

## Pancreas Sparing Distal Duodenectomy of Third and Fourth Part Duodenum for Neuroendocrine Tumor (NET): A Case Report and Review of Literature

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The authors describe a case of the surgical treatment of duodenal neuroendocrine tumor (d-NET) at third and fourth part (D3-D4) of duodenum. A woman 42 years old presents us with severe intermittent vomiting for 2 months. Her work-up found a large duodenal wall mass size 8 cm at D3-D4 of duodenum with displaced pancreatic body. We performed a pancreas sparing distal duodenectomy (PSDD) with end to side duodenojejunostomy with lymphatic dissection. Post-operative course found mild pancreatitis with infected intra-abdominal fluid collection. She developed partial gut obstruction 2.5 months after operation but complete resolution by conservative treatment. PSDD is feasible as an operative option to avoid pancreaticoduodenectomy. This procedure can make adequate negative margin for d-NET. 1 year follow-up has not found any recurrence.

**Keywords:** Duodenal neuroendocrine tumor, Third and fourth part duodenum, Pancreas sparing distal duodenectomy

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Role of surgical treatment is the primary treatment of neuroendocrine<sup>(1)</sup>. Duodenal neuroendocrine tumor (d-NET) is a rare tumor and found less than 2% of gastrointestinal neuroendocrine tumors (GI-NET)<sup>(2)</sup>. They are a slow growing tumor but they can become malignant<sup>(1,3)</sup>. Pancreaticoduodenectomy (PD) is operative of choice for duodenal tumor but associated high morbidity and mortality. Several literatures purposed superior outcome of pancreas sparing distal duodenectomy (PSDD) for duodenal tumor but few report on the role of PSDD for NET of D3-D4 of duodenum<sup>(4-6)</sup>. We present success in treating the case of d-NET of D3-D4 of duodenum by PSDD and literatures reviewed on the surgical role of PSDD.

### Case Report

A 42 year old female presents severe intermittent vomiting for 2 months. She had vague abdominal pain for 6

months before it becoming severe abdominal pain. Her underlying disease is hypertension, thyroid nodule planned thyroidectomy due to present hurthle cell by fine needle aspiration (FNA) biopsy. Laboratory work up: CA19-9 is 19.8 U/ml and other unremarkable. Esophagogastroduodenoscopy (EGD) found gastritis. Abdominal ultrasound has found a pancreatic mass size 7.7 cm and then computed tomography (CT) scan has found a 7.7x7.4 cm exophytic mass of third and fourth part (D3-D4) of duodenum wall medial to uncinate process of pancreas with multiple mesenteric lymphadenopathy, but has not found distant metastasis or vascular invasion (Figure 1).

The authors performed midline incision and then opened lesser sac to identify duodenal wall mass. The characteristic of mass protrudes from D3-D4 of duodenum wall and adheres with uncinate process, head, and body of pancreas. The mass was removed through meticulous dissection from pancreas without injury. We found multiple lymphadenopathy around mass and para aorta and then performed lymph node biopsy for intra-operative pathologic examination. The intra-operative pathologic report is metastatic lymph node and then performed wide lymph node dissection along lesion and para aorta. The authors use gastrointestinal anastomosis (GIA) staple cut proximal and distal segment of D3-D4 of duodenum and then reconstructed end to side duodenojejunostomy by hand sewing double

