Case Report

Periampullary Carcinoma-A Rare Histopathological Variant

Fazl QP, Mozzain IK, Shakeel ur RK, Mubashir AS, Sajjad AD, Abdul Rashid L, Zubaida R, Zeba C

Department of Surgery, Sher-i-Kashmir Institute of Medical Sciences, Soura, Srinagar-190011, Jammu and Kashmir, India.

Abstract

Signet ring cell adenocarcinomas may be encountered in various parts of gastrointestinal tract but are almost unheard of in the region of ampulla of vater. In the region of ampulla of vater even if we come across an adenocarcinoma, it is usually a well differentiated variant. A 56-year-old female with features of obstructive jaundice on evaluation was diagnosed to have a peri-ampullary carcinoma.The patient was subjected to a laparotomy. On exploration patient was found to have a malrotation of gut. Patient was subjected to a pylorus preserving pancreatico-duodenectomy. Histopathology of the resected specimen revealed a signet ring morphology which is a rare variant in periampullary region.

Keywords: Periampullary, ampulla, adenocarcinoma, malrotation, pancreas

Correspondence:

Fazl Q Parray, Department of Surgery, Sher-i-Kashmir Institute of Medical Sciences, Soura, Srinagar-190011, Jammu and Kashmir, India. Tel: +91 09419008550 Fax: +91 01942403470 Email: fazlparray@rediffmail.com / fazlparray@gmail.com.

Date of submission: 28 Sept, 2014 Date of acceptance: 28 Nov, 2015

Introduction

Periampullary carcinoma is used to define a heterogeneous group of neoplasms arising from the head of the pancreas, the distal common bile duct and the duodenum. Ampullary carcinoma tumor on the other hand is centered in the region of the ampulla of Vater, which is formed by; the ampulla (common channel), the intraduodenal portion of the bile duct and the intraduodenal portion of the pancreatic duct. Hence, it may show intestinal and pancreatobiliary morphology. The unequivocable establishment of ampullary origin is possible in small lesions applying strict topographical criteria obtained at gross and histological examination. Rarely ampulla of vater is known to harbour various types of malignant tumors and most of them are usually well-differentiated adenocarcinomas. Even though as surgeons we frequently come across signet ring cell carcinoma (SRCC) in some parts of gastrointestinal tract but the very occurrence of this histological variant is quite rare at ampulla of vater (1). After a thorough search on pubmed and google we could find that till date only 30 such cases are reported in literature.

Case Report

A 56-year-old female presented with chief complaints of jaundice since two months. It was gradual in onset, progressively deepening in nature, not associated with pain. It was associated with mild generalised itching. The patient was also suffering from hypertension and hypothyroidism for which she was already on medication. Physical examination revealed significant pallor, icterus and hepatomegaly.

Liver chemistry tests showed raised bilirubin and alkaline phosphatase levels. Patient was evaluated and an USG abdomen was done which showed dilated CBD about 1cm after which ERCP was performed which showed a periampullary growth and stenting was done. No metastasis was found on further imaging procedures.

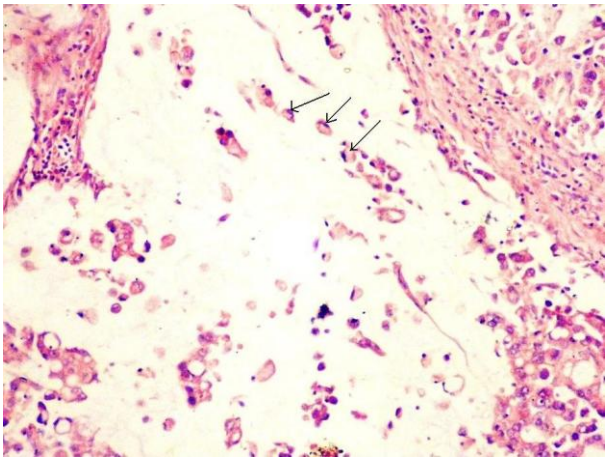

Figure 1: Signet ring cells in mucinous background - H&E (20X)

