

Retroperitoneal Non Functional Neuroendocrine Tumor Of Pancreas Encircling Duodenum Without Obstruction

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Citation

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Abstract

A 50-year-old lady with the history of generalized weakness and abdominal discomfort for one month was investigated in a local hospital and found to have severe anemia with a hemoglobin level of 3g/dl. She was treated with multiple blood transfusions. Upon admission to our hospital, the patient continued to have abdominal discomfort. On examination there was a single ill-defined mass palpable in the right hypochondrium and right lumbar area. CT of the abdomen revealed a multilobulated mass lesion in the right paraspinal location extending superiorly from the porta hepatics to the aortic bifurcation inferiorly, located anterior to the inferior vena cava and right kidney. There was extrinsic compression of the second and third part of the duodenum in upper GI scopy. On exploratory laprotomy there was a mass in the retroperitoneum posterior to the second and third part of the duodenum, anterior to the inferior vena cava and right kidney extending from porta hepatis to right iliac fossa. Part of the tumor was encircling the third part of the duodenum for about 270 degrees, displacing the uncinate process and the head of the pancreas superiorly. The mass was carefully separated from duodenum and pancreas with sharp dissection, and the whole mass was excised completely. Histopathology of the mass was reported as pancreatic neuroendocrine tumor.

INTRODUCTION

Endocrine tumors of the pancreas belong to the broad group of neuroendocrine tumors that are located in a variety of locations. They occur infrequently with an incidence of 1 to 5 per million annually. (1, 2) Traditionally, they were thought to arise from APUD (amine precursor uptake decarboxylase) cells. (3, 4) More recently, studies suggest that they arise from pluripotent stem cells in the pancreatic ductal and acinar epithelium. (5)

They manifest with well-defined clinical syndromes due to hormones they secrete like hypoglycemia attributed to insulinoma, the fulminant peptic ulcer disease of Zollinger-Ellison syndrome, the watery diarrhea, hypokalemia, and achlorhydria of Verner-Morrison syndrome, the diabetes and skin rash of glucagonoma, or the steatorrhea and cholelithiasis of somatostatinoma. Some endocrine tumors have no clinical evidence of hormone secretion and they are called non-functioning tumors. (6) These tumors are being increasingly identified and diagnosed because of widespread abdominal imaging.

There are no histological characteristics that distinguish benign from malignant tumors; identification of malignancy requires evidence of local invasion or distant spread. (7, 8)

Even the malignant tumors have an indolent course.

Pancreatic endocrine tumors are associated with MEN-type I, Von Hippel-Lindau disease, (9), Von Recklinghausen's disease (10) and tuberous sclerosis (11).

CASE SUMMARY

A 50-year-old lady presented with pain in the right upper quadrant of the abdomen for one month. The pain was dull aching, continued, mild to moderate in severity, not radiating and not related to food. There were also complaints of generalized weakness for one month, but there was no history of fever, vomiting, jaundice, abdominal distention, loss of weight, and loss of appetite. Bowel and bladder habits were normal. Two months earlier, the patient had been diagnosed as anemic and treated with multiple blood transfusions.

On examination there was a single mass palpable occupying the right hypochondrium, right lumbar, and umbilical area, about 15x12cm. The upper border was extending under the subcostal margin; the lower border was extending up to the level of the iliac crest. The mass was non-tender, firm to hard in consistency; the surface was nodular, and not moving with respiration.

All her routine blood investigations were within normal

limits except albumin which was 2.9mg/dl. CT of the abdomen revealed a large single multilobulated mass lesion measuring about 15x10x6cm in a right paraspinal location extending from the inferior surface of the liver from the level of the porta hepatics to vertebra L4. The mass was located anterior to the right kidney and IVC. (Figure 1). The mass was extending posterior to the second and third part of the duodenum and lifting the uncinate process and head of the pancreas anteriorly (Figure 1, 2). The mass appeared to compress the IVC abutting the portal vein and superior mesenteric vein (Figure 2, 3). The mass was highly vascular receiving blood supply from the superior mesenteric artery and with venous drainage to the portal vein. There was no evidence of metastatic deposits in the liver, ascites or regional lymphadenopathy. The findings were suggestive of GIST, paraganglioma, and lymphoma.