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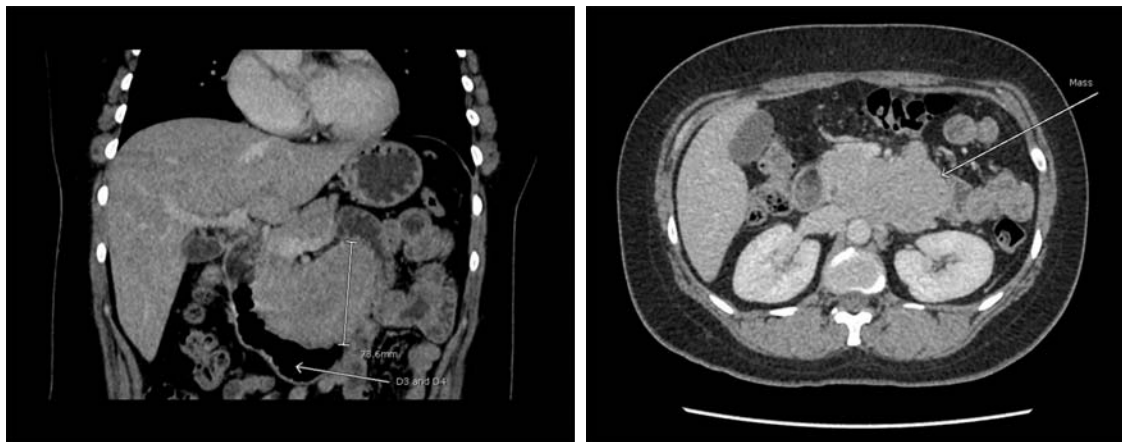
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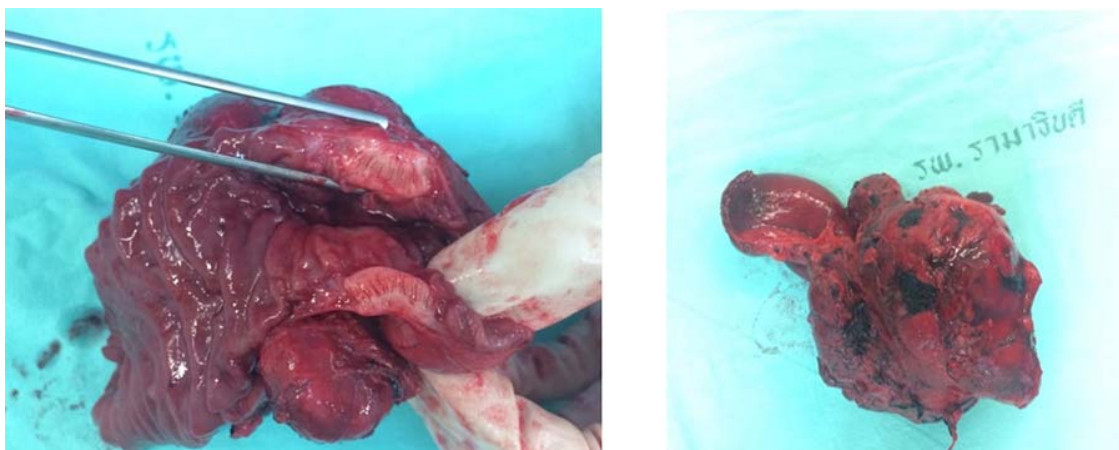
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**Figure 1.** CT scan shown 7.7x7.4 cm exophytic mass of third and fourth part duodenum wall medial to uncinate process



**Figure 2.** The specimen after resection shown intra luminal stenosis (Forceps).

layers. The authors palpable tamarind seed (needs clarification) in the specimen that cause intermittent obstruction at intraluminal stenosis (Figure 2). To close a suction drain was placed at paraduodenal anastomosis and under pancreas. Total blood loss is 800 ml. First week, she had mild pancreatitis. Second week, she had abdominal pain with fever from infected intra-abdominal collection which was resolved by percutaneous drain and intravenous antibiotic. The patient was discharged at 45 days after operation. She was readmitted for partial gut obstruction at 1 month after discharge, but improved by conservative treatment. At one year follow-up, recurrent condition was not found.

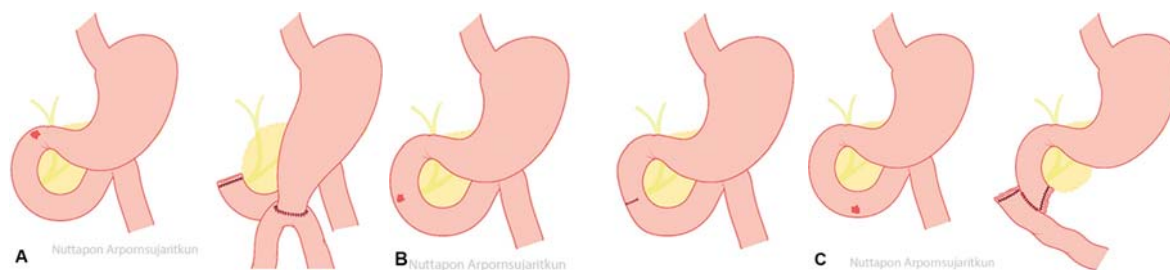
### Discussion

The original pancreas sparing duodenectomy (PSD) first described in 1995 by Chung RS et al<sup>(7)</sup>. This

procedure was treated in familial adenomatous polyposis in duodenum. The detail of procedure must detach duodenum from head of pancreas and then re-implant bile duct and pancreatic duct. Now, PSD was classified in several types due to position of duodenal tumor (Figure 3). It frequently is confused. PSDD is one type procedure of PSD.

The procedure of duodenectomy at D3-D4 of duodenum has several names in the literature. Example, pancreas sparing distal duodenectomy (PSDD)<sup>(5)</sup>, limited distal duodenectomy<sup>(4)</sup>, pancreas sparing duodenectomy (PSD)<sup>(8)</sup>, pancreas preserving distal duodenectomy (PPDD)<sup>(9)</sup>, segmental resection<sup>(6)</sup>. In this literature, we used PSDD due to the position of lesion, below ampulla of Vater.