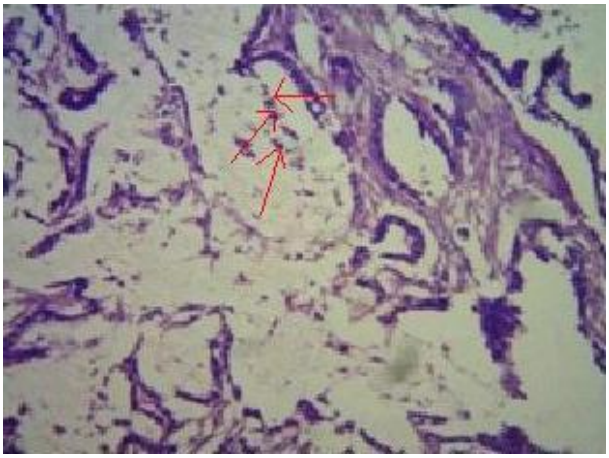

Figure 2: Signet ring ampullary carcinoma – H&E (10X)

The patient on exploration was found to have malrotation of gut. Patient underwent a pylorus-preserving pancreaticoduodenectomy. Intra operative findings revealed about 2x3cm firm mass in periampullary region with desmoplastic changes and enlargement of periportal lymph nodes.

On histopathological examination of the surgical specimen adenocarcinoma of signet cell morphology (Fig. 1, 2) was found involving the periampullary region which had infiltrated through the serosa, not infiltrating the underlying pancreas or CBD. One of the five removed lymph nodes were involved. However, the tumor margins were negative. Tumour staging was T2N1M0.

Patient is on adjuvant treatment at present and is on our follow up; doing well with no active complains and has been relieved of all symptoms.

Discussion

Ampulla of vater is rarely known to harbour a malignancy and even though occasionally it does get involved with adenocarcinoma but the incidence is < 6 cases per million per year (2). In periampullary tumors it is seen hardly in 6% and comprises only .2% of all gastrointestinal cancers (3,4). SRCC, a rare variant of adenocarcinoma, we usually come across in stomach, colon, rectum and even at times in breast, urinary bladder, pancreas and gall bladder. It is considered to be one of the worst cancers and is associated with a poor prognosis (5,6).

SRCC of ampulla of vater has been first of all reported in literature in 1979 by Sekoguchi and Mizumoto (7) followed by Gardner et al. (8) in 1990. SRCC in light microscopy is characterized by the presence of more than 50% characteristic signet ring cells. These cells an eccentrically located, crescent shaped nuclei with intracytoplasmic mucin (4,9). Where from these cells originate is still a controversial issue. Some believe these cells originate from congenital heterotrophic gastric mucosa whileas others believe that the site of origin for such cancers is from areas of gastric type metaplastic epithelia (10).

Since, SRCC is quite a rare entity in ampulla of vater, we thought it worth reporting and sharing it with our fellow community. After a thorough search on pubmed, google, google scholar, pubget, we could only retrieve 30 cases till date reported in literature. These cases are in the age group of 32-83 years. Amongst the reported cases, one patient presented with a metastatic disease after radiological evaluation whereas one more patient presented with T4 disease. In reported cases 10 patients were in T3 stage and 8 patients in T2 stage (11). In one patient multiple pulmonary deposits were seen (12) while as another presented with disseminated carcinamatosis (13). One patient during the course of adjuvant treatment following surgery presented with leptomeningeal metastasis (14).

Experience from previously reported cases suggests good prognosis in localised disease till the first postoperative year if a Classical Whipple's resection (15), local transduodenal excision (16) and pylorus-preserving pancreatoduodenectomy (17) have been done. We performed pylorus-preserving pancreatico-duodenectomy on our patient and she is at present on the follow up of our department and department of medical oncology for last 11 months after completing her adjuvant treatment.

Even though these patients are routinely given adjuvant chemotheraphy especially in a node positive disease but its role in a localised or metastatic disease is still debatable. A good evidence in support of adjuvant treatment is still lacking (18). Chemotherapy with 5-flurouracil and leucovorin have been have been reported to increase survival and QOL (13).

