1130-0108/2012/104/9/501-502
Revista Española de Enfermedades Digestivas

Rev Esp Enferm Dig (Madrid)
Vol. 104, N.° 9, pp. 501-502, 2012

Letters to the Editor

Signet ring cell adenocarcinoma of the ampulla of Vater: A rare pathology

Key words: Ampulla of Vater. Signet ring cell. Adenocarcinoma.

Dear Editor,

Tumors of the ampulla of Vater are uncommon, representing 0.2% of malignant tumors of the gastrointestinal tract and 8% of malignant tumors of the biliopancreatic area. They arise from the intestinal mucosa of the ampulla of Vater, being adenocarcinoma the most frequent (99% of cases). However, the signet-ring cells subtype at this location is a very rare variant, with only 20 cases reported in the literature (1-9).

Case report

We report the case of a 78-year-old woman with painless jaundice of recent onset. The abdominal CT showed bile duct dilatation and, after the performance of EUS and ERCP with biopsy, an adenocarcinoma of the ampulla of Vater was diagnosed.

A cephalic pancreaticoduodenectomy (CPD) was then performed with Whipple reconstruction, with pancreatico-jejunal anastomosis made using polypropylene mesh following the technique described by Wang (10). The postoperative followup was uneventful. The resected specimen confirmed the presence of a mucosecretor adenocarcinoma with areas of signet-ring cells at the ampulla of Vater, with diffuse growth pattern of 1.1 cm in diameter infiltrating the wall to the muscle layer without pancreatic involvement. Only 1 of 21 lymph nodes had neoplastic infiltration. The patient received adjuvant chemotherapy with gemcitabine and after 14 months follow-up remains free of disease.

Discussion

Signet ring cell adenocarcinoma is a rare variant of gastrointestinal adenocarcinoma, in which the intracellular mucin displaces the nucleus of tumor cells to the periphery. The stomach is the most frequent localization (90%), with generally more aggressive behavior and worse prognosis associated. Other locations of this disease include the breast, gallbladder, pancreas or colon. The development of signet ring cells carcinoma at the level of the ampulla of Vater is truly exceptional. Since the first description made in 1979 by Sekoguchi (1), the cases reported in the literature are limited to 20 (Table I).

Surgical resection is the only curative treatment, being the pancreatoduodenectomy or the ampullectomy the most common options. There are several factors to consider before the choice of treatment, mainly the tumor stage and the surgical and anesthetic risk. Some authors advocate in all cases for the performance of a CPD because of the risk of lymph node spread that even in T1 stage can reach up to 10%.

Because of the few cases described of this tumor it can not be defined if it is associated with more aggressive behavior, which would be a factor to take into account to perform a CPD resection. In the 20 cases reported to date, only 1 of them was treated by ampullectomy with good results, although with a short follow-up period of only 12 months (3).

Several prospective studies have shown an improved survival in patients resected with curative criteria, which are treated with adjuvant chemotherapy, usually employing gemcitabine and 5-fluorouracil.

Lucía Lesquereux-Martínez, Aquilino Fernández-Pérez and Manuel Bustamante-Montalvo

Department of Gereral Surgery and Digestive Diseases. Hospital Clínico Universitario de Santiago de Compostela, A Coruña. Spain

Table I. Published cases of signet-ring cell adenocarcionoma of the ampulla of Vater

Author	*Year*	*Age*	*Sex*	*Size*	*TNM*	*Stage*	*Surgery*	*Follow up (months)*	*Results*
Sekoguchi	1979	47	Male	2.0 x 0.8	T3N0M0	IIA	Unknown	Unknown	DFS
Gardner	1990	69	Female	2.0 x 1.5	T3N0M0	IIA	CPD	Unknown	DFS
Arnal-Monreal	1994	71	Male	2.5	T2N0M0	IB	CPD	24	DFS
Casella	1994	70	Male	Unknown	TxNxM0		Ampulectomy	12	DFS
Tseng	2002	47	Male	2.0	T3N0M0	IIA	CPD	6	DFS
Hara	2002	68	Male	1.5 x 0.8	T2N0M0	IB	CPD	10	DFS
Nabeshima	2003	49	Male	0.8	T3NxM1	IV	No	12	Died
Eriguchi	2003	83	Male	1.5 x 1.2	T3N0M0	IIA	CPD	18	DFS
Ramia	2004	67	Female	1.8	T2N0M0	IB	CPD	12	DFS
Fang	2004	53	Male	Unknown	T2N0M0	IB	CPD	25	DFS
Li	2004	56	Female	1.5 x 1.0	T2N1M0	IIB	CPD	12	DFS
Purohit	2005	18	Female	Unknown	TxNxM1	IV	No	Unknown	Unknown
Bloomston	2005	58	Female	1.0 x 0.8	T2N0M0	IB	CPD	134	DFS
Valeri	2005	66	Male				CPD		
Akatsu	2007	43	Female	2.0 x 1.8	T2N0M0	IB	CPD	90	DFS
Gao	2009	38	Female		T3N0M0	IIA	CPD	6	DFS
Ishibashi	2009	59	Male		T3N0M0	IIA	CPD	18	Died
Tas	2011								
Burgos-García	2011	73	Male	2.1x1.5	T2N1M0	IIB	CPD	14	DFS
Burgos-García	2011	74	Male	Unknown	T3N0Mx	IIA	Total Pancreatectomy	3	DFS

DFS: disease free survival. CPD: cephalic pancreaticoduodenectomy.

References

1. Sekoguchi T, Mizumoto R. Clinicopathological study of papilla of Vater. Geka Chiryo 1979;41:1-5.
2. Arnal Monreal FM, Lorenzo Patiño MJ, Sacristán F, Ghanimé Saide G. Carcinoma de celulas en anillo de sello de ampolla de Vater. Rev Esp Enferm Dig 1994;85:391-3.
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.
4. Tseng LJ, Jao YT, Mo LR. Signet ring cell carcinoma of major papilla. Gastrointest Endosc 2002;56:733.
5. 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.
6. 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.
7. 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.
8. Taş A, Ozer E, Köklü 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.
9. Burgos García A, Martín Arranz E, Rey Sanz R, Marín Serrano E, Martín Arranz MD, González Sanz-Agero P, et al. Carcinoma de células en anillo de sello de la ampolla de Vater. Gastroenterol Hepatol 2011; 34:141-6.
10. Wang X, Zhou W, Xin Y, Huang D, Mou Y, Cai X. A new technique of polypropylene mesh-reinforced pancreaticojejunostomy. Am J Surg 2007;194:413-5.