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Case Report

A case report of primary small bowel neuroendocrine tumor with large metastatic lymph node at root of mesentery with tumor fibrosis

Saravana Kumar I.*, Gopikrishna M.

Department of Surgery, J. R. Medical College and Hospital, Tindivanam, Tamil Nadu, India

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***Correspondence:**
Dr. Saravana Kumar I.,
E-mail: sarsandivis12@gmail.com

ABSTRACT

Small bowel neuroendocrine tumors (NETs) are rare but are increasing in incidence advances in imaging techniques. NETs are a heterogeneous group of tumors with diverse clinical and imaging presentations. Although these tumors are typically smaller in size, they can cause significant fibrotic reactions in the bowel wall, lymph nodes, and mesentery. Due to the relatively rare occurrence of these tumors, limited awareness among physicians and nonspecific clinical symptoms, patients with small bowel NETs often experience considerable delays in diagnosis and inappropriate management. We present a case of a 58-year-old female who came with recurrent abdominal pain, distension, vomiting, significant weight loss and poor appetite. Although imaging findings were suggestive of a mesenchymal tumor, intraoperatively we could find a stricturising lesion in the ileum with a mesenteric mass. She underwent resection and anastomosis of the small bowel lesion along with mesenteric nodal excision. Interestingly, the postoperative biopsy revealed a well-differentiated Grade 2 intermediate-grade ileal NET with mesenteric nodal involvement. On immunohistochemistry (IHC), tumour cells show low Ki-67 index with positive for chromogranin and synaptophysin.

Keywords: Neuroendocrine tumor, Mesenteric fibrosis, Giant lymph node

INTRODUCTION

Neuroendocrine tumors (NETs) are rare epithelial neoplasms originating from enterochromaffin cells. The neuroendocrine tumors are rare slow-growing tumors with distinct histological, biological, and clinical behaviours. NETs accounting for approximately 2% of all malignancies with the gastrointestinal tract and lungs being the most common sites of occurrence.[1,2] Among gastrointestinal NETs, the ileum is the most frequent site, followed by the rectum and appendix.[3] NETs were first described by Langhans in 1867 from a polypoidal mass in small intestine.[4] In 1890, Ransom provided the first description of the carcinoid syndrome in a patient who experienced diarrhoea and dyspnoea aggravated by food and who, on autopsy, had diffuse hepatic metastases and a distal ileal mass.[5] NETs were first described by Langhans in 1867 from a polypoidal mass in small intestine.[4] In 1890, Ransom provided the first description of the carcinoid syndrome in a patient who experienced diarrhoea and dyspnoea aggravated by food and who, on autopsy, had diffuse hepatic metastases and a distal ileal mass.[5] In 1907, the term “karzinoide” was first used by Oberndorfer to describe a series of six patients who had small bowel tumors.[6] The behaviour of NETs differs by anatomic site and is very heterogeneous.[7] Nowadays relative increase in diagnosis of NET may be because of more frequent use of imaging and endoscopy.[10]Although the term "carcinoid" has historically been used to refer to cumulative symptoms associated with NETs, most patients with small bowel NETs initially present with nonspecific abdominal pain rather than symptoms of hormone excess.[14,15] The rarity of small bowel NETs, coupled with limited awareness among healthcare providers and non specific clinical symptoms, often leads to substantial delays in diagnosis. In this report, we report a rare case of a well-differentiated grade 2 intermediate-grade ileal NET with stricturising lesion causing sub acute small bowel obstruction who presented as mesenteric nodal mass.

CASE REPORT

A 58-year-old female presented to our emergency medical services on 08 November 2024, with complaints of generalized abdominal pain and distension. These symptoms were associated with multiple episodes of bilious vomiting over the past five days. Two days after the onset of vomiting, she developed loose stools, occurring 3–4 times daily. The abdominal pain was insidious in onset, aggravated by food intake, and relieved by vomiting. She reported a significant weight loss of over 10 kg in the past three years and a loss of appetite. The patient also gave a history of multiple similar episodes since 2021, requiring several hospital visits. She had previously been managed conservatively with nil per oral instructions, analgesics, and intravenous fluids at a local hospital.