Figure 1

Figure 1: Abdominal CT: Rt. kidney and superior mesenteric vein

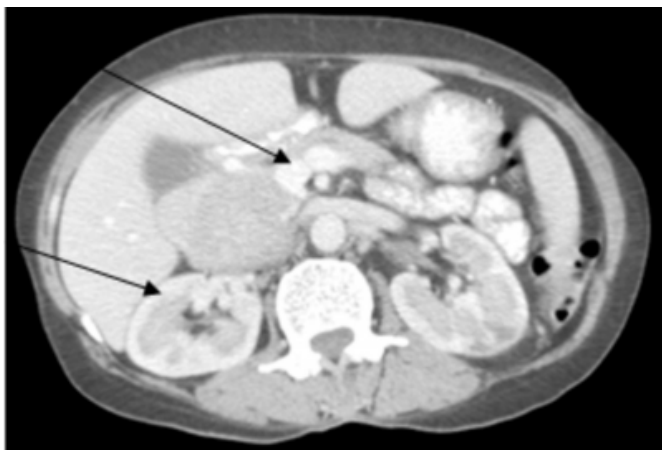


Figure 2

Figure 2: Abdominal CT: IVC and aorta

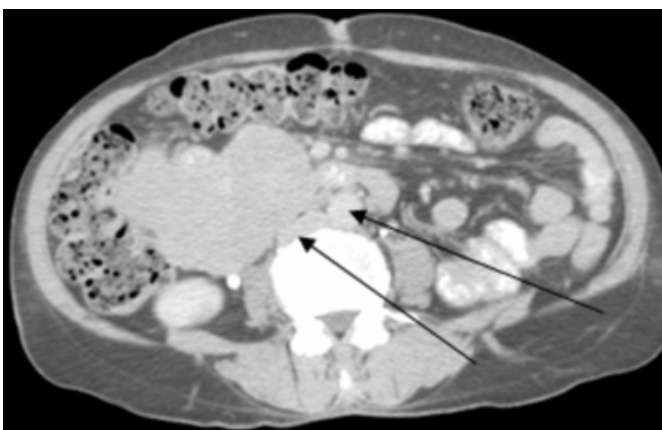
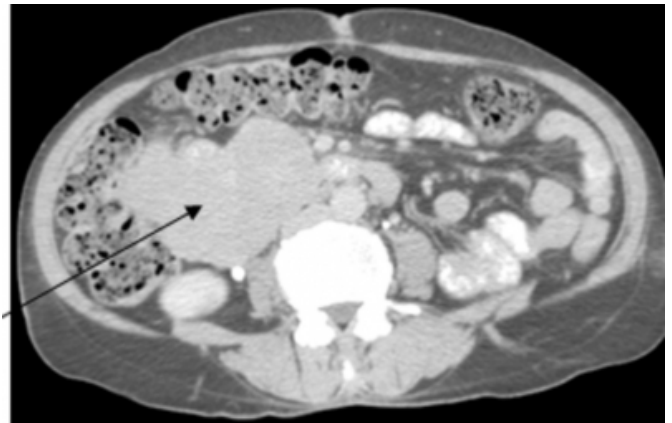


Figure 3

Figure 3: Abdominal CT: Retroperitoneal tumor



On exploratory laparotomy, the tumor was mobile and we could easily separate it from the IVC and right kidney; anteriorly, the second and third part of the duodenum was looking completely engulfed in the tumor. As we carefully separated the duodenum from the tumor by sharp dissection, it was found that a part of the tumor was encircling the third part of the duodenum to about 270 degrees and separating the duodenum from the uncinate process (Figure 4, 5). The uncinate process and head of the pancreas was pushed anteriorly and upwards (Figure 6). With careful sharp dissection the tumor was separated from the duodenum, pancreas, and portal vein and excised completely. Histopathology after special stains and immunohistochemistry, which was positive for synaptophysin, chromogranin, and focally positive for cytokeratin, was suggestive of non-functional neuroendocrine tumor of the pancreas.

