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Multiple Intestinal Neuroendocrine Tumour Presented as a Large Mesenteric Mass: Case Report and Literature Review

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Authors' contributions

This work was carried out in collaboration among all authors. Authors RB and AE designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors AE and AL managed the analyses of the study and the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Most Neuroendocrine tumours (NETs) are of digestive origin (70%), they can involve from any organ of the digestive system.
The small intestine and rectum are the most common locations, (small intestine 30%, colorectal 30%, appendix 20%, pancreas 10%, stomach 5%).
The clinical presentation may be secondary to the secretion of biologically active peptides or related to the tumour syndrome (pain, and palpable mass).
Chromogranin A or synaptophysin are useful markers for the diagnosis and follow-up of NEN.
The radiological diagnosis of intestinal neuroendocrine neoplasm (NEN) has been greatly improved by the advent of new isotope imaging techniques.
The fact that this tumour is rare and that its symptomatology is varied is a cause of many errors in its diagnosis resulting in therapeutic delay.
Curative surgery remains the "gold standard" for the management of small bowel NETs.

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We report the case of an intestinal neuroendocrine tumour presented as a large mesenteric mass and discuss its epidemiological, clinical, biochemical, histological and therapeutic features in the light of the literature.

Keywords: Neuroendocrine Neoplasm (NEN); chromogranin A; Ki67; octreoscan.

1. INTRODUCTION

Neuroendocrine neoplasm (NEN) of the small intestine are rare tumors with an increasing incidence.

They are characterised by the proliferation of cells expressing structural proteins (chromogranin, synaptophysin).

Clinical manifestations may be secondary to the secretion of biologically active peptides (carcinoid syndrome), or related to the symptoms of tumour mass.

They are often multiple and non-functional and discovered at the stage of metastasis, most often in the liver.

Surgery is the gold standard treatment for intestinal NET and should include surgical resection of the primary tumour and associated mesenteric lymph nodes.

2. CASE REPORT

A 60 year old male patient presented with abdominal pain and vomiting for duration of ten days prior to admission. On examination, he was concious, hemodynamically and respiratorily stable, having no signs suggestive of carcinoid syndrome and no external gastrointestinal bleeding.

Abdominal examination revealed a 6 cm deep mass in the umbilical region, hard and tender, fixed in relation to the deep plane, the rectal exam showed no abnormalities.

The abdominal scan showed the presence of a voluminous mesenteric mass measuring 60*48 mm, containing calcifications and necrosis zone with multiple centimetric and infra-centimetric preaortic lymph nodes, the most voluminous measuring 1.8 cm of diameter.

