CASE REPORT

A Rare case of Poorly Differentiated Adenocarcinoma (Signet Ring Cell type) of Duodenum- Nonampullary Region: A Case Report

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ABSTRACT

Signet Ring Cell type Adenocarcinoma (SRCC) of Duodenum is a rare pathological entity to occur and most of these tumors which have been reported in surgical literature known to occur in ampullary region of second part of duodenum. These are extremely rare to occur in non-ampullary regions of duodenum and only few cases have been reported in English literature so far across the world. We are submitting a rare case report of an elderly lady who presented to our institution with clinical profile consistent with gastric outlet obstruction. An esophagogastroduodenoscopy revealed a circumferential growth at D1-D2 junction with deep ulcer. Histopathological examination from biopsied specimen revealed poorly differentiated adenocarcinoma-Signet Ring cell type. On CT imaging she was found to have localized resectable disease and subsequently underwent standard Whipple's pancreaticoduodenectomy. She had uneventful post-operative recovery. Final Histopathology confirmed poorly differentiated adenocarcinoma -signet ring cell type. We conclude that SRCC nonampullary region remains extremely rare entity. These tumors are usually diagnosed with advanced disease on presentation and can be managed well with surgical resection, by Whipples Pancreaticoduodenectomy in majority, which are localized.

Keywords: Poorly Differentiated Adenocarcinoma, Signet Ring Cell type, Duodenum, Nonampullary Region.

INTRODUCTION

Primary Duodenal cancer is a rare entity comprises 0.3-0.5% tumour of all gastrointestinal cancer.[1,2]Biologically most duodenal tumours are mucin producing adenocarcinoma of various grades.[3] Very Few case reports of signet ring cell carcinoma(SRCC) in the duodenum have been reported in surgical literature, majority of them describe occurrence of this rare variety in ampullary region.[4-6] SRC is even rare to occur in nonampullary region of

duodenum such as second part of duodenum or duodenal bulb. So far only 5 cases of nonampullary SRCC of duodenum have been reported in English literature across the world. We report a rare case of nonampullary duodenal SRC, successfully managed with duodenopancreatic resection in our institution.

CASE PRESENTATION

68-year-old lady from rural background presented to us with features of Gastric outlet obstruction, anorexia, significant weight loss. A gastroduodenoscopy revealed a circumferential thickening of D1-D2 junction with deep ulcer at lateral aspect of growth. Biopsy from growth revealed poorly differentiated adenocarcinoma-Signet Ring cell type. After evaluating with cross sectional imaging which revealed localized disease, patient was taken up for surgery. Intraoperative findings revealed a hard bulky circumferential growth involving first and second part of duodenum free from adjacent major vasculature. Multiple enlarged subcentimetric sized lymph nodes noted along common hepatic artery, right side of portal vein and in retropancreatic region.

A standard Whipple's pancreaticoduodenectomy was performed along with LN clearance.Cut section revealed growth involving D1-D2 with normal duodenal ampulla. She had uneventful post operative recovery. RT removed on 4th POD and semisolids allowed on POD 6. DFA on Day-3 revealed values of less than 3 times of upper normal limit. She discharged to home with left drain in situ draining 150 ml serous content on 7th POD which wasremoved on subsequent OPD visit. Final Histopathology confirmed poorly differentiated adenocarcinoma -signet ring cell type (T4N2M0). patient is on follow up and after 30 months of surgery doing well without any evidence of disease.

DISCUSSION

SRCC of duodenum in non-ampullary region is extremely rare entity and so far, only 5 cases have been reported across the world over a period of last one and half decade. This rare entity was first reported in 2006 by Luis J et al where they found a confluent growth in 2nd part of duodenum in 68-year-old man who had bilobar liver metastasis on exploration and received a palliative surgery.[1]

After that few reports were came into literature describing SRCC in ampullary region of 2nd part of duodenum which almost exclusively present with obstructive jaundice and has got better prognosis as compared to those which are developed in non-ampullary region.[6]

SRCCs are predominantly found in gastric cancers; these tumors might originate from heterotopic gastric mucosa[7] or gastric-type metaplastic epithelia, which are considered to be a protective response to elevated acidity and are observable in the duodenal bulb of peptic ulcer patients[8], although our patient did not have any history of peptic ulcer disease.

A case report by Carasca et al[5] (2018) described that non-ampullary duodenal SRCC produces histopathological challenge for diagnosis and special stains/ immunostaining remains the cornerstone in the diagnosis where difficulties arise. They described microscopic examination which revealed a proliferation of atypical cells, with enlarged, irregular, eccentric nuclei, pushed to the periphery of the cells by large, intracytoplasmic Periodic Acid Schiff (PAS)-Alcian blue positive mucin vacuoles. The tumor cells were CDX2, cytokeratin (CK) 7 and CK 18/8 positive, which suggested a primary upper gastrointestinal tract site of origin. Immunostaining for mucin (MUC) 2 and MUC5AC was also positive demonstrating the duodenal goblet cells differentiation with a mixed gastric-foveolar and intestinal phenotype. Based on the morphological features and the immunohistochemical profile, a diagnosis of SRC carcinoma associated with poorly differentiatedadenocarcinoma of the non-ampullary duodenum was set.

SRCC nonampullary region remains extremely rare entity. These tumors are diagnosed to have advanced disease on presentation and can be managed well with surgical resection, by Whipples Pancreaticoduodenectomy in majority, which are localized.

