A large functional somatostatinoma in the pancreatic tail: Atypical CT appearances

Pankreas kuyruğunda büyük fonksiyonel bir somatostatinoma: Atipik BT görünümleri

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Somatostatinomas are extremely rare endocrine tumors, and those with diameters above 2 cm are reported to increase the risk of metastasis significantly. We report a case of a large functional somatostatinoma in the pancreatic tail without metastases. A 46-year-old woman with a history of recurrent mild upper abdominal pain and diarrhea for 10 months was admitted to our hospital. Multiple-phase spiral computed tomography revealed a 10 cm x 8 cm, ill-defined, elliptic mass in the body and tail of the pancreas. There was a slightly heterogeneous enhancement on hepatic arterial phase and isodensity to the pancreatic parenchyma with small dotted necrosis within the middle region of the mass on hepatic portal venous and parenchymal phase, with patent splenic vein, dilated collaterals at the splenic hilum and no dilated pancreatic duct, resembling a diffuse infiltration tumor. To the best of our knowledge, this is the first description of multiple-phase spiral computed tomography findings of a functional somatostatinoma in the pancreatic tail and the largest thus far on reported computed tomography, with some differences compared with the previous reports.

Key words: Computed tomography, pancreatic neoplasms, somatostatinoma, liver neoplasms, endocrine tumors

Somatostatinomalar çok nadir endokrin tümörlerdir ve 2cm'den büyük çapa sahip olanlarda metastaz riskinin belirgin olarak arttığı rapor edilmiştir. Burada, pankreas kuyruğunda büyük, fonksiyonel ancak metastaz yapmamış bir somatostatinoma vakasını sunuyoruz. Kırkaltı yaşında bir kadın hasta, 10 aydır tekrarlayan hafif üst karın ağrısı ve ishal şikayetiyle hastanemize başvurdu. Multi – faz spiral Bilgisayarlı Tomografi pankreasın gövde ve kuyruğunda 10 x 8 cm'lik sınırları belirsiz elips şeklinde bir kitle lezyonu gösterdi. Kitlenin, hepatik arteriyel fazda hafif heterojen kontrastlandığı, hepatik venöz ve parenkimal fazda ise ortasında küçük noktasal nekrozlarla, pankreas parenkimiyle isodens olduğu izlendi. Bunun yanında splenik venin açık, dalak hilusunda dilate kollateraller ve pankreatik kanalda dilatasyon olmaması diffüz infiltre bir tümör izlenimi vermekteydi. Bildiğimiz kadarıyla bu, pankreas kuyruğunda yerleşen, şimdiye kadar Bilgisayarlı Tomografi ile rapor edilenlerin en büyüğü olan fonksiyonel somatostatinomanın, ilk multi – faz spiral Bilgisayarlı Tomografi tanımlamasıdır ve geçmiş raporlarla karşılaştırıldığında bazı farklılıklar içermektedir.

Anahtar kelimeler: Bilgisayarlı tomografi, pankreas tümörleri, somatostinoma, karaciğer tümörleri, endokrin tümörleri

INTRODUCTION

Somatostatinomas are extremely rare endocrine tumors that comprise around 1% of all gastroenteropancreatic endocrine neoplasms (1). Most somatostatinomas (56%) originate from the pancreas (2). The majority of patients (92.7%) with this tumor type are symptomatic and may present with the clinical somatostatinoma syndrome (1). Although there have been some reports describing the radiological characteristics of functional somatostatinomas (1, 3, 4), the imaging features with multiple-phase spiral computed tomography (CT) have not been addressed previously. We herein describe the multiple-phase spiral CT findings of a functional somatostatinoma in the body and tail of the pancreas in a 46-year-old woman, which after a literature review, appears to be the largest reported thus far.

CASE REPORT

A 46-year-old woman with a history of intermittent vague upper abdominal pain for 10 months was admitted to our hospital. The patient presented with mild persistent diarrhea, nausea, vomiting and jaundice. The laboratory tests indicated normal blood and urine amylase. The blood, urine, and regular stool tests also revealed no abnorma-

