Neurofibroma of Pancreas Presenting with Low Back Pain

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ABSTRACT

Abdominal neurogenic tumor is most commonly located in the retroperitoneum along the paravertebral sympathetic plexus and in the adrenal glands. Pancreatic neurogenic tumor is very rare. We present a case of pancreatic neurofibroma in a 44-year-old female who suffered from low back pain and epigastric discomfort. Serial imaging findings revealed a cystic tumor in the pancreatic body. Pancreatic neurofibroma was confirmed by the pathological report. The CT appearance was nonspecific and showed a well-defined hypodense mass with mild contrast enhancement and cystic component. Sonographic feature was a hypoechoic mass with both cystic and solid parts. Owing to the difficulty in imaging differential diagnosis and the rarity of such pathology of pancreas, surgical resection of the tumor should be considered for both symptomatic relief and risk of malignancy.

Abdominal neurogenic tumor commonly manifests radiologically as a well-defined, smooth or lobulated mass. The most common abdominal location is in the retroperitoneum along the paravertebral sympathetic plexus and in the adrenal glands. Pancreas is a very rare position for neurogenic tumor. From this case report, we will describe the clinical presentation, ultrasonography and computed tomography (CT) imaging findings, pathologic features and treatment of pancreatic neurofibroma.

CASE REPORT

A 44-year-old female came to our outpatient department with complaints of intermittent low back pain and upper abdominal discomfort of three-week duration. She was quite healthy before without any major systemic disease, except one episode of acute pancreatitis with hospitalization about one year ago. Physical examination revealed mild tenderness over epigastrium without significant rebound tenderness or muscle guarding. The analyses of blood chemistry and complete blood cell count were all within normal limits. Tumor markers including CA-199 and carcinoembryonic antigen (CEA) were also not elevated. Esophagogastroduodenoscopy showed negative finding, but a hypoechoic cystic lesion with solid component was observed at the body of pancreas on the ultrasonography (Fig. 1). Subsequent CT scan demonstrated a big cystic mass measuring 5.7 × 8 × 5.8 cm in the pancreatic body with partial contrast enhancement of the solid component (Fig. 2).

From the imaging findings, pancreatic tumor such as cystadenoma, cystadenocarcinoma, intraductal papillary mucinous tumor, solid pseudopapillary tumor or pseudocyst were considered. She was then admitted to our general surgery ward for surgical intervention. After admission, subtotal distal pancreatectomy of pancreatic body and tail and splenectomy were performed smoothly.
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Figure 1. Abdominal ultrasound revealed a cystic lesion with solid component (arrow) in the body of pancreas.

Figure 2. Unenhanced CT scan a. showed a round hypoattenuating mass lesion in the pancreatic body. Contrast-enhanced CT scan b. of this pancreatic cystic mass showed mild contrast enhancement of its solid component (white arrows) with displacement of adjacent visceri. No calcification was detected in the tumor. Incidentally, an adrenal adenoma was seen in the left adrenal gland (black arrow).
Gross pathology revealed a well-delineated yellowish tan soft tissue mass over pancreatic body, measuring 7.5 × 6 × 4.9 cm in size. Neurofibroma with focal xanthomatous change was confirmed under microscopy (Fig. 3). No cellular atypia was noted. Nerve fibers were demonstrated by Bodian stain. The tumor cells were immunoreactive to S100 protein, but non-reactive to the other immunostains. The spleen showed no remarkable change except congestion. There was no evidence of malignancy in the specimen examined. The patient was discharged uneventfully 8 days after the operation.

**DISCUSSION**

Tumors originating from peripheral nerve sheath include neurilemmoma (schwannoma), neurofibroma and malignant peripheral nerve sheath tumor. Among them, neurofibromas usually appear as well-circumscribed tumors of the nerve system, and consist of fibroblasts, Schwann cells, nerve fibers, collagen in a myxoid/mucinous matrix [1, 2]. About 10% of neurofibromas are in association with neurofibromatosis type I (NF1), whereas the remainder are of the solitary type [3].

