

Case Series

Limited Resection in Duodenal Malignancy: A single Center Experience

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Abstract

Article information		Background: Pancreas-sparing duodenectomy [PSD] has come a long way since its conception in 1990s. Since then, less than 200 cases have been published.				
Received:	18-10-2024	Aim of the study: This study designed to share the outcomes of 3 Lynch Syndrome [LS], 2 gastrointestinal stromal tumors [GIST] and 1 case of duodenoieiunal adenocarcinoma				
Accepted:	13-12-2024	who had successful PSD, aiming to highlight the efficacy and safety of this challengin operation.				
DOI: <u>10.21608/ijma.2024.329327.2055.</u>		Patients and Methods: This case series included 6 patients with duodenal neoplasms who underwent PSD from 2022-2024. After obtaining patient's consent, their clinical and histopathological data was reviewed retrospectively using hospital records. Details like patient demographic, location of the tumor, past surgical history, associated syndrome, and surgical resection were obtained. All patients had biopsy specimens to prove GIST or adenocarcinoma. Patients with associated syndromes had specimens validated with immunohistochemistry. Selective duodenal resection was performed after excluding the involvement of the pancreatic head and other major vessels. The outcomes and complications were recorded				
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Email: m_asar@windowslive.com		Results: The mean age of the patients was $57.6 + 9.6$ years with a range of $45-69$ years, and the				
Citation: Asar MM, Hafez AS. Limited Resection in Duodenal Malignancy: A single Center Experience. IJMA 2024 Dec; 6 [12]: 5161-5164. doi: <u>10.21608/ijma.</u> <u>2024.329327.2055.</u>		female: male ratio was 1:5. The most common presentation was weight loss [66.6%, n=4 followed by obstruction [33%, n=2]. Three cases were associated with Lynch Syndrom According to the tumor location, two cases were D3, two cases were DJ, one case was D and one case was D4. In terms of the histopathological investigation, four cases were adenocarcinoma, and two cases were GIST. All patients underwent doudeno -jejunostor with Wide local excision in one case.				
		Conclusion: Pancreatic Sparing Duodenectomy is safe technique for the management of cases with duodenal adenocarcinoma, and the long term surveillance will add more evidence to the literature on how to better approach and manage cases with Lynch Syndrome following Pancreatic Sparing duodenectomy.				

Keywords: Pancreas-sparing Surgery; Duodenectomy, Lynch Syndrome, Adenocarcinoma; GIST.

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INTRODUCTION

Cancer of the small intestine is rare but dangerous malignancy. It accounts for less than 5% of all gastrointestinal malignancies in the United Stated of America $[USA]^{[1]}$, and less than 5% of gastrointestinal cancers. The median age at diagnosis is in the sixth decade of life ^[2].

The clinical presentation and diagnosis of small bowel tumors are usually delayed, and most patient present initially with non-specific pain ^[3]. Thus, they carry a poor prognosis regardless of stage, with a 5-year overall survival [OS] rate ranging from 14% to 33%. The duodenum is the most frequently involved segment, [55–82%], followed by the jejunum and ileum ^[2], and surgery for complete resection [R0] remains the only potentially curative treatment. The Duodenum is a complex organ; it is related to the ampulla of Vater, the pancreas and major vessels ^[4].

Pancreaticoduodenectomy [PD] has been used increasingly as a safe and appropriate resection option in selected patients with tumors of the pancreas and periampullary region ^[5]. However, due to the high morbidity [30%-50%] and mortality [5%] of PD, alternatives to this invasive procedure have been developed. In certain pathologies and tumor locations such as duodenal adenoma and adenocarcinoma confined to the mucosa, as well as duodenal non-epithelial tumors that are difficult to remove by wedge resection, pancreas-sparing duodenectomy [PSD] can be safely performed ^[6].

PSD surgery has come a long way since its conception in 1990s^[7]. Since then, less than 200 cases have been published^[8]. Depending on the site of the tumor, PSD can include the entire length of the duodenum, or a small portion ^[9]. In addition, it can be performed laparoscopically ^[10], or in an open surgery setting. It classically involves simple bile and pancreatic ductal anastomoses, and gastro jejunostomy. Carcinoid tumors, large supra-ampullary adenomas, multiple duodenal gastrinomas in patients with Multiple Endocrine Neoplasm [MEN-1] syndrome, mucosa-associated lymphoid tissue lymphoma, Crohn's disease, and duodenal gastrointestinal stromal tumors [GISTs] are all common indications of PSD. Another important indication is familial polyposis [FAP], in which PSD has shown safer and more impressive results when compared to its more invasive alternatives ^[11].