The d-NET is rare type of duodenal tumor. From our review and search from published literature about the role of PSDD for d-NET of D3-D4 of duodenum. We found only three published literatures<sup>(4-6)</sup>. This is possibly due to



**Figure 3.** Type of pancreatic sparing duodenectomy due to position of lesion (A, B, C).

suboptimal reports on the definition of d-NET and types of procedure. At the time of diagnosis, most of d-NET present regional lymph node metastasis of 40 to 60% with liver metastasis at less than 10%, especially, with a d-NET size of more than 2 cm which risks to metastasis<sup>(1)</sup>. From literature review and our case (Table 1), we found that most patients have non-functional tumors with size of more than 2 cm and positive regional lymph nodes or liver metastasis. Only 2 cases do not have regional or liver metastasis. It confirms that d-NET is usually slow growing without symptomatic. High risk metastasis is when tumor size is more than 2 cm<sup>(1)</sup>. No good research compared outcomes between aggressive versus non-aggressive locoregional resection. However, d-NET at D3-D4 of duodenum is the part of non-ampulla neuroendocrine tumor (naNET) that shows low aggressive behavior and differed histologically from ampulla neuroendocrine tumors (aNET). Therefore, d-NET at D3-D4 of duodenum must be less aggressive, locoregional resection<sup>(6,10)</sup>.

PSDD for d-NET of D3-D4 of duodenum should be beneficial for patients to avoid excessive resection from pancreaticoduodenectomy. This procedure is a feasible and safe procedure. A general surgeon can due this.

### What is already known on this topic?

Duodenal neuroendocrine tumor is very rare. A few case reports about pancreas sparing distal duodenectomy of third and fourth part duodenum.

### What this study adds?

We reviewed literatures about duodenal neuroendocrine tumors that were operated on with pancreas sparing distal duodenectomy (PSDD) at third and fourth part duodenum. The result from literature can conclude that PSDD can be safe and avoid pancreaticoduodenectomy with good outcome.

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### Potential conflicts of interest

The authors declare no conflict of interest.

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**Table 1.** Literature review of PSDD for d-NET of D3-D4 of duodenum

|                                  | Case 1                | Case 2                                         | Case 2                                     | Case 3                                                         | Case 4                      | Case 5                          | Our case                                                      |
|----------------------------------|-----------------------|------------------------------------------------|--------------------------------------------|----------------------------------------------------------------|-----------------------------|---------------------------------|---------------------------------------------------------------|
| Author                           | Zyromski NJ et al (6) | Zyromski NJ et al (6)                          | PA Sali et al (5)                          | PA Sali et al (5)                                              | A Golhar et al (4)          | A Golhar et al (4)              | Arpornsujaritkun et al                                        |
| Reference                        | (6)                   | (6)                                            | (5)                                        | (5)                                                            | (4)                         | (4)                             |                                                               |
| Age                              | 67                    | 48                                             | 57                                         | 43                                                             | 44                          | 58                              | 42                                                            |
| Sex                              | Female                | female                                         | Male                                       | Female                                                         | Male                        | Male                            | Female                                                        |
| Tumor size                       | 8 cm                  | 4 cm                                           | 6.6 cm                                     | No data                                                        | 11 cm                       | 2 cm                            | 8 cm                                                          |
| Regional/Distant metastasis      | No                    | No                                             | Lymph node, N1a                            | Lymph node and liver metastasis                                | Lymph node                  | Lymph node and liver metastasis | Lymph node                                                    |
| Function/non-function tumor      | No data               | No data                                        | Non-function                               | Non-function                                                   | Non-function                | Non-function                    | Non-function                                                  |
| Site of tumor                    | D3-D4 of duodenum     | Fourth part duodenum                           | Duodenum below ampulla                     | Fourth part duodenum                                           | Duodenojejunal flexor       | Third part duodenum             | D3-D4 of duodenum                                             |
| Surgical treatment               | Segmental resection   | Segmental resection                            | Pancreas sparing distal duodenectomy       | Pancreas sparing distal duodenectomy with liver metastatectomy | Limited distal duodenectomy | Limited duodenectomy, liver     | PSDD                                                          |
| Comorbidity                      | No                    | No                                             | History of nephrectomy due to kidney stone | No                                                             | No                          | No                              | Second primary papillary thyroid cancer                       |
| Pathology                        | Carcinoid tumor       | Carcinoid tumor                                | d-NET, stage IIIA                          | d-NET, stage T3N1M1                                            | d-NET, grade 1, Ki-67 <3%   | d-NET, grade 1, Ki-67 <3%       | d-NET                                                         |
| Complication                     | No data               | No data                                        | No                                         | No                                                             | Delay gastric emptying time | Delay gastric emptying time     | Infected collection, intraabdominal partial bowel obstruction |
| Follow up (data from literature) | Recurrent at 2 years  | Recurrent at 9 years but follow-up to 22 years | Alive without recurrent at 10 months       | No data                                                        | Alive at 29 months          | Alive at 3 months               | 12 months, Alive without recurrent                            |

D3-D4 = third and fourth part duodenum, PSDD = pancreas sparing distal duodenectomy