Original Article

Whipple's Procedure For A Duodenal Neuroendocrine Tumour – An Uncommon Disease – Case Report

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DOI: 10.47750/pnr.2023.14.S01.14

Abstract

Duodenal NETs (Neuroendocrine Tumours) are rare neoplasms that represent a clinical challenge due to heterogeneity of their biological behaviour, diagnosis and treatment options. They represent 1-3% of all primary duodenal tumours. Our case report aims to increase awareness and highlight some issues related to the diagnosis and management of duodenal Neuroendocrine tumours.

We present the case of a 55-year-old man with a large, slowly-growing Neuroendocrine tumour of the duodenum which was incidentally identified after a screening USG of the abdomen. A Whipple's procedure with complete tumour excision was performed. Patient was comfortable on discharge and on regular follow up.

The mainstay of treatment of duodenal NET is surgery with the intent to cure. The management protocol is based on ENETs guidelines. Surgical resection of D-NETs should be followed up by CECT of A+P and Chromogranin A levels performed at 6 and 12 months after surgery, and then every year for at least 3 years

KEY WORDS

1. DUODENAL NEUROENDOCRINE TUMOUR
2. CASE REPORT
3. WHIPPLE'S PROCEDURE
4. CHROMOGRANIN

INTRODUCTION

Neuroendocrine tumours (NETs) are rare tumours with an annual incidence of 3–5 cases/1,00,000 population. However, with improvements in diagnostic tools, the frequency of its incidence has been steadily increasing. [1,2,3] The neuroendocrine (NE) system includes endocrine glands and scattered cells in the exocrine parenchyma, such as endocrine cells of the respiratory and digestive tracts. These specifically comprise what is known as the diffuse endocrine system. [4,5] Around 60% of NETs are located in the gastrointestinal tract, primarily in the small bowel (30%) [6]. This case report is of a patient diagnosed with a Duodenal Neuroendocrine Tumour and managed in a community practise setting.

CASE REPORT

Patient Information:

A 55yr old man with no apparent complaints presented with an ultrasonography of the abdomen showing a mass in the stomach seen in during a routine health check-up. He had no history of abdominal pain , distension or any other bowel complaints. He is a known hypertensive, controlled on medication. He has had no significant complaints in the past. His personal and family history was unremarkable.

Clinical Findings:

On examination, he was well nourished and vitally stable. Abdominal examination was normal with no evidence of abdominal lump or tenderness in the abdomen.

Diagnostic Assessment:

The Ultrasonography suggested 2 solid hypoechoic masses related to the gastric antrum measuring 2.8x2.7x2.4cm and 4.9x4.9x4.2 cm respectively with increased vascularity. Gastroscopy showed a well circumscribed mass in the antrum with normal mucosa. Biopsy taken from the mass showed chronic superficial gastritis. CECT(A+P) showed a well defined exophytic heterogeneously enhancing lesion present in relation to the antro-pyloric region of the stomach, measuring 5x4.5cms. Fat planes with surrounding structures appeared well maintained. The ultrasonography and CECT showed normal duodenum, pancreas and surrounding structures with no lymphadenopathy.

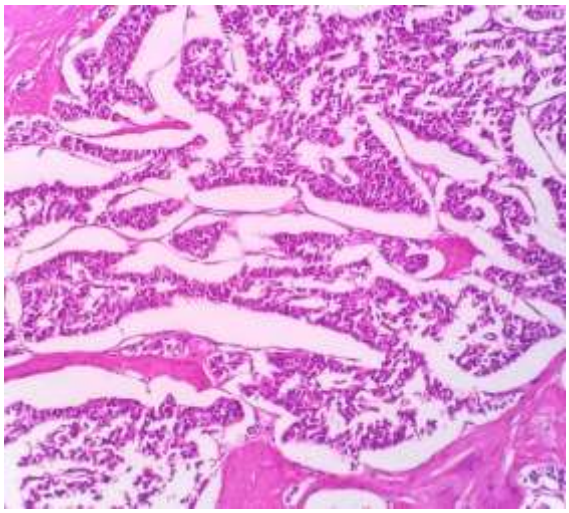
Therapeutic Intervention:

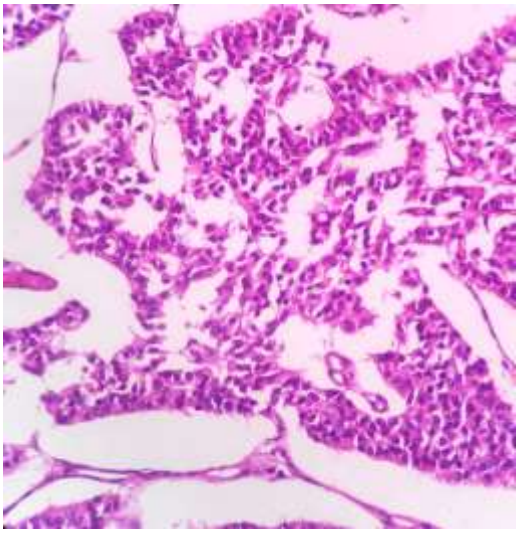
Patient was explained about the condition and counselled for a distal gastrectomy with a gastro-jejunal bypass. On entering the peritoneum through a midline incision, the internal organs were examined. There was evidence of a growth 2x3cm involving the anterior wall of the antropyloric region of the stomach. Another growth arising from the first part of duodenum D1 involving the mesocolon and superior border of head of pancreas. Rest of the abdominal organs were normal with no evidence of distant metastasis. Intra operative decision was made that the patient will require a whipple's surgery for adequate resection of the tumour. Whipples procedure was done. Additionally, a segmental colectomy with colo-colic anastomosis was also was done due to the involvement of the transverse mesocolon. A temporary feeding jejunosotomy was also made to enhance post op recovery.