Patients with other hereditary caner, such as Lynch Syndrome [LS], can also benefit from this operation. LS is the commonest hereditary colon cancer; it is characterized by non-polypoid adenomas that quickly develop into colorectal carcinoma, and affects the duodenum in about 4.5% of cases ^[12].

GIST, mesenchymal or non-mesenchymal tumor of the intestinal tract, arises in the duodenum in 10% of cases, and shows excellent results when treated with PSD ^[13].

In this article, we shared the procedures, and outcomes of 3 LS cases, 2 GIST cases, and 1 case of duodenojejunal adenocarcinoma who have had successful PSD operations, aiming to highlight the efficacy and safety of this challenging operation.

PATIENTS AND METHODS

This case series study included six patients with duodenal neoplasms who underwent PSD from 2022-2024. After obtaining

patient consent, their clinical and histopathological data was reviewed retrospectively using hospital records. Details such as patient demographic, location of the resected tumor, past surgical history, associated syndrome, and surgical resection were obtained. All patients had biopsy specimens to prove GIST or adenocarcinoma, and patients with associated syndromes had specimens validated with immunohistochemistry. Selective duodenal resection was planned after excluding the involvement of the head of the pancreas and other major vessels. The outcomes and complications were also assessed.

Surgical techniques

With exception of one GIST case with tumor in the 2nd part of the duodenum, all cases were operated on laparoscopically. The port placement was four quadrant port configurations [10 mm epigastric line, 5 mm mid-clavicular line, 5 mm anterior axillary line, and 10 mm umbilicus], in addition to supra pubic port used early in the procedure as optical port.

In the 5 laparoscopic cases, the procedure starts by mobilizing the root of the mesentery starting from the bifurcation of the abdominal aorta, thus exposing the third part of the duodenum, further mobilization of the right mesocolon gives us more exposure of the surgical field. As the dissection continues, the duodenojejunal [DJ] flexure is completely dissected and the distal margin is marked and transected; central lymphadenectomy around the superior mesenteric vessels is done with control of branches directed to the part of the duodenum to be excised. The attention is then directed to separate the duodenum from the pancreas; all efforts should hemostasis using energy vascular sealing devices. When the proximal margin is reached, the main concern is to exclude the limit of the ampulla to avoid its injury or in advert closure by staplers, this is easily achieved if the location of the tumor is away from the second part, or by probing of the Common Bile Duct [CBD], or by Indocyanine green [ICG] dve injection.

A single GIST case with tumor in the second part of the duodenum directly opposite to the ampulla was operated on in a conventional open setting. Due to this proximity and the open setting of the surgery, vital structures were under vision all the time, even during reconstruction of bowel integrity.

Bowel reconstruction was done as stapled duodenojejunal anastomosis in all cases, with exception of one aforementioned GIST case which was done in a conventional open setting and reconstructed in a double layered, hand-sewn fashion

RESULTS

The clinicopathological data of patients is outlined in [Table 1]. The mean age of the patients was 57.6 ± 9.6 years with a range of 45-69 years, and the female: male ratio was 1:5. The most common presentation was weight loss [66.6%, n=4], followed by obstruction [33%, n=2].

Three cases were associated with Lynch Syndrome. According to the tumor location, two cases were in the third part of the duodenum [D3], two cases were DJ, one case was in the second part of the duodenum [D2], and one case was in the fourth part of the duodenum [D4]. In terms of the histopathological investigation, four cases were adenocarcinoma, and two cases were GIST. All patients underwent doudeno -jejunostomy with wide local excision in one case. All patients with Lynch Syndrome had past history of colon cancer and colectomy, two of those patients showed polypoidal lesions on upper gastrointestinal endoscopy, and one patient demonstrated a stenotic lesion in the third part of duodenum and presented with symptoms of gastric outlet obstruction. All LS lesions proved to be adenocarcinoma after biopsy and pathology. One GIST patient complained of sudden onset anemia and computed tomography [CT] imaging showed a mass highly suggestive of stromal tumor in the fourth part of the duodenum, after exclusion of invasion, PSD was performed. Another GIST patient had a more severe presentation with tumor located within a few centimeters to the Ampulla of Vater, pancreas sparing resection was possible only after open surgery and after safeguarding all vital structures. As regards the surgical outcomes, the mean operative time was 2.8 ± 0.8 hours, and the mean blood loss was 171.6 ml with a range of 100 - 400 ml.

