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Rare Yet Relentless: Unusual Presentation of Jejunal Neuroendocrine Tumors - Case Reports

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ABSTRACT

Neuroendocrine tumors (NETs) are rare neoplasms arising from neuroendocrine cells primarily located in the gastrointestinal tract, pancreas, and lungs. Among gastrointestinal NETs, those originating in the jejunum are particularly uncommon. Their vague and nonspecific symptoms often delay diagnosis. Although these tumors are painless and slow growing, they have a metastatic potential, especially to the liver and lymph nodes. We report two rare and diagnostically challenging cases of jejunal NETs: one presenting as a hepatic abscess and another associated with markedly elevated serum alpha-fetoprotein (AFP). These cases emphasize the necessity of considering NETs in the differential diagnosis of atypical hepatic presentations and highlight the importance of advanced imaging and histopathological evaluation.

Keywords: Neuroendocrine tumor, jejunum, hepatic metastasis, alpha-fetoprotein, hepatic abscess.

INTRODUCTION

Neuroendocrine tumors (NETs) are rare neoplasms originating from neuroendocrine cells which are present in gastrointestinal tract, pancreas, and lungs [1]. Among gastrointestinal NETs, tumours of jejunum are uncommon and often present with nonspecific symptoms such as vague abdominal discomfort, anemia, which frequently leads to delayed diagnosis [2]. Despite their painless and slow growing nature, these tumors possess significant metastatic potential, particularly to regional lymph nodes and the liver [3]. Histologically, NETs are categorized into well-differentiated and poorly differentiated types. Well-differentiated tumors are graded based on mitotic count and Ki-67 proliferation index: Grade 1 tumors exhibit <2 mitoses/10 high-power fields (HPF) and

a Ki-67 index <3%, Grade 2 tumors show 2–20 mitoses/10 HPF or a Ki-67 index of 3–20%, and Grade 3 well-differentiated tumors exceed 20 mitoses/10 HPF with Ki-67 >20%[4,5,6,7]. Poorly differentiated neuroendocrine carcinomas, are aggressive high-grade neoplasms with mitotic counts >20/10 HPF and typically respond poorly to conventional therapies [6,7].

We report two diagnostically challenging cases of jejunal NETs with rare presentations: one case presenting as a hepatic abscess, and another associated with markedly elevated serum alpha-fetoprotein (AFP), a tumor marker not typically expressed by neuroendocrine neoplasms. These cases highlight the heterogenous picture of jejunal NETs and the necessity of including them in differential diagnoses for atypical hepatic lesions or elevated AFP levels.

Management of jejunal NETs depend on its stage and grade [8,9]. For localized Grade 1 and 2 tumors, surgical resection with regional lymphadenectomy is the curative treatment [12]. In cases of metastatic disease, surgical intervention is indicated, particularly if the primary tumor causes local symptoms or complications [10,11]. For unresectable or advanced disease, medical management include somatostatin analogues (octreotide or lanreotide) for symptom control and tumor stabilization, liver directed therapies such as transarterial chemoembolization or radiofrequency ablation for hepatic metastases, and peptide receptor radionuclide therapy (PRRT) for somatostatin receptor-positive tumors[12,13]. Systemic chemotherapy like capecitabine-temozolomide combinations for well-differentiated NETs or platinum-based regimens (cisplatin or carboplatin with etoposide) for high-grade NECs are used in advanced cases [14]. Imaging modalities, like contrast-enhanced MRI, somatostatin receptor PET/CT (68Ga-DOTATATE PET) and immunohistochemical markers such as synaptophysin and chromogranin A, have significant role in tumor staging and characterization [15,16]. In this case series, we aim to highlight two atypical presentations of jejunal NETs and emphasize the importance of a high index of suspicion and a multidisciplinary approach for timely diagnosis and optimal management.

RESULT

CASE 1:

A 63-year-old male, a known case of Type 2 diabetes mellitus, hypertension, and obstructive sleep apnea, presented with fever, cough, breathlessness, and generalized tiredness for one week. On examination he was febrile, tachypnoeic, rhonchi present over infra scapular and infra mammary areas on both sides and had tenderness on right hypochondrium. Blood investigations showed elevated inflammatory markers. A chest X-ray showed bilateral reticulonodular opacities on both sides (Figure 1). He was started on intravenous antibiotics, nebulization and other supportive measures. However, as abdominal pain and cough were persisting, CT thorax with abdomen cuts was taken which showed a well circumscribed hypodense lesion of size 8 cm x 7.5 cm in segment VIII of liver which was suggestive of hepatic abscess (infective etiology), necrotic multicentric hepatocellular carcinoma (malignant etiology). To differentiate between the two, further imaging and laboratory tests was done. Ultrasound of abdomen showed multiple hyperechoic lesions in both lobes of liver with areas of liquefaction (Figure 2). Laboratory tests for infectious work up like blood culture, amoebiasis IgG antibodies, were negative. Tumor markers like alpha fetoprotein, carcinoembryonic antigen, carbohydrate antigen 19-9 were negative. A contrast enhanced CT scan abdomen showed peripherally enhancing lesion of size 10 x 8.8 x 8 cm in segment VIII of liver with possibility of abscess (Figure 3). A pigtail catheter drainage for the abscess was initially considered but later deferred due to increased vascularity of the lesion, instead a USG guided biopsy of lesion was done for histopathological evaluation (Figure 4).