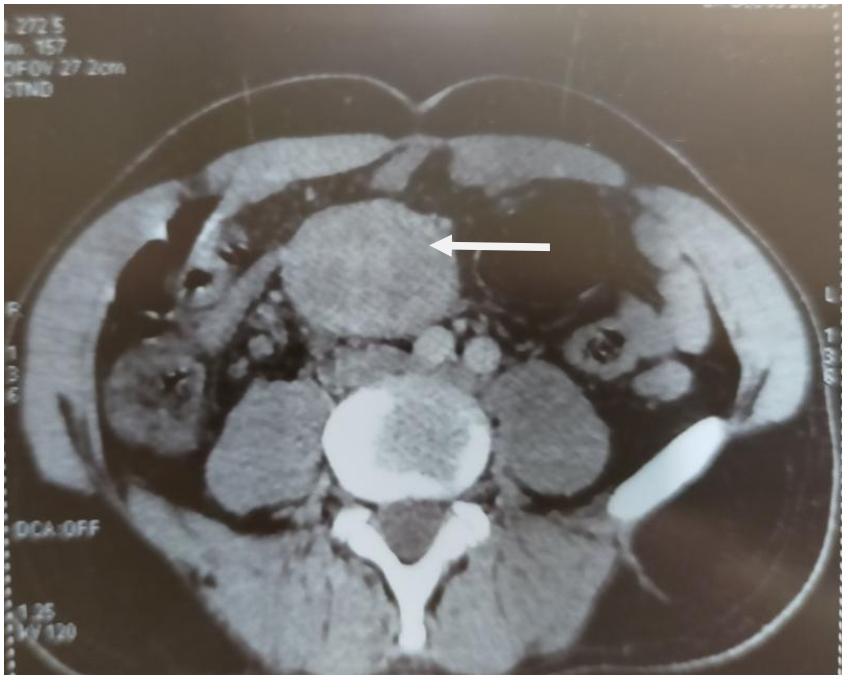

Fig. 1. Axial slide of abdominal CT scan showing voluminous mesenteric mass measuring 60*48 mm, containing calcifications and necrosis zone (arrow)

Surgical exploration found a 6 cm long mesenteric mass with several mesenteric nodules and multiple nodules staggered over the small bowel, one of which was stenosing, he had benefited from a segmental resection at 3.80 of the duodenojejunal flexure, anastomosis and biopsy of the mesenteric mass.

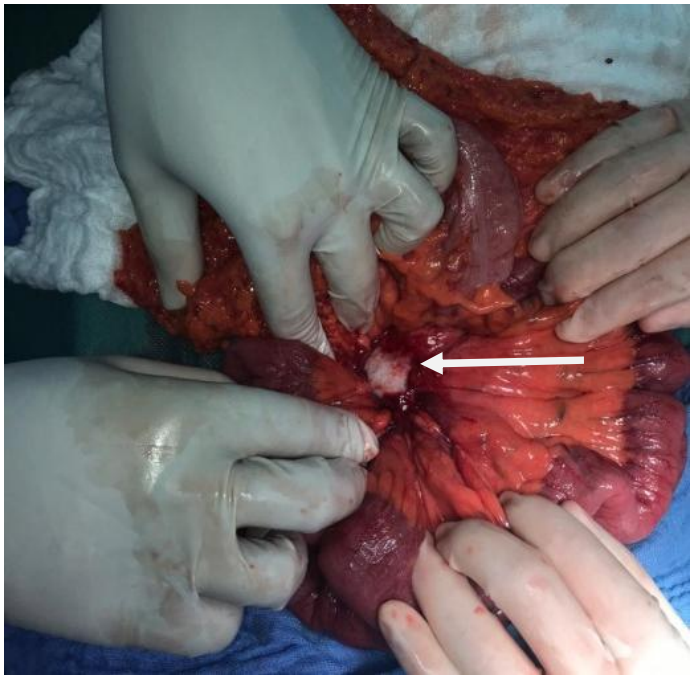

Fig. 2. Per-operatory image showing the mesenteric mass (arrow)

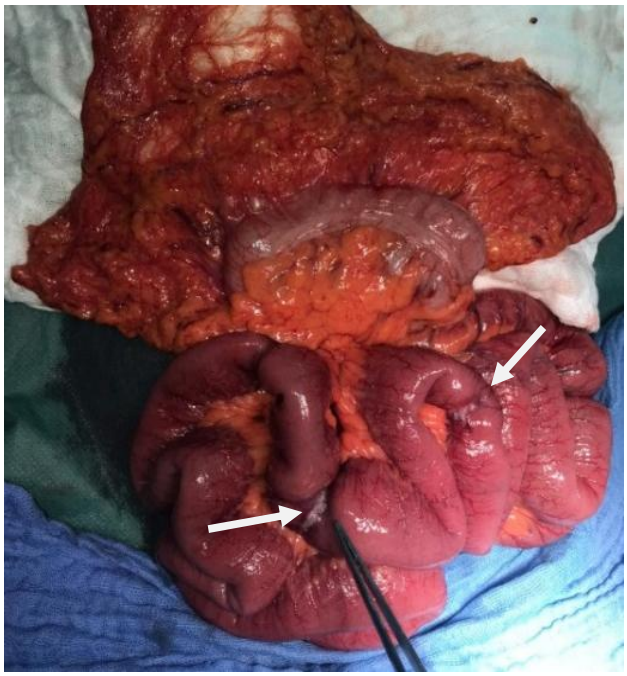

Fig. 3. Per-operatory image showing grey coloured nodules (arrows)

Pathology: Well differentiated neuroendocrine tumour, grade G1.