Figure 4

Figure 4: Tumor encircling duodenum

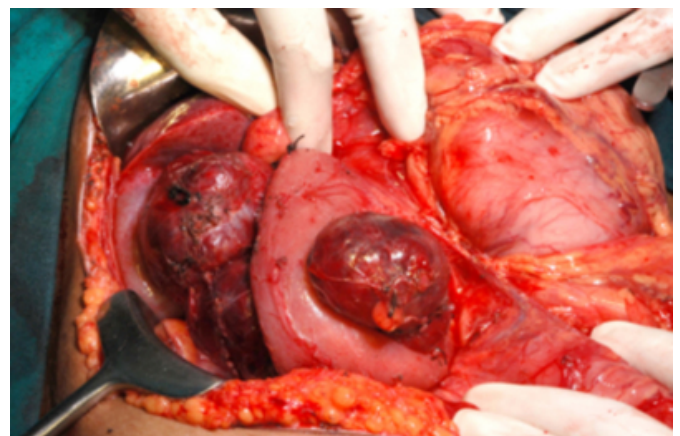


Figure 5

Figure 5: Tumor separated from porta hepatis

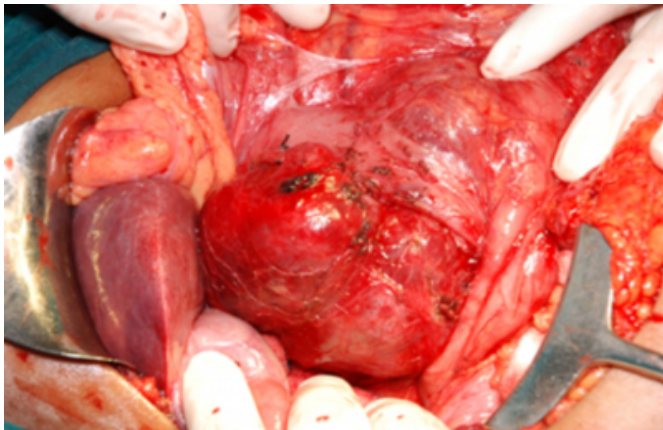
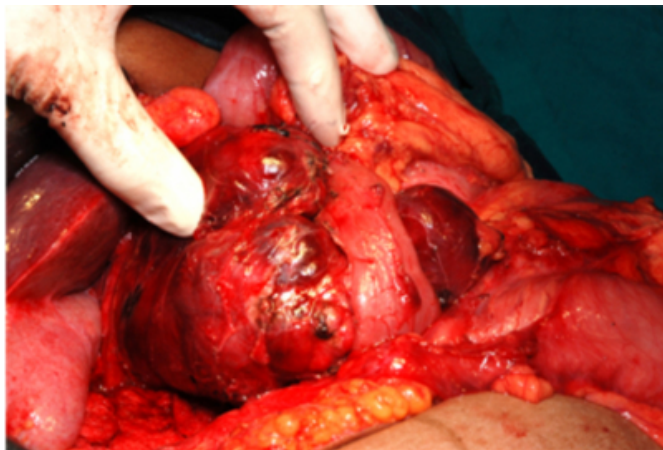


Figure 6

Figure 6: Tumor separated from IVC and right kidney



DISCUSSION

Non-functioning neuroendocrine pancreatic tumors (NFNEPT) may be the most common pancreatic endocrine tumors (12), usually seen in the fourth and fifth decade, with equal incidence in males and females. They usually present as large tumors and 60 to 80% metastasize at the time of presentation (13).

NFNEPTs do not secrete any known active hormones. They are often asymptomatic and may present with abdominal pain, weight loss and jaundice. These tumors can secrete a variety of hormones including pancreatic polypeptide, neurotensin, and calcitonin, and proteins like neuron-specific enolase, and chromogranin A. These hormones do not produce any symptoms. They have high degree of vascularity, cystic degeneration, and calcification on imaging. EUS with biopsy will confirm the diagnosis. NFNEPT usually presents at an advanced stage such as large

mass, gross local invasion and distant metastases, and are often misdiagnosed as pancreatic exocrine tumors (14).

Surgery remains the treatment of choice with a high rate of resectability reported up to 60%. NFNEPTs are usually treated by pancreaticoduodenectomy for tumors of the head of the pancreas or by distal pancreatectomy for tumors of body and tail. Debulking of the tumor and resection of hepatic metastasis is advised as it increases the disease-free survival rate (14).

CONCLUSION

As this patient was not having clinical evidence of secretion of any active hormones, this tumor was diagnosed as non-functioning neuroendocrine tumor of the pancreas; these are the most common NEPT. As these tumors do not secrete any active hormones, they usually present as large tumors with metastasis, in contrast to our patient: even though the tumor was of large size, there was no evidence of metastasis, and a neuroendocrine tumor of the pancreas encircling the duodenum with no evidence of obstruction has never been described earlier. The natural course of these tumors is usually indolent and complete surgical excision is the treatment of choice. Hepatic resection is indicated in these tumors if there is metastasis to the liver because of its indolent course and added survival advantage.

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