >14 months and 3 months	obstructive jaundice, weight loss
Hani *et al*	61/F	Pure SRCC	T2N0M0 Stage IB	Pancreaticoduodenectomy	Unknown	obstructive jaundice, weight loss

(ADC: Adenocarcinoma, SRCC: Signet ring cell carcinoma, M: Male, F: Female)

it seems that pure signet ring carcinoma is gastric type, that is, negative for CK 7 and positive for CK 20 and MUC 2, but mixed signet ring carcinoma and adenocarcinoma is pancreaticobiliary type, that is positive for CK 7 and negative for CK 20 and MUC 2.[4] The later type is the similar CK pattern to case of extra hepatic bile duct Poorly Differentiated Adenocarcinoma with Signet-ring Cell Carcinoma.[14]

The common type of treatment is Pancreaticoduodenectomy, if the patients have discovered at early stage of carcinoma of the ampulla of Vater, a pyloric-preserving pancreaticoduodenectomy is better to be done because the preservation of the whole stomach and pyloric ring may reduce postoperative complications, without decreasing curability.[1,9] There is no standard postoperative adjuvant therapy regimen for patients with this tumor. Nabeshima *et al*. reported the effectiveness of 5-fluorouracil and leucovorin in increasing the survival time with a good quality of life.[1,12]

For our patient the whipple surgery was done but without pyloric - preserving procedure because of unknown stage of tumor.

The prognosis of signet ring carcinoma of ampulla of Vater is better than stomach and discover in the earlier stage due to precocious jaundice of disease. Of 21 patients with signet ring carcinoma of ampulla of Vater, seven cases were at stage IB and six cases were at stage IIA. The 5-year survival rate for gastric signet ring carcinoma is

16.2%. Although the previous cases did not have long-term follow up but findings are suggested that signet ring carcinoma of ampulla of Vater has better prognosis.[1]

CONCLUSIONS

The signet ring carcinoma rarely occurs in ampulla of Vater but it should be in different possible diagnosis for the patients with obstructive jaundice. Despite the insidious pattern of development and poor outcome that this tumor shows in stomach but this tumor present itself at the early stage of disease in ampulla of Vater that may be due to its specific site and early symptom of jaundice.

REFERENCES

1. Ishibashi Y, Ito Y, Omori K, Wakabayashi K. Signet ring cell carcinoma of the ampulla of Vater. A case report. JOP 2009;10:690-3.
2. Sekoguchi T, Mizumoto R. Clinicopathological study of papilla of Vater. Geka Chiryo 1979;41:1-5.
3. Greene FL, Page DL, Fleming ID, Fritz AG, Balch CM, Haller DG, *et al*. AJCC cancer staging handbook: TNM classification of malignant tumors. 6th ed. New York: Springer-Verlag; 2002. p. 171-7.
4. Maekawa H, Sakurada M, Orita H, Sato K. Signet-Ring cell carcinoma co-existing with adenocarcinoma of the ampulla of Vater. A case report. JOP 2011;12:162-6.
5. Gao JM, Tang SS, Fu W, Fan R. Signet-ring cell carcinoma of ampulla of Vater: Contrast-enhanced ultrasound findings. World J Gastroenterol 2009;15:888-91.
6. Li L, Chen QH, Sullivan JD, Breuer FU. Signet- ring cell carcinoma of the ampulla of Vater. Ann Clin Lab Sci 2004;34:471-5.
7. Gardner HA, Matthews J, Ciano PS. A signet-ring cell carcinoma of the ampulla of Vater. Arch Pathol Lab Med 1990;114:1071-2.
8. 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.
9. Eriguchi N, Aoyagi S, Jimi A. Signet-ring cell carcinoma of the ampulla of Vater: Report of a case. Surg Today 2003;33:467-9.
10. Arnal Monreal FM, Lorenzo Patiño MJ, Sacristán F, Ghanimé Saide G. Signet ring cell carcinoma of the Vater's ampulla. Rev Esp Enferm Dig 1994;85:391-3.
11. Purohit RC, Kant K, Bhargava N, Kothari N, Purohit V. Signet ring cell carcinoma of ampulla of Vater in a young adult. Indian J Gastroenterol 2005;24:222-3.
12. Nabeshima S, Kishihara Y, Nabeshima A, Yamaga S, Kinjo M, Kashiwagi S, *et al*. Poorly differentiated adenocarcinoma with signet-ring-cells of the Vater's ampulla, without jaundice but with disseminated carcinomatosis. Fukuoka Igaku Zasshi 2003;94:235-40.
13. Ramia JM, Mansilla A, Villar J, Muffak K, Garrote D, Ferron JA. Signet-ring-cell carcinoma of the Vater's ampulla. JOP 2004;5:495-7.
14. Ogata S, Kimura A, Hatsuse K, Yamamoto J, Shimazaki H, Nakanishi K, *et al*. Poorly differentiated adenocarcinoma with signet-ring cell carcinoma of the extrahepatic bile duct in a 42-year-old Japanese female: A case report. Acta Med 2010;64:63-5.
15. Fang CL, Chu JS, Hsieh MC, Wu MS. Signet-ring cell carcinoma of the ampulla of Vater. J Formos Med Assoc 2004;103:793-6.
16. Bloomston M, Walker M, Frankel WL. Radical resection in signet ring carcinoma of the ampulla of Vater: Report of an 11-year survivor. Am Surg 2006;72:193-5.
17. Akatsu T, Aiura K, Takahashi S, Kaneyama K, Kitajima M, Kitagawa Y. Signet-ring cell carcinoma of the ampulla of Vater: Report of a case. Surg Today 2007;37:1110-4.
18. 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.
19. Valeri S, Caricato M, Ripetti V, Crucitti P, Ausania F, Garberini A, *et al*. Signet-ring cell carcinoma of the Vater`s ampulla: Report of a clinical case. Suppl Tumori 2005;4:S61.
20. Tas A, Ozer E, Koklu S, Kocak E. Signet ring cell carcinoma of the ampulla of Vater: A rare cause of acute pancreatitis. Scand J Gastroenterol 2011;46:126-7.
21. García AB, Arranz EM, Sanz RR, Serrano EM, Arranz MD, Sanz-Agero PG, *et al*. Signet ring cell carcinoma of the ampulla of Vater. Gastroenterol Hepatol 2011;34:141-6.
22. Tseng LJ, Jao YT, Mo LR. Signet ring cell carcinoma of major papilla. Gastrointest Endosc 2002;56:733.

Source of Support: Nil, **Conflict of Interest:** None declared.