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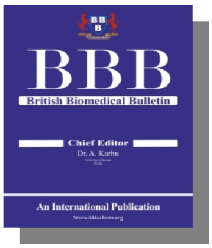

Case Report

Unusual Presentation of Jejunal Neuroendocrine Tumor with Cystic Hepatic Metastasis

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A R T I C L E I N F O

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A B S T R A C T

Neuroendocrine tumors are derived from primitive stem cells in the gut wall, but also can be seen in other organs. Most Neuroendocrine tumors are slow growing and indolent without symptoms. Nevertheless, aggressive and metastatic disease (e.g., in the brain) does occur. Here we report a case of jejunal neuroendocrine tumor with cystic metastasis in liver presented with progressive right sided abdominal distension.

Introduction

Carcinoid tumors are of neuroendocrine origin and derived from primitive stem cells in the gut wall, but they can be seen in other organs[1]. The Jejunal neuroendocrine tumor is midget tumor. Midgut tumors are argentaffin positive and can produce high levels of serotonin, 5-hydroxytryptamine (5-HT), kinins, prostaglandins, substance P (SP), and other vasoactive peptides[2]. On CT, hepatic metastases of neuroendocrine tumors are hypervascular-enhancing lesions and may assume a variety of patterns; such as finely nodular, coarsely nodular, mixed single large mass and rarely pseudocystic appearance, which probably is due to ischemic necrosis[3]. At the time of diagnosis, 58%-64% of patients with small intestinal neuroendocrine tumors have regional lymph node or liver metastasis[4].

Case Report

We present a 55 year old woman with chief complaints of progressive distension of the abdomen and pain for last one month. Physical examination revealed hepatomegaly and mass arising from right hypochondrium to the epigastrium. Blood analysis revealed anaemia (Hb-4g/dl, RBC (3.24) with normal leukocyte counts. Serology for hydatid disease was negative. In view of the deranged coagulation profile, the patient was not planned for percutaneous liver biopsy.

On abdominal ultrasound, multiple echogenic mass lesions were found to have cystic components, of which the biggest one was in the right lobe of liver measuring 10x7x7cms. Abdominal CT revealed heterogeneously enhancing hepatic mass lesions with areas of central necrosis and lobulated contours **(Fig 1)**. With these findings, differential diagnosis of multiloculated hydatid liver, polycystic liver disease or cystic liver metastasis with unknown primary was made. All procedures like EGD, colonoscopy, and enteroclysis / small bowel series were normal. Exploratory laparotomy was planned and hepatic resection of the right lobe and segment V was performed. Peroperatively, jejunal extra mucosal mass 2.3 cm x 1.0 cm was identified and resection was done. A look for lymph nodes proved to be negative. Gross examination of the right liver lobe and segment V, measuring 10cm x 7cm x 7cm revealed multiple grey white nodules ranging from 3mm to 2 cm in size on the surface and the liver was grossly lobulated **(Fig 2)**. On cut section, the liver parenchyma was a cystic mass, cysts ranging in size from 1cm to 5cm filled with hemorrhagic degenerative material, which were lined by a thin rim of liver parenchyma. Jejunal tumor was grossly 2.3cm overlying mucosa was unremarkable. On cut section, it was grey white, limited to subserosa. **(Fig 3)**.

Histopathologically jejunal mass showed normal mucosal architecture with underlying sheets, trabeculae, tubules and rosette like pattern of cells. Tumour cells were round or polygonal with central nucleus, punctate chromatin and infrequent mitosis. The cytoplasm was eosinophilic and capillaries were prominent **(Fig 4)**. Immunohistochemically, the tumour was positive for Chromogranin **(Fig 5)**. Liver histology also revealed metastatic deposits of neuroendocrine tumor with extensive areas of hemorrhage and necrosis. As per WHO classification, the tumor was classified as "Well differentiated neuroendocrine carcinoma".

Discussion

Primary malignant tumors of the small intestine are uncommon and Gastrointestinal Carcinoid (Carcinoid Tumor) is the most common primary tumor of the