The length of hospital stay was 7.8 ± 2.4 days. According to the post operative complications, case 2 developed pneumonia, and case 5 developed intestinal leakage, which was reoperated for control of the ligation, however this case died.

Table [1]: Clinicon	pathological features an	d surgical strategy of	f patients who underwen	t pancreas sparing duodenectomy
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No.	Age	Sex	Presentation	Associated syndrome	Tumor location	Pathology	Surgery performed
1	53	F	Weight loss Melena	Lynch Syndrome	D3	Adenocarcinoma	Doudeno -jejunostomy
2	48	М	Gastric outlet obstruction	Lynch Syndrome	D3	Adenocarcinoma	Doudeno -jejunostomy
3	61	М	Weight loss Melena	Lynch Syndrome	DJ	Adenocarcinoma	Doudeno -jejunostomy
4	66	М	Anemia	None	D4	GIST	Doudeno -jejunostomy
5	69	М	Weight loss Intestinal obstruction	None	DJ	Adenocarcinoma	Doudeno -jejunostomy
6	45	М	Weight loss Melena	None	D2	GIST	Wide local excision, Doudeno-jejunostomy

GIST: Gastrointestinal Stromal Tumor. D2/D3/D4: second, third, and fourth parts of the duodenum

DISCUSSION

Due to the increase utilization of complicated and elaborate surgical methods, especially in patients with small, resectable, non-invasive tumors near the pancreas, it is becoming increasingly clear that organ preservation should the gold-standard ^[14]. We presented six cases, with syndromal and non-syndromal disease, who showed favorable response to pancreas conserving surgery, and better post-operative results than those expected in a pancreaticoduodenectomy.

Regarding GIST, previous studies show the feasibility and safety of this approach ^[15,16]. One study even compares PSD and PD in a series of GIST cases, and finds superior oncological benefits for patients receiving PSD, with only 11.1% patients experiencing minor complications ^[17]. However, research on PSD in patients with Lynch Syndrome is lacking and insufficient, and no conclusive studies show the benefit of PSD on LS cases. In this study we demonstrated the advantage of PSD in LS.

The laparoscopic, minimally invasive approach is one of the main reasons PSD showcases very low mortality and morbidity compared to PD. Five of our patients who were operated on laparoscopically confirmed this. One GIST patient who received open surgery still showed lower risk of common PD complications such as chyle leak and pancreatic fistula ^[18]. This is supplemented by the findings of **Busquets** *et al.*, ^[19] who had only 14% of cases experience major complications, even in the setting of open surgery. This is a great leap from the high [>50%] morbidity seen in open PD surgeries^[18]. Additionally, the open approach allowed for the wide local excision of the tumor in the second part of the duodenum, closely related to the ampulla, giving the confidence and ability to preserve the pancreas which would not have been possible had the operation been done laparoscopically. A few reports exist laparoscopic resection of ampullary and periampullary tumours while also preserving the

pancreas ^[20,21]; however, a portion of the operation has to be completed extracorporially to keep the field and view and limit complications.

Despite the general safety and efficacy of PSD, one should be careful as not overlook the limited but clear indications for this surgery. Select tumors with clear margins and no signs of aggressive malignancy only should be recommended for this procedure [22]. Patient general state should also be considered; in fact, one study in 2016 implicates age as an important prognostic factor for the success of this operation ^[23]. Resection margins should also be taken into account, and that, although ampullary and peri-ampullary tumors can be resected using PSD, caution should be taken as not to emulate the high mortality and morbidity of PD, as demonstrated in one study by Liu et al, where PSD did not show any statistically different overall survival [83%] over PD [81%] ^[22]. PSD is not a complication free surgery; Otsuka et al describes a case acute obstructive pancreatitis on day one post-surgery ^[24], highlighting the importance of pancreatography to exclude congenital variations. Finally, it is worth mentioning that the corroboration of the overall safety of PSD without acknowledging the stability and orthodoxy of the patient selection is not the aim of this study. Care should be taken when selecting the patients. On top of that, Lynch Syndrome patients should be monitored for at least 5 years, as recurrence in the digestive tract and other locations is commonly seen in these patients [25,26].