On examination, the patient appeared dehydrated, though her vital signs were within normal limits. Her abdomen was distended with a resonant note on percussion, and a palpable, firm, tender mass with ill-defined margins was noted in the infraumbilical region. Laboratory investigations were within normal limits. An abdominal X-ray revealed dilated bowel loops (Figure 1). Contrast-enhanced computed tomography (CECT) of the abdomen and pelvis demonstrated a relatively well-defined, heterogeneously enhancing soft tissue density lesion measuring 3.7×3.2×2.4 cm with areas of necrosis or cystic change at the root of the mesentery near the umbilicus (Figure 2a). Coronal sections showed the lesion with mild surrounding desmoplastic reaction (Figure 2b). Posteriorly, the lesion abutted adjacent jejunal bowel loops with suspected infiltration, suggestive of a mesenteric gastrointestinal stromal tumor (GIST). Retrospective evaluation revealed suspicious eccentric mural thickening in one of the small bowel loops in the umbilical region within the lesion's drainage territory (Figure 2c).

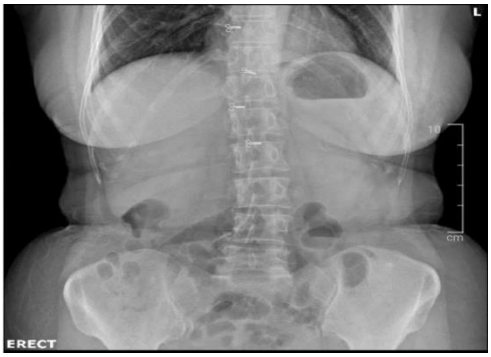

Figure 1: Plain X-ray abdomen erect.

An exploratory laparotomy with a midline incision was performed. Intraoperatively, a 5×4 cm mass was identified at the root of the small bowel mesentery near the superior mesenteric artery (SMA) and vein (SMV). The mass was hard, mobile, and located over the third part of the duodenum without infiltration (Figure 3). Additionally, a stricturizing lesion measuring 2×2 cm was found in the ileum, 40 cm from the ileocecal junction, in close proximity to the mesenteric lesion (Figure 4). A 1×1 cm firm, mobile lymph node adjacent to the SMA was also identified. The mesenteric mass was carefully dissected and separated from the SMA and SMV, and intraoperative ultrasound Doppler indicated desmoplastic reaction rather than vascular infiltration. En bloc resection, including 50 cm of the small bowel and mesentery, was performed to ensure complete nodal clearance. The resected segment included a 10 cm margin for the ileal mass, located 170 cm from the duodenojejunal flexure and 30 cm from the ileocecal junction (Figure 5). A hand-sewn, double-layer anastomosis was performed, and abdominal drains were placed before closing the abdomen in layers. The patient's postoperative period was uneventful.

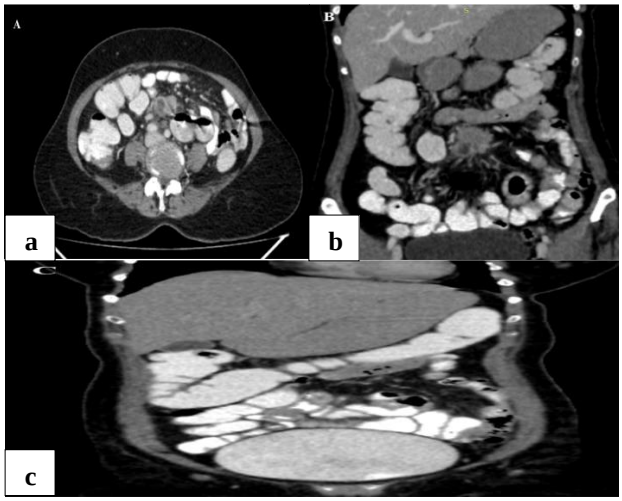

Figure 2: CECT abdomen with pelvis (a) axial section, and (b and c) coronal section.