Biopsy showed sinusoidal trabecular pattern with hyperchromatic nuclei favouring cholangiocarcinoma (Figure 5) and rosetting of cells favouring of neuroendocrine tumour (Figure 6). An immunohistochemistry staining was done, for which CK -7 was negative (Figure 7) and chromogranin A was positive (Figure 8). The possibilities from biopsy were cholangiocarcinoma with neuroendocrine differentiation, mixed hepatocellular-cholangiocarcinoma, metastatic neuroendocrine carcinoma. A whole body 18-flurodeoxyglucose positron emission tomography (PET-CT) scan revealed somatostatin receptor expressing lesion in proximal jejunum representing a primary malignancy (Figure 9) with hepatic metastasis (Figure 10). From a clinical picture of lower respiratory tract infection with hepatic abscess, patient was diagnosed with jejunal neuroendocrine tumor with liver metastases and was started on octreotide therapy, leading to symptomatic improvement.

Figure 1: Chest X ray showing bilateral reticulonodular opacities.

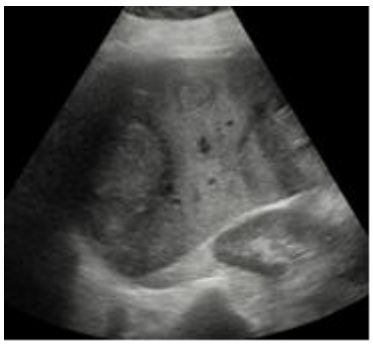

Figure 2: USG abdomen showing multiple lesions in liver.

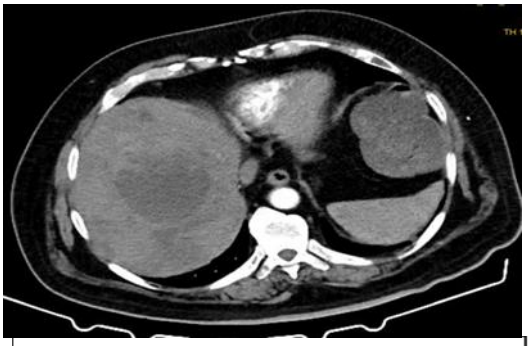

Figure 3: CECT abdomen showing peripherally enhancing lesion in liver.