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Manuscript received: 14.02.2007 **Accepted:** 27.10.2008

doi: 10.4318/tjg.2009.0030

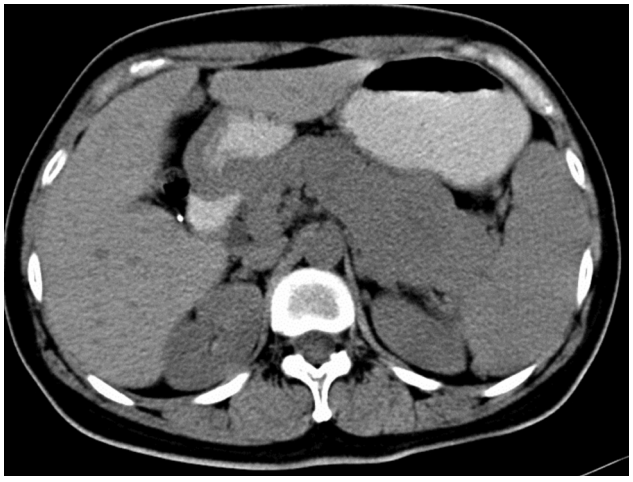

Figure 1. Unenhanced spiral CT shows a large, ill-defined, isodense mass in the body and tail of the pancreas.

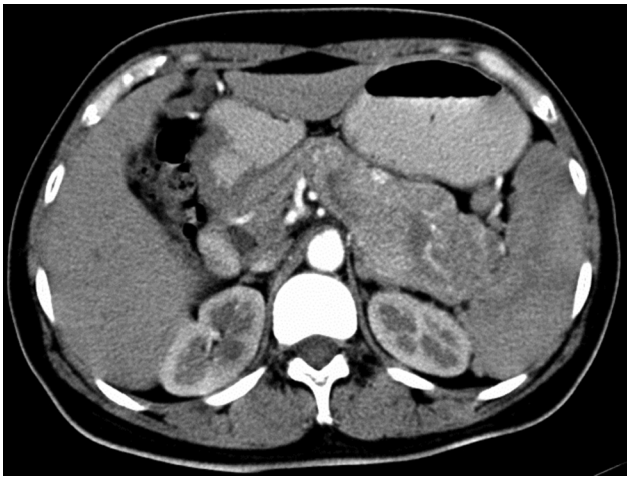

Figure 2. Enhanced CT on hepatic artery phase shows a slightly heterogeneously enhanced tumor in the body and tail of the pancreas.

lity. The tumor markers showed carcinoma embryonic antigen 0.7 ng/ml (normal, <5 ng/ml), alpha-fetoprotein 4.0 ng/ml (normal, <20 ng/ml), and CA 19-9 3.8 U/m (normal, <37 U/ml). Chest X-ray was normal.

An unenhanced spiral CT (Siemens Somatom Sensation 16, Germany) scan revealed a large, ill-defined, elliptic mass measuring 10 cm x 8 cm in the body and tail of the pancreas. The mass showed the same attenuation as the pancreatic parenchyma except for a small region of slightly low attenuation in the middle of the mass (Figure 1). Following intravenous administration of 100 ml of contrast medium at a rate of 3 ml/sec, the tumor revealed a slightly heterogeneous enhancement on hepatic artery phase (Figure 2) and isodensity to the pancreatic parenchyma with small dotted necrosis within the middle region of the mass on hepatic portal venous and parenchymal phase. Associated features included patent splenic vein, dilated collaterals at the splenic hilum, without dilated pancreatic duct (Figures 3A, 3B and 4), resembling a diffuse infiltration tumor. No metastases were detected in the extra-pancreatic region.

Because there was no evidence of metastatic disease, localized pancreatectomy plus splenectomy were undertaken. Pathologic examination revealed a 10 cm x 10 cm x 6 cm somatostatinoma in the body and tail of the pancreas. Microscopic examination showed uniform cells in trabecular and pac-