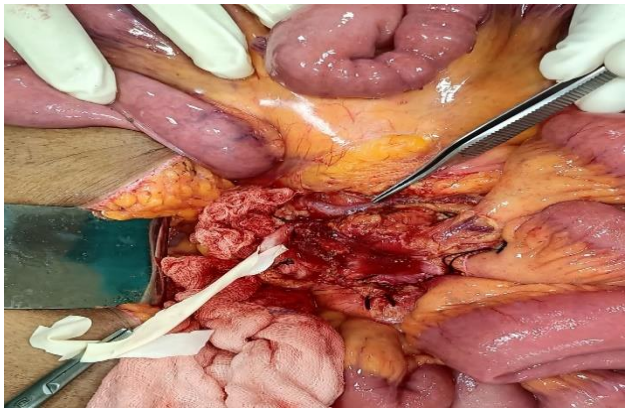

Figure 3: Mesenteric mass in relation to the superior mesenteric vein shown with tip of adson forceps and superior mesenteric artery by a vessel loop.

Histopathological examination of the resected specimen revealed an ill-defined, pale-yellow tumor measuring 2.5×1.5×1 cm within the bowel wall, with adequate resection margins (Figure 6a). The mesenteric mass, measuring 4×3.5×3 cm, appeared yellowish and firm and corresponded to a lymph node (Figure 6b).

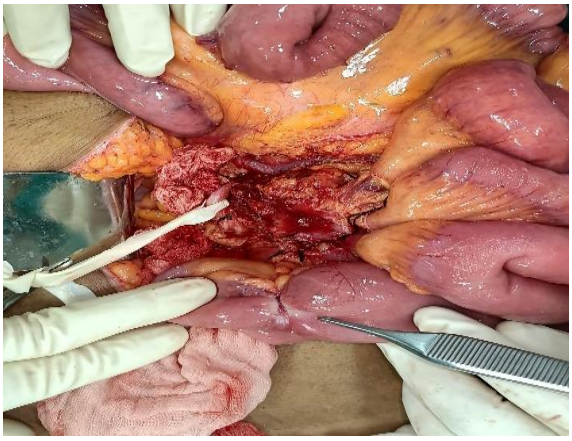

Figure 4: Small bowel mass shown with tip of adson forceps and close relation to mesenteric mass.

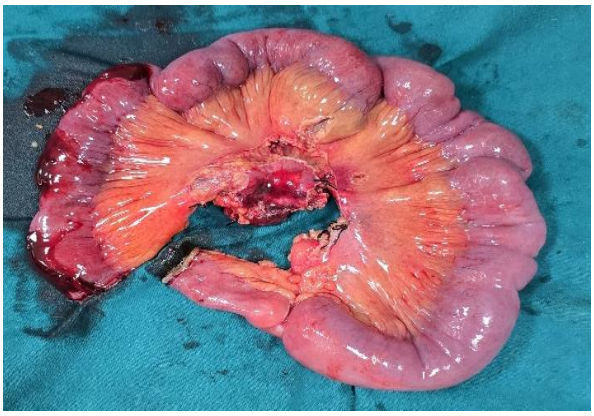

Figure 5: Resected specimen en-bloc showing mesenteric mass and small bowel mass with adequate gross margin.

Microscopic examination showed an infiltrative neoplasm in the submucosa composed of nests, cords, and trabeculae of tumor cells with abundant eosinophilic granular cytoplasm and round nuclei exhibiting salt-and-pepper chromatin (Figure 6d). Tumor infiltration of the muscularis propria was observed, along with lymphovascular and perineural invasion at multiple foci (Figure 6e).

The mitotic count was 8/mm^2. The mesenteric mass represented lymph node Involvement with complete effacement by tumor cells of similar morphology. The final histopathological diagnosis was a well-differentiated neuroendocrine tumor (grade 2) with lymphovascular and perineural invasion. Regional lymph node involvement was noted in 2 of 7 nodes, and the tumor was staged as pT2 pN2 (AJCC version). Immunohistochemistry (IHC) revealed a low Ki67 index, (Figure 7a and b) with tumor cells testing positive for chromogranin A (Figure 7c and d) and synaptophysin (Figure 7e and f). The case was discussed in a tumor board of our institution and a DOTANOC PET scan was advised to rule out subclinical lesions elsewhere, with further follow-up planned.

