

Rare Case of Multifocal Ileal Neuroendocrine Tumour Clinically Presenting as Mesentric Mass

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Abstract: *Small intestinal neuroendocrine tumors (NETs) are generally slow-growing tumors and often multiple with a relatively high propensity for local and systemic metastases. We present a case of multifocal ileal NET with initial presentation as mesenteric mass.*

Keywords: NET, mesentric mass, ileum, multifocal

1. Introduction

Gastrointestinal NETs are typically low grade malignancies that arise from the diffuse neuroendocrine system scattered throughout the gut mucosa [1,2]. The first description of a small bowel NET was made by Langhans, in 1867, who described a polypoid tumor of the small intestine [3]. The term "karzinoide" was first used by Oberndorfer to describe a series of six patients who had small bowel tumors [4]. The incidence of small intestinal NETs has risen steadily over the past 30 years to 1.2 cases per 1,00,000 person years clinically. Jejunoileal NETs are multifocal (2-100 tumours) in at least 1/3rd of cases [5]. We present a very rare case of multifocal NET ileum who primarily presented with a mesentric mass.

2. Case Report

A 46 year old female presented with abdominal pain of 4 months duration with no associated features of obstruction or systemic manifestations. On examination there was abdominal guarding with no organomegaly. CT scan abdomen revealed a well defined rounded homogenous lesion in mesentery in umbilical region measuring 36x33mm with fat stranding (fig1) with suspicion of mesentric carcinoid.

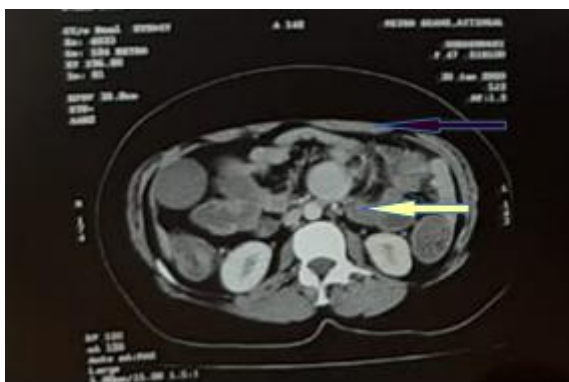


Figure 1: CT scan abdomen showing a well defined rounded homogenous lesion in mesentery

Subsequently patient underwent resection of small bowel with mesentery along with mesentric mass. Intraoperatively

surgeon identified structure involving 3cm length of ileum which was about 80 cm from ileocaecal junction (fig2).



Figure 2: Mesentric mass

Gross pathological examination revealed mesentric mass measuring 4.5 x4x2.5 cm which on cut section was yellowish solid and firm. Adjacent segment of small bowel showed multiple (9) similar yellowish submucosal nodules largest measuring 2.3x1.5x1cm (fig3).



Figure 3: Small bowel with multiple NETs along with mesentric mass

The mesentery also showed 6 lymph nodes largest measuring 1x1cm. Histopathology of mesentric mass and

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small bowel nodules showed neoplastic cells arranged in insular and trabecular pattern with monomorphic cells having moderate eosinophilic cytoplasm and nuclei with speckled chromatin (fig4and5).

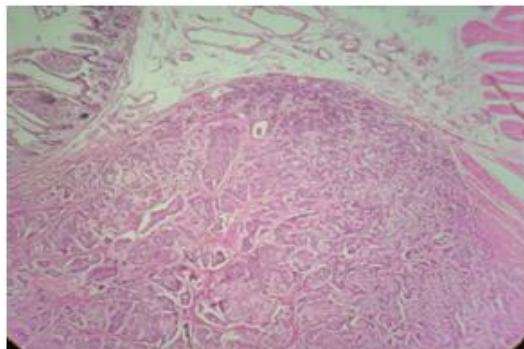


Figure 4: Neoplastic cells arranged in insular and trabecular pattern

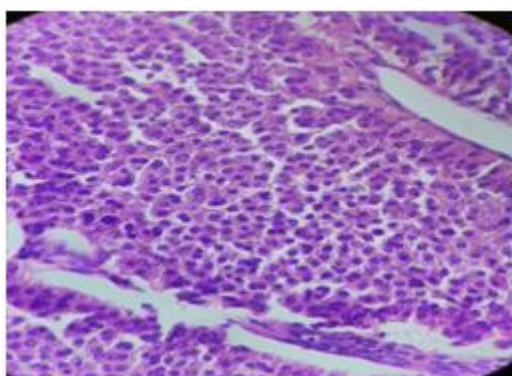


Figure 5: High power showing the speckled chromatin.

In addition the mesenteric mass showed dense fibrosis around the neoplasm with infiltration into surrounding fat and a focus of perineural invasion. The neoplasm is seen infiltrating in to the subserosa in the largest mucosal growth. The mitotic rate was 0-1 /10HPF. There was no necrosis. One out of 6 mesenteric lymphnodes show metastasis. So histopathological examination confirmed this case as well differentiated neuroendocrine tumour ileum grade 1, multifocal(9 in number) with infiltration in to subserosa and mesenteric mass measuring 4.5 cm in greatest dimension with 1/6 lymphnodes showing metastasis. The most recent WHO classification includes NET grade 1, NET grade 2, and NEC; these are distinguished from each other on the basis of proliferative index, which is assessed by the percentage of cells that stain positive for Ki-67, and mitotic rate. Hence T₃(_m)N₂M₀ - Stage III was assigned.

3. Discussion

46% to 64% of GIT carcinoid tumors arise in the midgut and most midgut carcinoid tumors originate in the terminal ileum [6]. Midgut carcinoid tumors commonly spread to the mesentery, reported as occurring in 40% to 80% of cases in various series [7]. Our patient presented with abdominal pain and CT scan revealed a mesenteric mass. On exploration surgeon identified a constriction in the ileum adjacent to mesenteric mass and only on gross pathology examination multiple mucosal nodules were revealed. Tumors of the small intestine are usually discovered after resection of the bowel

for symptoms of obstruction, or during exploration of the small intestine in search of a primary tumor after distant metastases have occurred [8]. So when mesenteric mass or lymph-node metastasis in the mesentery is suspected, we need to check the small intestine for primary lesions. [8] There was difficulty in diagnosis of mesenteric mass from a completely replaced lymphnode. Since the mesenteric mass has got irregular borders with invasion to fat and discontinuous from the primary neoplasm a diagnosis of mesenteric mass was preferred over lymph node metastasis.

MTDs were defined as discrete but irregular mesenteric tumor nodules frequently located adjacent to neurovascular bundles and discontinuous from the primary neoplasm; direct mesenteric extension from the primary tumor or extranodal extension of an involved lymph node was not considered as an MTD. Mesenteric deposits with a rounded contour or associated with a surrounding rim of lymphocytes were considered as LN metastases. MTDs are a strong predictor for liver metastasis and as a corollary, decreased disease-specific survival. In contrast, LN metastasis was not significantly associated with liver metastasis or disease specific survival. [9]

4. Conclusion

In our case the patient presented with a mesenteric mass and vague abdominal pain. Only gross pathological examination revealed multiple neuroendocrine tumours of ileum.

The recently published 8th edition of TNM classification acknowledges the importance of the number of lymph nodes metastasis and the presence of mesenteric mass, incorporating for the first time this information in the N category of the TNM system. According to the new classification, presence of mesenteric neoplastic mass measuring more than 2cm in maximum diameter corresponds to N2 category, even in the absence of lymph node metastasis [10]. It is imperative to know the significance of neuroendocrine tumours presenting as a mesenteric mass and its role in staging and prognosis.

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