Conclusion: Pancreatic Sparing duodenectomy is safe technique for the management of cases with duodenal adenocarcinoma, and the long term surveillance will add more evidence to the literature on how to better approach and manage cases with Lynch Syndrome following PSD.

Conflict of interest and financial disclosure: none

REFERENCES

- Siegel R, DeSantis C, Virgo K, Stein K, Mariotto A, Smith T, et al. Cancer treatment and survivorship statistics, 2012. CA Cancer J Clin. 2012 Jul-Aug;62[4]:220-41. doi: 10.3322/caac.21149.
- Neugut AI, Jacobson JS, Suh S, Mukherjee R, Arber N. The epidemiology of cancer of the small bowel. Cancer Epidemiol Biomarkers Prev. 1998 Mar;7[3]:243-51. PMID: 9521441
- Dabaja BS, Suki D, Pro B, Bonnen M, Ajani J. Adenocarcinoma of the small bowel: presentation, prognostic factors, and outcome of 217 patients. Cancer. 2004 Aug 1;101[3]:518-26. doi: 10.1002/cncr.20404.
- Sharma A, Nagar A, Varshney P, Tomar M, Sarin S, Choubey RP, Kapoor VK. Pancreas-preserving limited duodenal resection: Minimizing morbidity without compromising oncological adequacy. Ann Hepatobiliary Pancreat Surg. 2022 May 31;26[2]:149-158. doi: 10.14701/ahbps.21-124.
- Yeo CJ, Cameron JL, Sohn TA, Lillemoe KD, Pitt HA, Talamini MA, et al. Six hundred fifty consecutive pancreaticoduodenectomies in the 1990s: pathology, complications, and outcomes. Ann Surg. 1997 Sep;226 [3]: 248-57. doi: 10.1097/00000658-199709000-00004.
- Nakayama Y, Konishi M, Gotohda N, Kato Y, Aizawa H, Kudo M, et al. Comparison of postoperative early and late complications between pancreas-sparing duodenectomy and pancreatoduodenectomy. Surg Today. 2017 Jun;47[6]:705-711.
- Vallance S. Duodenectomy without pancreatectomy for extensive benign villous adenoma of the duodenum. Aust N Z J Surg. 1990 Apr;60 [4]:3 11-4. doi: 10.1111/j.1445-2197.1990.tb07374.x.
- Cantalejo-Díaz M, Ramia-Ángel JM, Palomares-Cano A, Serradilla-Martín M. Pancreas-Preserving Total Duodenectomy: A Systematic Review. Dig Surg. 2021;38[3]:186-197. doi: 10.1159/000515718.
- Konishi M, Kinoshita T, Nakagohri T, Takahashi S, Gotohda N, Ryu M. Pancreas-sparing duodenectomy for duodenal neoplasms including malignancies. Hepatogastroenterology. 2007 Apr-May;54[75]:753-7.
- Stauffer JA, Adkisson CD, Riegert-Johnson DL, Goldberg RF, Bowers SP, Asbun HJ. Pancreas-sparing total duodenectomy for ampullary duodenal neoplasms. World J Surg. 2012 Oct;36[10]:2461-72. doi: 10.1007/ s00268-012-1672-3
- Mackey R, Walsh RM, Chung R, Brown N, Smith A, Church J, Burke C. Pancreas-sparing duodenectomy is effective management for familial adenomatous polyposis. J Gastrointest Surg. 2005 Nov;9[8]:1088-93; discussion 1093. doi: 10.1016/j.gassur.2005.07.021.
- Hammoudi N, Dhooge M, Coriat R, Leblanc S, Barret M, Bordacahar B, et al. Duodenal tumor risk in Lynch syndrome. Dig Liver Dis. 2019 Feb;51[2]:299-303. doi: 10.1016/j.dld.2018.10.005.
- Zhou Y, Wang X, Si X, Wang S, Cai Z. Surgery for duodenal gastrointestinal stromal tumor: A systematic review and meta-analysis of pancreaticoduodenectomy versus local resection. Asian J Surg. 2020 Jan;43[1]:1-8. doi: 10.1016/j.asjsur.2019.02.006.
- Parab TM, DeRogatis MJ, Boaz AM, Grasso SA, Issack PS, Duarte DA, et al. Gastrointestinal stromal tumors: a comprehensive review. J Gastrointest Oncol. 2019;10[1]:144-154. doi: 10.21037/jgo.2018.08.20.