3. DISCUSSION

Primary neuroendocrine tumours (NETs) can arise in any organ of the digestive tract, most commonly in the small intestine and rectum [1]. The incidence of small bowel (ileal and jejunal) NETs is 0.29/100,000 population [2]. They derive from enterochromaffin cells that produce serotonin. They are heterogeneous tumours developed at the expense of the diffuse endocrine system'cells in the digestive wall.

Clinical symptoms may be related to the secretion of biologically active peptides (carcinoid syndrome) or secondary to the symptoms of tumour mass (pain,palpable mass, vomiting, weight loss, melena, and occlusive syndrome).

Small bowel NETs are responsible of the secretion of multiple biomarkers, the determination of which can be useful for diagnosis or postoperative survey [3]. Chromogranin A (CgA) is the most sensitive and specific biomarker for the diagnosis of NET and its values correlate with both tumour volume and disease prognosis [4].

CT scan is the most common morphological examination performed for the diagnosis of NET of the small intestine [5]. On arterial time small bowel NETs and liver metastases are typically hypervascular and appear bright; however, some may be hypovascular and may appear more prominent (dark) on venous time [6,7].

Abdominopelvic MRI with gadolinium injection and diffusion sequences, which is more sensitive than CT scan for the detection of liver metastases [8]. MRI is recommended in combination with thoracic-abdominopelvic CTscan to perform a comprehensive search for metastases. Spinal MRI is indicated in the case of hepatic metastases to search for extrahepatic localisations, particularly bones [9]. It is recommended for the evaluation of the extension of well-differentiated digestive NET [9].

Somatostatin receptor scintigraphy (SRS) (Octreoscan®) detects intra- and extra-abdominal metastases [10].

Positron emission tomography coupled with CT (PET-CT) using radioactive 68 Ga-labelled SST analogues has shown better performance than SRS [11].

Other isotope techniques also have a wide indication in the extensional assessment of NET:

FDG-PET-CT performs better than SRS in patients with well-differentiated NETs with high Ki67 index (>10%) [12], it is indicated as first-line for CNPD or if the SRS scan is negative, or if the Ki67 proliferation index is >10% [8,9].

PET-CT using FDOPA has demonstrated better diagnostic performance in small bowel NETs compared to CT and SRS [13].

It is currently recommended for use in the case of negative SRS or as part of the preoperative work-up for small bowel NET [14].

European guidelines recommend a thoracic-abdominal-pelvic CT scan followed if possible by a 68Gallium-DOTATOC positron emission tomography (PET) scan which is the most specific for somatostatin receptors.

NENs should be classified according to the WHO classification, which is based on histological differentiation (NET vs. NEC) and tumour grade, based on the proliferation index which is measured by the Ki67 index and the mitotic index.

It is important to note that the Ki-67 index should be determined for both primary tumours and secondary lymphatic and hepatic locations, as prognosis and survival is more accurately predicted from the highest grade observed at a given site [15].

Curative surgery is the gold standard for the management of small bowel NETs. It should be considered as soon as possible, even at the metastatic stage [16,17].

There is no indication for adjuvant therapy in small bowel NETs after carcinological surgery with complete removal of the tumour [17].

Treatment of functional NETs initially involves control of symptoms related to hormonal hypersecretion.

Medical treatment should be simple and highly effective and relies heavily on somatostatin analogues.

Systemic chemotherapy should be reserved for patients with unresectable and progressive metastatic disease.

4. CONCLUSION

Intestinal NETs are rare but their incidence is increasing.

They are manifested by clinical polymorphism behaviour and specific pathology.

Some intestinal NETs may secrete bioactive hormones which will be responsible for a carci noid syndrome.

Histological diagnosis is confirmed immunohistochemically by positivity of CgA and synaptophysin.

Current treatment modalities include surgical resection which is the mainstay of management of intestinal NET, symptomatic treatment, somatostatin analogue therapy and chemotherapy for unresectable metastatic stages.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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