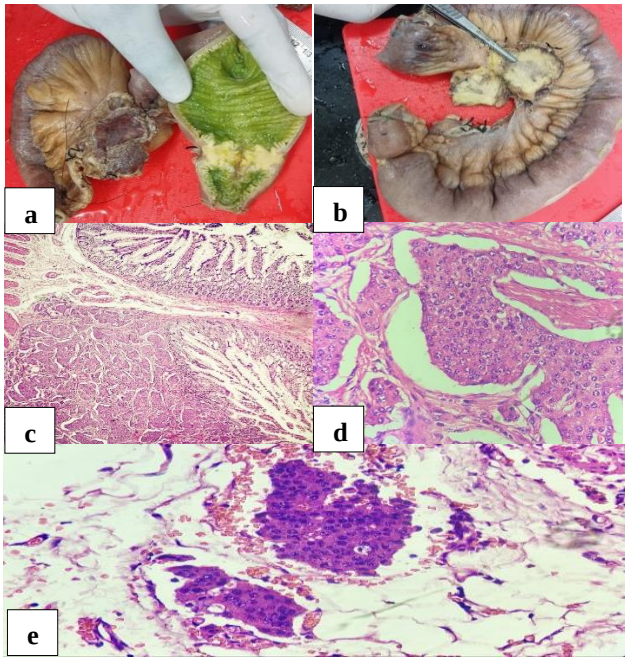

Figure 6: Gross features (a) cut surface of small bowel mass, (b) cut surface of lymph node mass, microscopic examination (100× magnification) showing small intestinal mucosa and tumor in submucosal location (haematoxylin and eosin), (d) microscopic examination (400x magnification) showing salt-and-pepper chromatin (haematoxylin and eosin), and (e) showing lymphovascular invasion of tumour cells.

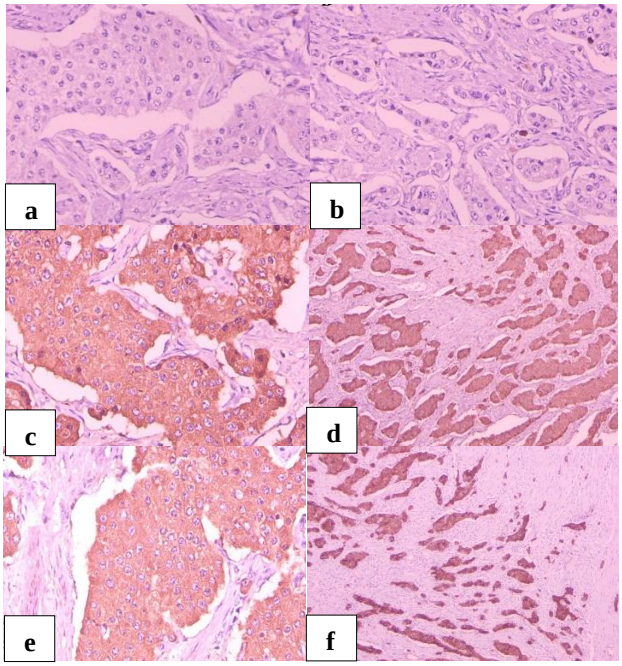

Figure 7: Immunohistochemistry (IHC) (a and b) microscopic picture of Ki 67 labelling index in 400x magnification is 8 per 2 mm^2, (c and d) microscopic examination showing tumour cells positive for chromogranin in 400x and 100x magnification respectively, and (e and f) microscopic examination showing tumour cells positive for synaptophysin in 400x and 100x magnification respectively.

DISCUSSION

In large Western studies, the median age at presentation of NETs is typically over 60 years. Although NETs are typically considered rare cancers, their rising incidence, coupled with their generally slow-growing nature, has led to a prevalence that surpasses that of many other gastrointestinal malignancies.[8] In a study using data from the SEER registry reported a 6.4-fold increase in the incidence of NETs since 1973 and increasing this trend primarily due to NETs in the rectum and small intestine, which are the two most common sites for primary gastroenteropancreatic NETs.[9] In a retrospective analysis of NETs in India, the most common primary tumor site was the stomach followed by the pancreas.[10] However, Kapoor et al reported as pancreas was the most common primary site.[11] These findings are consistent with those reported by Kulkarni et al.[12]

The relative increase in diagnosis of NET may be because of more frequent use of imaging and endoscopy.[9]Although the term "carcinoid" has historically been used to refer to both NETs and the syndrome, most patients with small bowel NETs initially present with nonspecific abdominal pain rather than symptoms of hormone excess.[13,14] Due to the relatively rare occurrence of these tumors, limited knowledge among physicians, and the nonspecific nature of early symptoms, patients with small bowel NETs often face significant delays in diagnosis.[15,16]