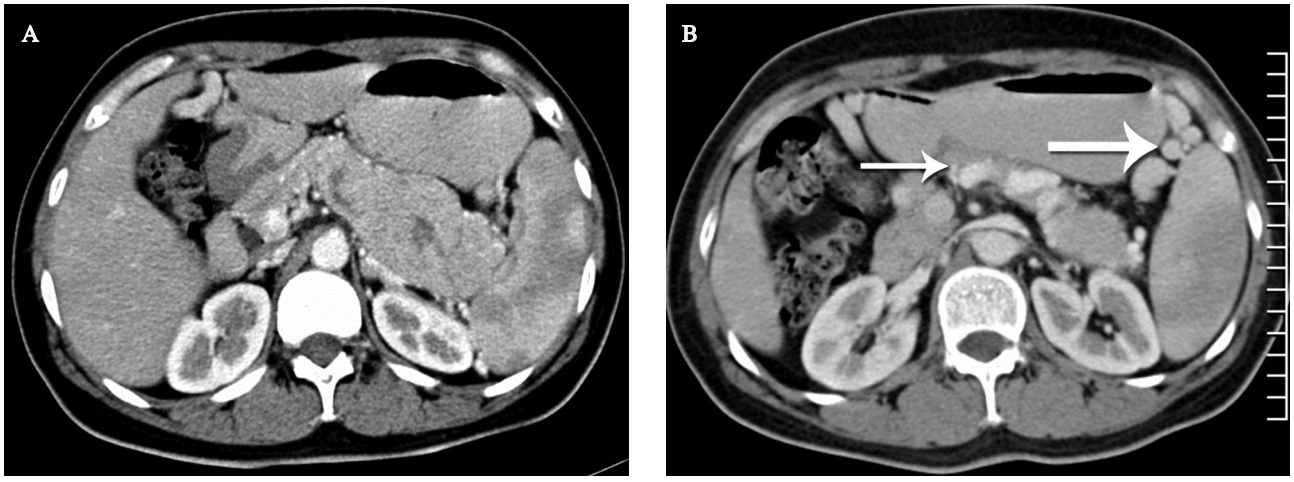

Figure 3. Enhanced CT on hepatic portal venous phase shows a tumor isodense to the pancreatic parenchyma with small dotted necrosis in the tumor **(A)**, associated with patent splenic vein (arrow) and dilated collaterals (arrow) at the splenic hilum **(B)**.

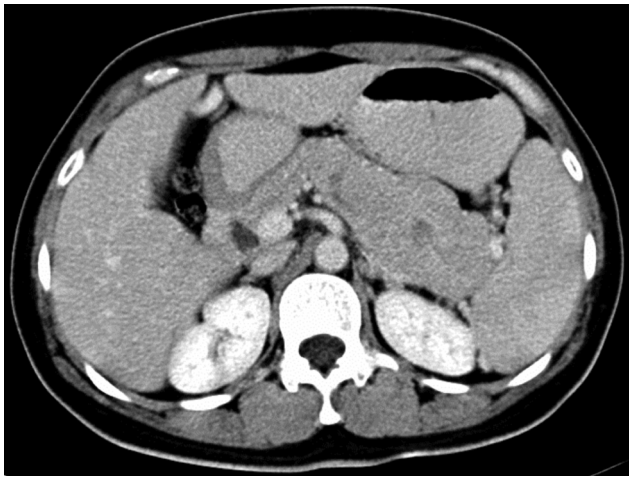

Figure 4. Enhanced CT on hepatic parenchymal phase shows a tumor isodense to the pancreatic parenchyma with small dotted necrosis in the tumor without dilated pancreatic duct.

keting patterns with prominent vascularity, spindle and epithelioid tumor cells with "salt and pepper" nuclei, and rare mitoses (Figure 5). Immunohistologic staining was negative for smooth muscle actin, neurofilaments, insulin, gastrin, and serotonin, and positive for somatostatin (Figure 6).

The patient's postoperative course was uneventful, and she was discharged on the seventh postoperative day. At the 12-month follow-up, the patient was doing well and no longer had the upper abdominal symptoms or mild diarrhea.

DISCUSSION

Somatostatinomas are extremely rare endocrine tumors first described in 1977 (5). They derive from the somatostatin-producing delta cells of the pancreas or the endocrine cells of the digestive tract (6, 7) and may be sporadic (93.1%) or familial (6.9%) in association with neurofibromatosis type 1 (NF1), multiple endocrine neoplasia type 1 (MEN1), and Von Hippel–Lindau syndromes (8-11). The estimated annual incidence is 1 in 40 million (12), with a median age at onset of 54 years. The tumor is malignant in 60–70% of cases (13), with tumor size being the most relevant predictive factor. A diameter of 2 cm is considered as the cut-off value, above which the risk of metastasis significantly increases (14, 15). Our case with a tumor size of 10 cm had no metastases in the extra-pancreatic region, which did not conform to the literature.

Clinically, the classic somatostatinoma or inhibitory syndrome of this tumor type includes diabetes mellitus, diarrhea/steatorrhea, and cholelithiasis. Pathological examination of the surgical specimen provides the definitive diagnosis (1). The immunohistologic staining was positive for somatostatin in our case, and combined with the mild 'inhibitory' syndrome, the functional somatostatinoma was definitely diagnosed.