Neurofibromas can occur in the skin, soft tissues, viscera, almost anywhere in the body [1]. However, pancreatic involvement of neurofibromas is extremely rare [1, 2, 4]. Pancreatic involvement by the neurofibromas could be due to extension of plexiform variant along the celiac plexus in NF1 patients, or the tumors may arise de novo from the pancreas [2, 5]. To our knowledge, only four cases of solitary neurofibromas in the pancreas have been reported in the literature [1, 4, 6, 7]. Among them, one case was plexiform type, but the patient lacked other features of NF1 [4]; the others were classical neurofibromas. The reported cases so far seemed to occur mainly in middle-aged people, without significant gender differences. The presenting symptoms were usually upper abdominal pain or back pain, and the tumors were mostly found in the head and neck of the pancreas. Nevertheless, our case presented with a mass lesion in the pancreatic body.

The characteristic CT appearance of neurofibromas is homogenously hypodense, smooth and round well-defined masses with the attenuation values of 20-25 HU on unenhanced scan, and shows mild contrast enhancement [1-3, 5]. The low attenuation of neurofibromas is attributed to lipid-rich Schwann cells, adipocytes, and entrapment of perineural adipose tissue [2, 3, 5]. Some neurofibromas contain cystic spaces inside them due to myxoid matrix or cystic degeneration [2-4], as in our case. The sonographic finding of neurofibroma is typically a well-defined hypoechoic mass with occasional heterogeneity related to myxoid stroma, hemorrhage, fibrosis or calcification. Cystic areas and posterior acoustic enhancement are also commonly seen [8]. Hypervascularity could be noted in neurofibromas on color Doppler ultrasonography [9].

In our case, the tumor comprised of mixed solid and cystic components. The cystic lesions of the pancreas
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contain a spectrum of benign, malignant, borderline neoplasms and tumor-like lesions. The preoperative differential diagnosis included mucinous cystic neoplasm, solid pseudopapillary tumor, cystic neuroendocrine neoplasm and intraductal papillary mucinous tumor according to the features and prevalence described by previous studies conducted for cystic lesions of the pancreas [10-13]. Pseudocyst was also considered in the differential diagnosis due to the patient’s history of pancreatitis, but the large solid component made it less likely to be a pseudocyst [10]. The diagnosis of neurofibroma didn’t enter our list of differential diagnosis because of its rarity. It is, however, still difficult to accurately diagnose a cystic lesion of the pancreas and to determine whether it is benign or malignant with current cross-sectional imaging [10-13].

The management of cystic neoplasms of pancreas has not reached a consensus. The characteristics associated with malignancy include the presence of solid component, increasing cyst size and symptoms. Only small asymptomatic cysts (<3cm) without suspicious features are suggested to be safely followed up with imaging studies. Fit patients with large or symptomatic cysts, and low surgical risk are indicated for surgical resection of the cystic lesion [10, 13, 14]. Surgical removal of the cystic tumor is the treatment of choice for both symptomatic relief and risk of malignancy. Prognosis after resection of the neurofibroma remains unknown due to scarcity of cases. No definite tumor recurrence was noted after surgery in the previous reported cases [6].

CONCLUSION

We report a rare case of pancreatic neurofibroma. The presenting symptoms of pancreatic neurofibroma are usually upper abdominal pain or back pain. CT appearance is a well-defined hypodense mass with mild contrast enhancement, which sometimes contains cystic areas. Sonographic feature is as hypoechoic mass with occasional heterogeneity or cystic parts. It is difficult to make a diagnosis of pancreatic neurofibroma by imaging due to its rarity. Surgical resection is considered as the treatment of choice for both symptomatic relief and risk of malignancy. Prognosis after surgery is undetermined.

REFERENCES