- 15. Goh BK, Chow PK, Ong HS, Wong WK. Gastrointestinal stromal tumor involving the second and third portion of the duodenum: treatment by partial duodenectomy and Roux-en-Y duodenojejunostomy. J Surg Oncol. 2005 Sep 15;91[4]:273-5. doi: 10.1002/jso.20311.
- Yamashita S, Sakamoto Y, Saiura A, Yamamoto J, Kosuge T, Aoki T, Sugawara Y, Hasegawa K, Kokudo N. Pancreas-sparing duodenectomy for gastrointestinal stromal tumor. Am J Surg. 2014 Apr;207[4]:578-83. doi: 10.1016/j.amjsurg.2013.05.009
- 17. Lu C, Jin W, Mou Y, Shao H, Wu X, Li S, et al. Optimal Laparoscopic Management and Oncological Outcomes of Gastrointestinal Stromal Tumors in Duodenum: Pancreaticoduodenectomy or Pancreas-Sparing Duodenectomy? Cancer Manag Res. 2020 Jun 19;12:4725-4734. doi: 10.2147/CMAR.S254972.
- Simon R. Complications After Pancreaticoduodenectomy. Surg Clin North Am. 2021 Oct;101[5]:865-874. doi: 10.1016/j.suc.2021.06.011.
- Busquets J, Lopez-Dominguez J, Gonzalez-Castillo A, Vila M, Pelaez N, Secanella L, Ramos E, Fabregat J. Pancreas sparing duodenectomy in the treatment of primary duodenal neoplasms and other situations with duodenal involvement. Hepatobiliary Pancreat Dis Int. 2021 Oct;20 [5]: 485-492. doi: 10.1016/j.hbpd.2021.02.007.
- 20. Abe N, Hashimoto Y, Kawaguchi S, Shimoyama H, Kojima Y, Yoshimoto E, et al. Successful treatment of large adenoma extending close to the papilla in the duodenum by laparoscopy-assisted pancreas-sparing duodenectomy. Asian J Endosc Surg. 2016 Feb;9[1]:52-6. doi: 10.1111/ases.12246.
- 21. Vega EA, Salehi O, Nicolaescu DC, Krishnan S, Alarcon SV, Kozyreva O, et al. Laparoscopic Pancreatic Head Preserving Total Duodenectomy: The Parenchymal Sparing Alternative to a Whipple. Ann Surg Oncol. 2021 Jan; 28[1]:131-132. doi: 10.1245/s10434-020-08715-z.
- 22. Liu B, Li J, Zhang YJ, Yan LN, You SY, Lau WY, et al. Pancreas-sparing duodenectomy with regional lymph node dissection for early-stage ampullary carcinoma: A case control study using propensity scoring methods. World J Gastroenterol. 2015 May 14;21[18]:5488-95. doi: 10.3748/wjg.v21.i18.5488.
- 23. Watanabe Y, Ishida H, Baba H, Iwama T, Kudo A, Tanabe M, Ishikawa H. Pancreas-sparing total duodenectomy for Spigelman stage IV duodenal polyposis associated with familial adenomatous polyposis: experience of 10 cases at a single institution. Fam Cancer. 2017 Jan; 16 [1]: 91-98. doi: 10.1007/s10689-016-9932-2.
- 24. Otsuka S, Sugiura T, Uesaka K. Acute obstructive pancreatitis after pancreas-sparing total duodenectomy in a patient with pancreas divisum: a case report. Surg Case Rep. 2016 Dec;2[1]:126. doi: 10.1186/s40792-016-0255-1.
- 25. Shinojima N, Ozono K, Yamamoto H, Abe S, Sasaki R, Tomita Y, et al. Lynch syndrome-associated chordoma with high tumor mutational burden and significant response to immune checkpoint inhibitors. Brain Tumor Pathol. 2023;40[3]:185-190. doi: 10.1007/s10014-023-00461-w.
- 26. Lonati C, Moschini M, Simeone C, Spiess PE, Necchi A. Lynch syndrome in urological practice: diagnosis, therapeutic strategies, and screening for upper tract urothelial carcinoma. Curr Opin Urol. 2022 Jan 1;32[1]:40-47. doi: 10.1097/MOU.00000000000936.