Due to smaller size, primary small bowel NETs are often difficult to detect on imaging. However, using targeted imaging protocols, such as a late arterial phase and contrast tomogram enterography can enhance the sensitivity of NETs. In CT NETs may have hyperenhancement in arterial phase, hairpin kink in the small bowel course and calcifications in most of the cases. Mesenteric metastasis can show a "spoke-like" appearance of mesenteric vessels due to fibrosis and the desmoplastic reaction induced by NETs.[17-19] In our case, imaging features were suggestive of mesenteric GIST due to presence of lobulated mass, minimal enhancement with solid/cystic areas inside.

Neuroendocrine cells typically express neural markers such as synaptophysin and chromogranin A. Plasma chromogranin A levels are elevated in 60-100% of patients with NETs, with sensitivity and specificity ranging from 70% to 100%. However, in cases of rapidly growing, poorly differentiated NETs, which often lose their characteristic structure and contain fewer secretory vesicles, the marker may not be released, resulting in false negative results. False positives can also occur in conditions like kidney disease, cardiovascular disease, and inflammatory bowel disease.[20,21]

The WHO classification of NETs has evolved over the past two decades and as per the most recent 2019 WHO classification NETs are graded as G1, G2, or G3, based on the mitotic rate and Ki-67 labelling index. The NETs are graded as - grade 1: as, low grade, having <2 mitoses/2 mm^2, Ki67 index: <3%; grade 2, as intermediate grade having 2-20 mitoses/2 mm^2, Ki67 index: 3-20%; and grade 3, as high grade having >20 mitoses/2 mm^2, Ki67 index: >20%.[22,23] Most neuroendocrine tumors of the small intestine are low-grade, characterized by indolent growth and a low proliferation rate. These tumors are often diagnosed at advanced stages with metastatic spread. In contrast, NEC are considered highly aggressive and have a poor prognosis. Metastasis is commonly observed in the liver, lymph nodes, lungs, and bones. Miller et al reported that NETs can have lymph node metastasis in 46.1% of G1 cases, 77.8% of G2 cases and 100% of G3 cases.[24] It is observed that, even smaller ileal NETs have a potential to spread to the regional lymph nodes.[25] A regional nodal spread has 5-year survival rate of 95%, while if distant metastasis is present then the survival rate is 67%.[9]

Most common true primary solid tumors of the mesentery include NETs, fibromatoses, neurofibromas, teratomas and germ cell tumors. While primary mesenteric NETs are very rare, secondary mesenteric involvement is more common, reported in 40–80% of cases.[26,27] Though neuroendocrine tumours are usually smaller in size, they can induce pronounced fibrotic reaction in the bowel wall, lymph nodes and mesentry.[28] They can be sometimes presenting like mesenteric mass or stricturising intestinal lesion. This was first described by Moertel et al in 1961.[29] This leads to misdiagnosis of NETs and leads to significant morbidity as a results of bowel obstruction and gut gangrene.

Morshitha et al described a rare case of a G2 NET involving the mesentery in a 54-year-old male, who underwent resection and anastomosis. The patient has been followed up for 1.5 years and reported no signs of recurrence.[30] He described about 4 different stages of mesenteric nodal metastasis with respect to superior mesenteric artery (SMA)/superior mesenteric vein (SMV) involvement. In our study, the mesenteric nodal mass had stage 1-2 without any SMA/SMV infiltration and we could able to resect the mass completely.

Surgery is the mainstay treatment for localized gastrointestinal NETs. Bowel resection combined with lymph node dissection is typically performed for NETs. In early stages I-III, R0 resection provides a curative approach, while in stage IV, it is usually palliative in nature to prevent local complications such as bowel obstruction and small bowel ischemia.[25,31,32]

CONCLUSION

Small bowel NETs can present as mesenteric nodal masses, which may resemble primary mediastinal neoplasms in imaging studies. Therefore, surgeons should remain vigilant and have a high level of suspicion for NETs during preoperative clinical assessment, imaging evaluation and intraoperative examination to ensure optimal patient management.

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