However, in the present case, in view of no metastasis found in imaging procedures, a surgical resection was done with pylorus preserving pancreatodudonectomy. Keeping in view her nodal involvement after a thorough discussion in tumor board the patient was put on adjuvant treatment and is showing a good recovery till date and continues to follow up with us.

Conclusion

It is very important for all surgeons to follow the histopathology of all operative specimens in order to frame a definitive diagnosis, prognosticates a patient and give the appropriate advice about the follow up treatment. Histopathological surprises may altogether alter the pre-planned management of the patient.

References

1. Gao JM, Tang SS, Fu W, Fan R. Signet-ring cell carcinoma of the ampulla of Vater: contrast-enhanced ultrasound findings. World J Gastroenterol 2009; 15(7): 888-91.

2. Li L, Chen QH, Sullivan JD, Breuer FU. Signet-ring cell carcinoma of the ampulla of Vater. Ann of Clin Lab Sci 2004; 34(4): 471-5.

3. Howe JR, Klimstra DS, Moccia RD, Conlon KC, Brennan M. Factors predictive of survival in ampullary carcinoma. Ann Surg 1998; 228(1): 87-94.

4. Albores-Saavedra J, Henson DE, Klimstra DS. Tumors of the gallbladder, extrahepatic bile ducts, and ampulla of Vater. In: Rosai J, Sobin L, eds. Atlas of Tumor Pathology. 3rd series, Fascicle 27. Washington DC: Armed Forces Institute of Pathology, 2000, pp-259-316.

5. Yokota T, Kunii Y, Teshima S, et al. Signet ring cell carcinoma of the stomach: a clinicopathological comparison with the other histological types. Tohoku J Exp Med 1998; 186(2): 121-30.

6. Kim JP, Kim SC, Yang HK .Prognostic significance of signet ring cell carcinoma of the stomach. Surg Oncol 1994; 3(4): 221-7.

7. Sekoguchi T, Mizumoto R. Clinicopathological study of papilla of Vater. Geka Chiryo 1979; 41: 1-5.

8. Gardner HA, Matthews J, Ciano PS. A signet-ring cell carcinoma of the ampulla of Vater. Arch Pathol Lab Med 1990; 114(10): 1071-2.

9. Lewin KJ, Appelman HD. Carcinoma of the Stomach. Tumors of the Esophagus and Stomach In: Rosai J, eds. Atlas of Tumor Pathology. 3rd series, Fascicle 18. Washington, DC: Armed Forces Institute of Pathology, 1996, pp-205, 209, 287-291.

10. Ramia JM, Mansilla A, Villar J, Muffak K, Garrote D, Ferron JA. Signet-ring-cell carcinoma of the Vater's ampulla. JOP 2004; 5(6): 495-7.

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(5): 222-3.

12. Taş A, Ozer E, Köklü S, Kocak E. Signet ring cell carcinoma of the ampulla of vater: rare cause of acute pancreatitis. Scand J Gastroenterol 2011; 46(1): 126-7.

13. Nabeshima S, Kishihara Y, Nabeshima A, 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(7): 235-40.

14. Paplomata E, Wilfong L. Signet ring cell carcinoma of the ampulla of Vater with leptomeningeal metastases: a case report. J Clin Oncol 2011; 29(21): e627-9.

15. Arnal Monreal FM, Lorenzo Patino MJ, Sacristan F, Ghanimé Saide G. Signet ring cell carcinoma of the Vater's ampulla. Rev Esp Enferm Dig 1994; 85(5): 391-3.

16. 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(6): 987-90.

17. Eriguchi N, Aoyagi S, Jimi A. Signet-ring cell carcinoma of the ampulla of Vater: report of a case. Surg Today 2003; 33(6): 467-9.

18. Acharya MN, Panagiotopoulos N, Cohen P, Ahmad R, Jiao LR. Poorly-differentiated signet-ring cell carcinoma of the ampulla of vater: report of a rare malignancy. JOP 2013; 14(2): 190-4.