small bowel accounting for more than 95% of all carcinoids[4]. The age-adjusted incidence of carcinoid tumors worldwide is approximately 2 per 100,000 persons[5,6]. The average age at diagnosis is 61.4 years[5]. Carcinoid tumors represent about 0.5% of all newly diagnosed malignancies[5,7]. The tumor arises from the enterochromaffin cells of Kulchitsky. Various sites of origin of this neoplasm are appendix - 30-45%, small bowel - 25-35% (duodenum 2%, jejunum 7%, ileum 91%, multiple sites 15-35%), rectum 10-15%, caecum - 5%, and stomach - 0.5%[8]. Our patient was 55 years old, and presented with progressive right sided abdominal distension and pain. Since Jejunal Neuroendocrine tumor is derivative of midgut and is argentaffin positive and produce increased levels of serotonin, 5HT, kinins, Prostaglandin, substance P, and can produce Neuroendocrine syndrome, when the liver or retroperitoneal nodes are involved. Usually Neuroendocrine tumors of the small bowel behave in a malignant fashion producing lymph node or/and liver metastasis. The small bowel neuroendocrine tumor is slow growing and is missed mostly on radiography. Barium studies (enteroclysis, follow through examination) may show intramural or intraluminal filling defects in the involved portion of the gut. There can also be narrowing of the lumen with stricture formation, thickening of the valvulae conniventes and increase in the inter bowel loop distance due to wall thickening. None of these features were present in our case. It was totally an extra mucosal mass with normal looking jejunal mucosa. Diagnosis can be made confidently on CT. It reveals a soft density mass with speculated borders and radiating strands with or without calcification in 80% of cases[9]. In our patient ultrasound revealed liver studded with multiple masses only and CT demonstrated multiple cystic masses within the liver parenchyma. Rest of the abdominal organs were abnormal and no jejunal mass was demonstrated. Patient underwent laparotomy for huge liver mass and jejunal mass (2.3 cm x 1cm) was found incidentally, which histopathologically proved to be neuroendocrine tumor.

The radiological appearance of neuroendocrine tumors varies according to size of tumor, extent of mesenteric involvement, nodal infiltration or liver metastases. Usually small Neuroendocrine tumors located in the mucosa and submucosa of the intestinal wall are best evaluated with enteroclysis or small bowel series, but in many cases, usually small nodular neuroendocrine tumors, which are submucosal without mucosal involvement, may be very difficult to detect with a conventional small bowel series. They are best demonstrated on gadolinium enhanced T1-weighted MR images obtained with fat suppression, where they manifest as nodules or focal areas of mural thickening with moderately intense gadolinium enhancement[10].

Conclusion

The primary jejunal neuroendocrine tumor is rare entity and cystic liver metastasis without producing any neuroendocrine syndrome is very rare. Small submucosal small bowel neuroendocrine tumor without mucosal and nodal involvement, but with extensive cystic liver metastasis may also masquerade various benign and malignant cystic hepatic masses.

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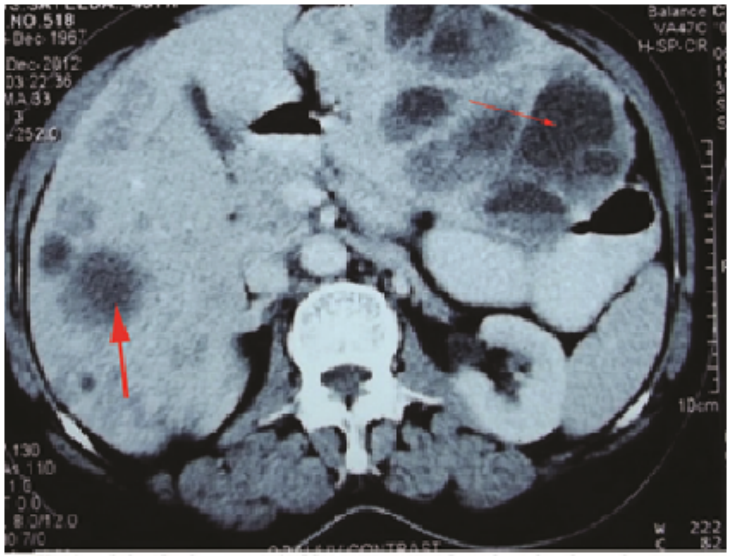

Figure 1. Abdominal CT revealed heterogeneously enhancing hepatic mass lesions with areas of central necrosis and lobulated contours

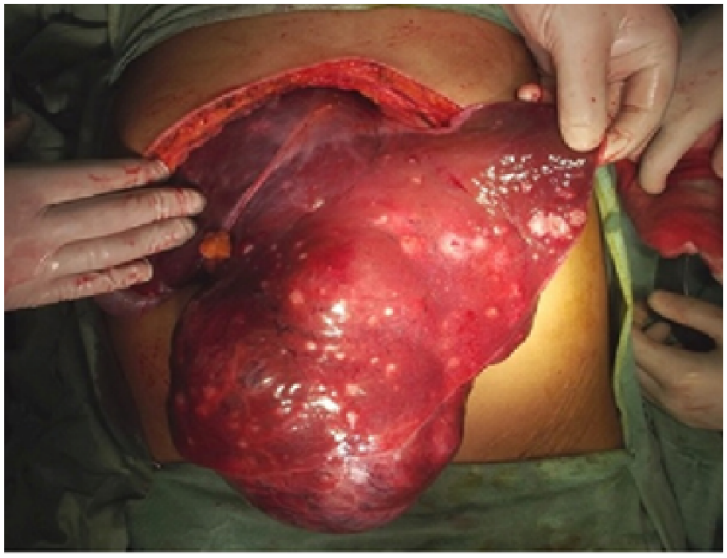

Figure 2. Gross examination of right liver love and segment V, revealing multiple grey white nodules ranging on the surface and the liver, grossly lobulated