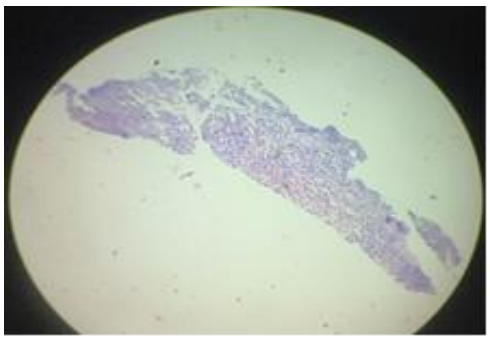

Figure 4: Biopsy of the core tissue

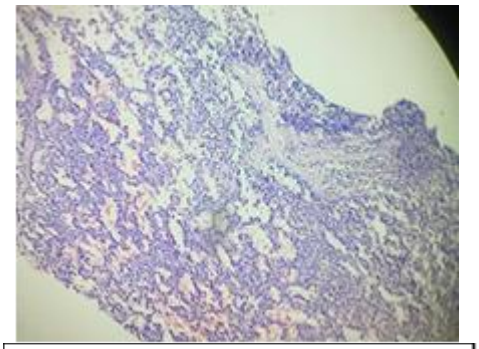

Figure 5: Scanner view of tissue, sinusoidal pattern of arrangement

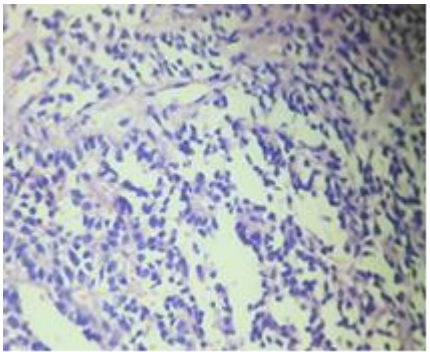

Figure 6: Cells with hyperchromatic nuclei and rosetting pattern

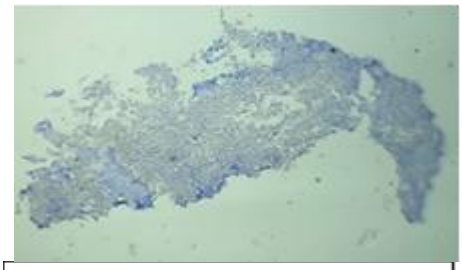

Figure 7: IHC stain, CK 7(-)

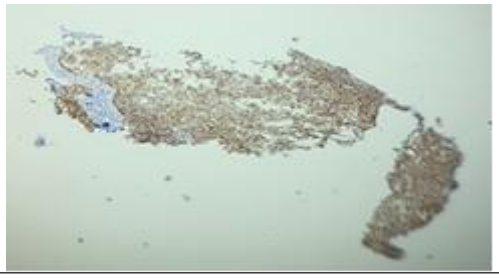

Figure 8: IHC stain, chromogranin A (+)

Figure 9: Somatostatin receptor expression in proximal jejunum

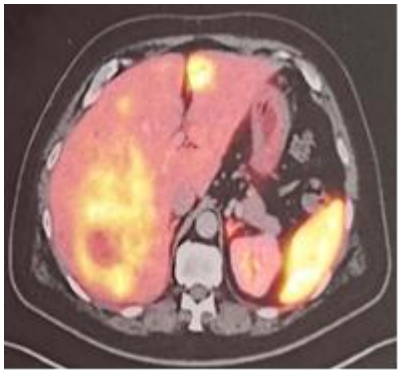

Figure 10: Somatostatin receptor expressing hepatic metastasis

CASE 2:

A 62-year-old male known case of type 2 diabetes mellitus, hypertension presented with lower back pain, weight loss of 1 month duration. On examination tenderness over right hypochondrium. Laboratory findings revealed neutrophilic leucocytosis, elevated ESR, and altered liver function tests. X ray spine was taken to rule out lytic lesions (Figure 10). Multiple myeloma was ruled out through serum protein electrophoresis and urine Bence Jones protein analysis. Out of the tumour markers sent, alpha feto protein was significantly elevated (>15000), gamma glutamyl transferase was slightly elevated and rest of the markers like carcinoembryonic antigen, carbohydrate antigen 19-9 were negative. Contrast enhanced computed tomography of (CECT) abdomen was done which showed hepatomegaly with multiple scattered circumscribed peripherally enhancing mildly hypodense lesions of size 7.4 cm x 3.4 cm in liver parenchyma which was suggestive of multicentric hepatocellular carcinoma (Figure 11). Magnetic resonance cholangiopancreatography (MRCP) was done to trace common bile duct and it showed ill-defined mild delayed enhancing lesion in periampullary region- likely ampullary malignancy (Figure 12,13). Over the course patient had constipation and worsening of liver function test including gamma glutamyl transferase, Cholangiogram revealed a distal common bile duct stricture. A self-expandable metallic stent was placed. Later an endoscopic retrograde cholangiopancreato-graphy was performed and an ampullary biopsy was taken. Histopathology from jejunal lesion had possibility of small cell neuro endocrine tumour. PET CT scan showed multiple liver lesions with soft tissue density lesion in peri-ampullary region and primary lesion from jejunum. Immunohistochemical analysis showed positivity for synaptophysin, chromogranin A confirming neuroendocrine tumour of jejunum with metastatic spread. Patient was

initiated on chemotherapy-etoposide and carboplatin, and is now doing well.