Table: Previous case reports across the world describing Non-ampullary Signet Ring Cell Carcinoma of Duodenum

S.No	Author	Year	Site of the lesion	Treatment
1	**Luis J et al[1]**	2006	2nd part of duodenum	Unresectable on Laparotomypalliative surgery
2	**Terada et al[2]**	2014	3rd part of duodenum & proximal jejunum	Chemoradiation, had brain metastasis
3	**Mochizuki et al[3]**	2015	2nd part of Duodenum	Pylorus Preserving Pancreaticoduodenectomy
4	**Hirano et al[4]**	2017	Duodenal Bulb	Standard Whipples Pancreaticoduodenectomy
5	**Carasca et al[5]**	2018	2nd portion of Duodenum	Standard Whipples Pancreaticoduodenectomy
6	**Our Study**	2019	D1-D2 part of Duodenum	Standard Whipples Pancreaticoduodenectomy

Fig 1: Surgical specimen of WPD showing normal ampulla, A feeding tube is inserted through CBD and coming out from Normal ampulla in duodenum. A hard circumferential growth with deep ulcer noted in D1/D2 junction (Circled). Normal stomach mucosa is apparent proximally.

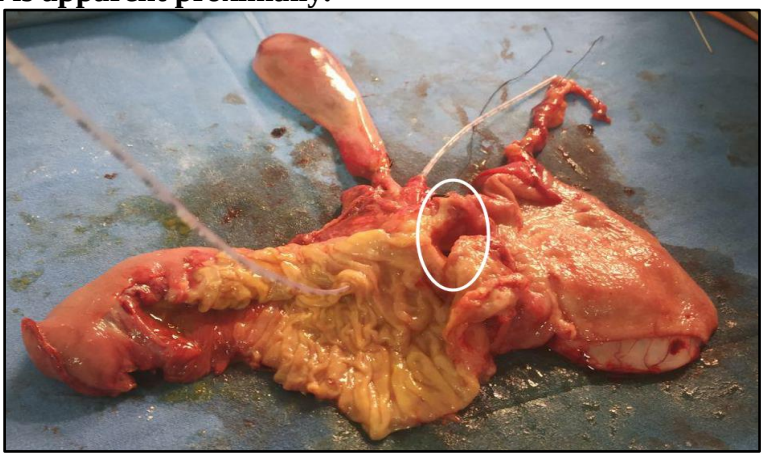
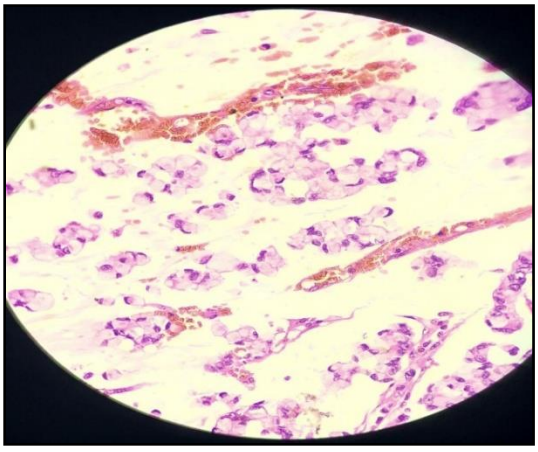


Fig 2: On histopathological examination Signet Ring Cell can be identified with submucosa of Duodenum.



REFERENCES

1. Lopez-Garcia LJ, Ojeda A, Toro DH. Primary Duodenal Signet Ring Cell Carcinoma Presenting as Gastric Outlet Obstruction. Puerto Rico health sciences journal. 2006 Dec 29;25(4).
2. Terada T. Signet-ring cell carcinoma of the nonampullary duodenum and proximal jejunum: a case report with an immunohistochemical study. Endoscopy. 2014;46(S 01):E348.
3. Mochizuki K, Kondo T, Tahara I, Inoue T, Kasai K, Oishi N, Nakazawa T, Katoh R. Signet ring cell carcinoma of the non-ampullary duodenum: A case report. Pathology-Research and Practice. 2015 Oct 1;211(10):801-4.
4. Hirano K, Yamagata Y, Tatsuoka T, Kubota Y, Saito K, Koketsu S, Okuyama T, Sugamata Y, Fujii A, Ban S, Tagaya N. A Case of Poorly Differentiated Adenocarcinoma with Signet Ring Cell Carcinoma of the Duodenal Bulb: A Case Report. International Surgery. 2017 Mar;102(3-4):141-4.
5. Caraşca C, Simion G, Nechifor-Boilă AC, Boeriu AM, Dobru ED. A rare case of signet-ring cell carcinoma associated with poorly differentiated adenocarcinoma of the non-ampullary duodenum. Romanian journal of morphology and embryology= Revue roumaine de morphologie et embryologie. 2018;59(1):311-5.
6. Damania R, Weaver J, Cocieru A. Signet ring cell carcinoma of the ampulla of vater with early development of bone metastasis: Case report and review of the rare malignancy. Journal of gastrointestinal cancer. 2016 Mar 1;47(1):89-92.
7. 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.
8. Wen X, Wu W, Wang B, Yao H, Teng X. Signet ring cell carcinoma of the ampulla of Vater: Immunophenotype and differentiation. Oncol Lett 2014;8(4):1687–92.