On the basis of imaging descriptions from previous literature (16, 17), functional pancreatic islet cell tumors are usually small and subtle, with low inherent contrast. They are usually isodense with the pancreas on pre-contrast images and hyperattenuating in the hepatic arterial phase and become inconspicuous in the venous phase. However, only a few CT findings of pancreatic functional somatostatinoma have been reported previously (1, 7, 8). CT of the abdomen shows solid tumors of 3.5-6 cm in diameter, some of which have calcification. The mass size in our case was 10 cm x 8 cm, which is the largest reported thus far. Contrast-enhanced CT shows nonhomogeneous enhancement of the pancreatic tumor with cystic areas of necrosis. To the best of our knowledge, the radiological manifestations of multiple-phase spiral CT of pancreatic functional somatostatinoma have not been described previously. The spiral CT findings of our case were a large, hypervascularized tumor within a small area of necrosis, resembling a diffuse infiltration tumor, and without calcifications or metastases, characteristics distinguishing this case from the previous reports.

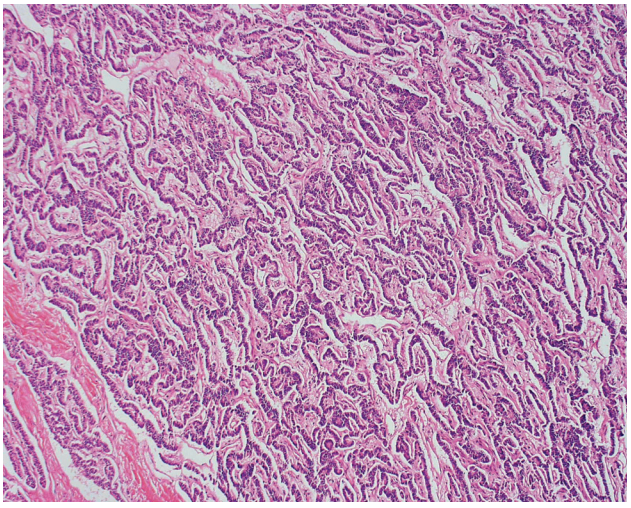

Figure 5. Microscopic examination illustrates uniform cells in trabecular and packeting patterns with prominent vascularity (hematoxylin-eosin [HE] x100).

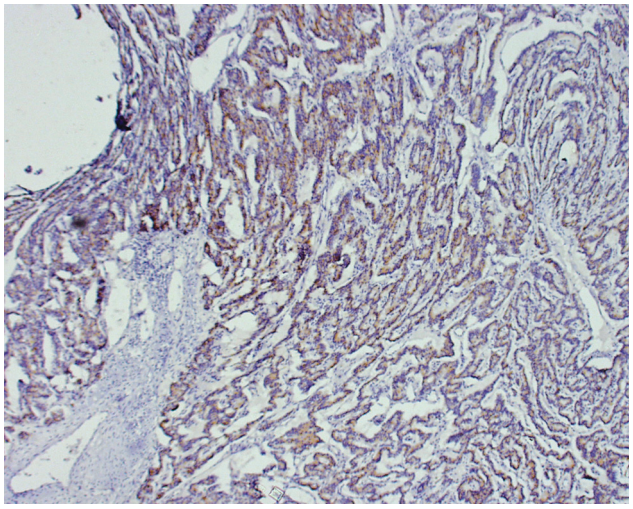

Figure 6. Immunohistologic staining is positive for somatostatin.

There are numerous differential diagnoses that should be considered when a pancreatic mass is detected without any hint of increased hormone release. Depending on the clinical history and the laboratory findings, the following diagnoses should be ruled out: diffuse infiltration tumor, e.g. pancreatic metastases, tuberculosis, and lymphoma; other pancreatic functional endocrine tumors, e.g. glucagonoma and vasoactive intestinal peptide-secreting tumor (VIPoma); non-secreting islet cell tumors; pancreatic adenocarcinoma; and focal pancreatitis. Glucagonomas and VIPomas should especially be excluded, because these two types of pancreatic functional endocrine tumors together with somatostatinomas are usually single, large, and accompanied with metastases at the time of diagnosis (18).

In summary, we have presented herein a 46-year-old female case with a large functional somatostatinoma in the body and tail of the pancreas that resembled a diffuse infiltration tumor without metastases. Although the incidence of functional somatostatinoma is low and the diagnosis can only be confirmed by pathological examination, it should be considered in the list of the possible differential diagnoses of a large, hypervascularized tumor that resembles a diffuse infiltration tumor without distinct necrosis in the pancreas.

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