Post-operative period was uneventful and patient recovered and discharged on POD 7

A) image showing excised specimen of gall bladder, duodenum and head of pancreas showing growth arising from first part of duodenum

B) Intra operative image post resection of the tumour





C) Histological image showing tumour cells with scanty cytoplasm and nuclear pleomorphism with salt and pepper chromatin.

The histopathological examination was suggestive of a Neuroendocrine tumour arising from the First part of Duodenum. The tumour was well differentiated, Grade 1 with invasion of the subserosa. The second swelling was a large paraduodenal metastatic nodal mass with no extranodal extension and no evidence of tumour extending into the colon or pancreas although seen adhered to both. The mass was composed of nests, cords and trabeculae of small monotonous epithelial cells in a 'salt and pepper' pattern. Mitotic activity was negligible (<2/10hpf). Following IHC analysis, they were found to be diffusely positive for Synaptophysin and Chromogranin with a MIB1 index of 1%(low). No specific hormones were detected.

DISCUSSION

Neuroendocrine Tumours are a very rare heterogenous group of disorder comprising approximately 2% of all malignancies. Neuroendocrine neoplasms can occur anywhere along the gastrointestinal tract, but are most commonly found in the stomach, duodenum, ileum, pancreas, appendix, and rectum. [7,8,9]

Duodenal NETs include all duodenal tumours with characteristics specific to neuroendocrine tumours such as Positivity for Synaptophysin and Chromogranin (secretory vesicle proteins) and also presence of specific GI hormones. [10,11] D-NETs are rare tumours with an overall incidence of 0.19/100000. Recent trends indicate an increase in the incidence, primarily due to improvements in diagnostic tools. These tumours comprise 1%-3% of primary duodenal tumours, 11% of small intestinal NETs, and 5%-8% of all GI-NETs. [12,13]

D-NETs include gastrinomas, somatostatin-producing tumours, non-functional serotonin-containing tumours, poorly differentiated neuroendocrine carcinomas, and gangliocytic paragangliomas. Non-functional duodenal NETs are the most frequent type comprising almost 90% of all Duodenal NENs. [15] They often produce peptides when examined on immunocytochemical studies (gastrin, serotonin, calcitonin, somatostatin) but do not produce a clinical syndrome.[11] Most of D-NETs are, therefore, asymptomatic and diagnosed incidentally on Oesophagogastroduodenoscopy. D-NETs arise in the deep mucosa and invade the submucosa having either a hemispherical or flatly elevated appearance.[15]

The differential diagnosis of D-NETs includes Gastrointestinal stromal Tumours (GISTs), Adenocarcinomas, Brunner's gland hyperplasia, lymphoid hyperplasia, heterotopic gastric mucosa, metastatic tumours, neurofibromas and schwannomas [16]

In 2015, the National Comprehensive Cancer Network (NCCN) recommended the inclusion of tumour differentiation, mitotic rate, and Ki-67 in the pathology report with specification of the particular classification and grading scheme to avoid confusion.[17]

This condition is suspected from routine histology and confirmed by immunohistochemical study assessing markers of neuroendocrine cells (CgA, NSE, synaptophysin) and the presence of specific hormones (gastrin, somatostatin, serotonin, calcitonin).[11]

Therapy for D-NETs is based on tumour size, location, histological grade, stage, and tumour type. [15] Resection is the primary treatment for duodenal NET. For lesions less than 1 cm Endoscopic resection is preferred. When the tumour is greater than 2 cm or when there is nodal metastasis a surgical resection is the appropriate method [18]. A tumour arising from the first or second portions of the duodenum indicates a pancreaticoduodenectomy due to the broader pattern of lymphatic drainage while, a tumour with its origins in the third or fourth portions may be treated with a segmental resection. [1] The management protocol of D-NETs based on the ENETS guidelines [19] However, if surgery is not feasible due to the extent and spread of disease, medical management to both alleviate symptoms and suppress tumour growth and spread is recommended.[20]

Surgical resection of D-NETs should be followed up by CECT, somatostatin receptor scintigraphy (SRS) and CgA levels performed at 6 and 12 months after surgery, and then annually for a minimum of 3 years. [15] The NCCN guidelines recommend re-evaluation of patients at 3 to 12 months following resection, and then, every 6 to 12 months for a duration of 10 years. [17]

The 5-year survival rate in patients with well-differentiated D-NETs is 80%-85%. However, this rate is only 72% in those with well-differentiated NET. [21,22]

CONCLUSION

Duodenal Neuroendocrine Tumour is a very rare heterogenous disease with a good prognosis if identified early. This is aided by improvements in diagnostic tools which enable its diagnosis in asymptomatic individuals. Adequate surgical resection and close follow up is essential for curative treatment.

Additional Information

No conflicts of interests.

DECLARATIONS

FUNDING – NIL

CONFLICTS OF INTEREST – NIL

AVAILABILITY OF DATA TRANSPARENCY – AVAILABLE

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