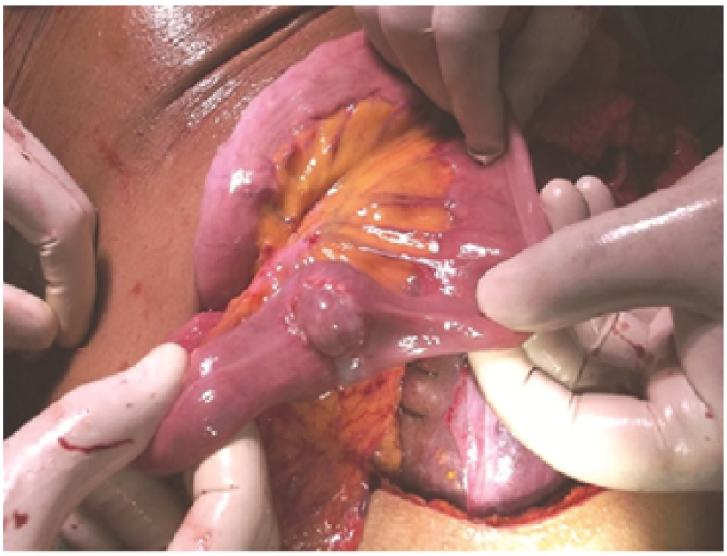

Figure 3. Peroperative picture of jejunal tumor (2.3 cm) of the patient, limited to subserosa

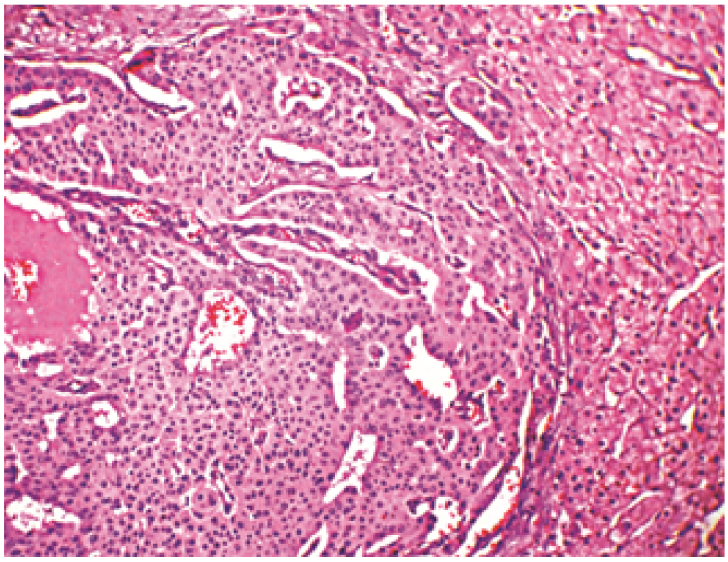

Figure 4. Metastatic deposits of neuroendocrine tumor in the liver

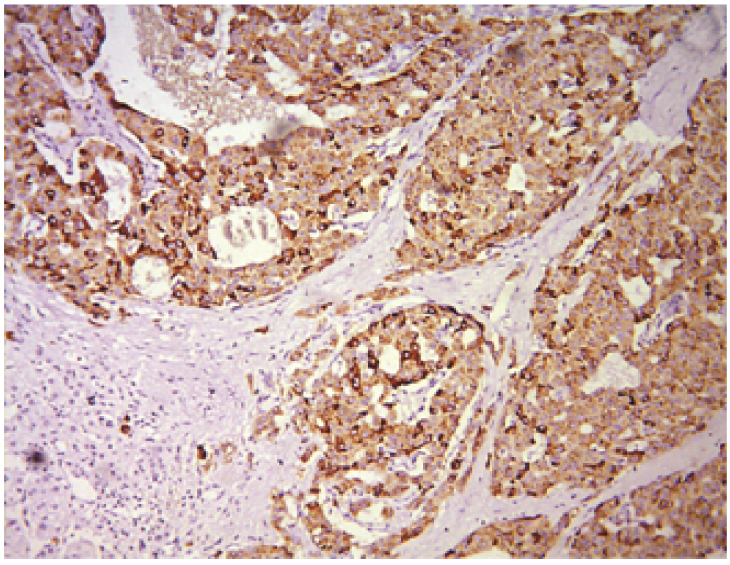

Figure 5. Immunohistochemical marker (chromogranin A), positive for tumor neuroendocrine