Figure 10: Xray spine

Figure 11: CECT abdomen showing multiple lesions on liver parenchyma

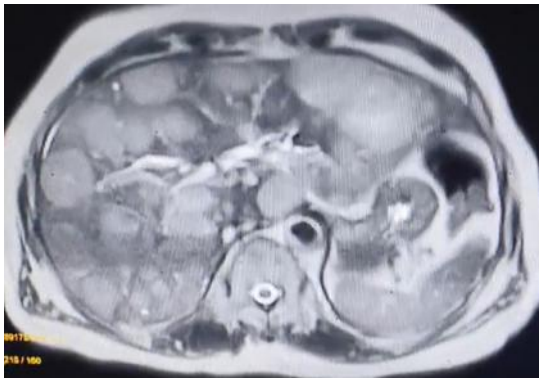

Figure 12: MRCP showing multiple lesions on liver lesion

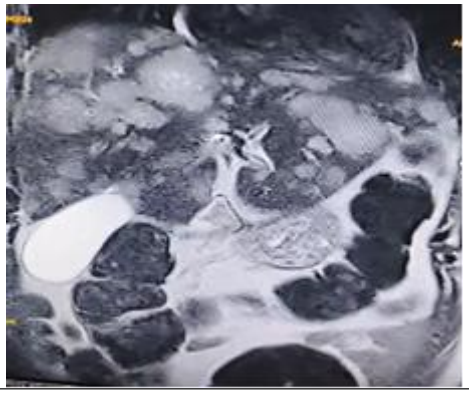

Figure 13: MRCP showing enhancing lesion in periampullary region

Figure 14: Ulcerated ampulla

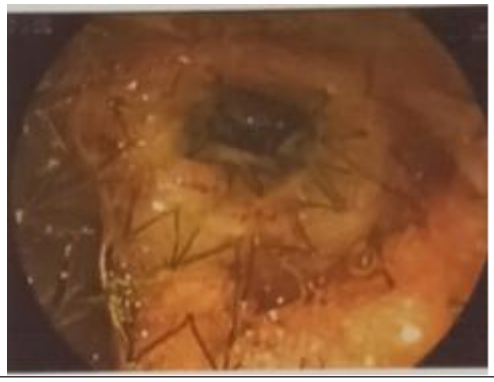

Figure 15: self-expandable metallic stent

DISCUSSION

In our case series, patient presented with atypical symptoms that was suggestive of conditions like lower respiratory tract infection with hepatic abscess and musculoskeletal pain. These nonspecific presentations highlight the challenges in recognizing NETs.

Case 1: Neuroendocrine Tumor Presenting as Hepatic Abscess

Neuroendocrine tumors presenting as a hepatic abscess are rare and has a diagnostic challenge. Patient presented with features of lower respiratory tract infection. However, lack of response to antibiotics and persistent systemic symptoms prompted further evaluation which confirmed a well-differentiated metastatic NET. This unusual presentation highlights the importance of considering malignancy in atypical or non-resolving hepatic abscesses, in patients with no clear infectious source [17,18,19]. Neuroendocrine markers like synaptophysin, chromogranin A, biopsy and histopathology of the lesion helped to confirm the neuroendocrine origin.

Case 2: Neuroendocrine Tumor with Elevated Alpha-Fetoprotein (AFP)

Neuroendocrine tumors (NETs) typically do not secrete alpha-fetoprotein (AFP), a marker more commonly associated with hepatocellular carcinoma, yolk sac tumors, and certain germ cell malignancies. In our case, a jejunal NET with elevated AFP represented an unusual finding [20,21,22]. This atypical biochemical profile highlights the importance of considering NETs in the differential diagnosis of AFP-elevated malignancies [23]. Immunohistochemistry was positive for synaptophysin and chromogranin A, confirming neuroendocrine tumour.

In the above cases, advanced imaging techniques like 18-flurodeoxyglucose positron emission tomography (PET-CT) and immunohistochemistry played a role in locating the tumor, extent of disease, and metastatic spread. The management of jejunal neuroendocrine tumors (NETs) requires a multidisciplinary and individualized approach, guided by tumor grade, stage, symptoms, and presence of metastasis. In cases of localized tumors, surgical resection is considered which involves segmental small bowel resection with mesenteric lymphadenectomy [24,25,26]. For advanced or metastatic disease, particularly with hepatic involvement, treatment include somatostatin analogues like octreotide, lanreotide as first-line therapy, peptide receptor radionuclide therapy with radiolabelled somatostatin analogues in case of inoperable, progressive, or metastatic disease with high somatostatin receptor expression. Other options include targeted therapies such as everolimus, liver-directed therapies (embolization, ablation), cytoreduction in hepatic metastases. Chemotherapy is generally reserved for high-grade or poorly differentiated NETs. In our case, first patient improved on octreotide therapy, and second patient improved with cisplatin and etoposide therapy without the need of any surgical intervention.

CONCLUSION

These case reports highlight rare presentations of jejunal neuroendocrine tumors with elevated alkaline phosphatase and hepatic lesions mimicking liver abscesses. Such atypical biochemical and radiological findings can easily lead to misdiagnosis and treatment. A high index of suspicion for NETs can be considered in patients with hepatic abnormalities unresponsive to conventional therapy.

Declaration by Authors

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Conflict of Interest: The authors declare no conflict of interest.

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