Case Report

Solitary neurofibroma of the pancreas body not associated with type 1 neurofibromatosis

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Abstract

Neurofibromas arise from peripheral nerve cells. They are rarely found within the pancreas, especially not associated with type I neurofibromatosis. Here, we report a case of a neurofibroma in a 44-year-old woman who initially presented with epigastralgia. Imaging revealed one large cystic mass of $5.7 \times 8 \times 5.8$ cm in the pancreatic body, which was resected with distal pancreatectomy. The postoperative course of treatment was without complication, and no signs of recurrence were observed after 1 year and 6 months’ follow-up.

Keywords: distal pancreatectomy; neurofibroma

1. Introduction

There are two types of neurofibromatosis, including neurofibromatosis type 1 (NF-1) and type 2 (NF-2). The two types have different genetic conditions. NF-1 is a tumor disorder that is caused by the malfunction of a gene on chromosome 17 that is responsible for control of cell division. NF-1 causes noncancerous lumps. NF-1 often comes with scoliosis (curvature of the spine), learning difficulties, eye problems, and epilepsy. NF-1 affects about 1 in 3000 to 5000 people and has a wide range of severity. It is difficult to predict who will remain only mildly affected or who will be more severely affected with neurofibromatosis type 1. People with NF-1 tend to develop varying numbers of neurofibromas. Neurofibroma is a benign nerve sheath tumor and little cancerous in the peripheral nervous system. It causes lumps on the skin which can occur anywhere on the body. While these skin changes do not lead serious medical complications, they can affect a person’s appearance. Neurofibromas can also develop under the skin or deeper in the body. These can grow quite large, result in significant medical problems and affect the structure of nearby bone, skin and muscles.1–5

2. Case report

A 44-year-old woman presented with intermittent dull epigastric pain for 2 weeks without experiencing nausea, vomiting, weight loss, yellowish skin discoloration, or changes in bowel habit prior to admission. One episode of acute pancreatitis was noted 1 year before this incident. After conservative treatment, no chronic sequelae had been observed. The patient had no other history of major systemic disease and did not smoke or consume alcohol.

On physical examination, mild bulging of the epigastric area with mild tenderness was noted. The laboratory data, such as amylase, lipase, total bilirubin, and tumor markers, such as
carbohydrate antigen 19-9 and carcinoembryonic antigen, were unremarkable.

Abdominal X-ray revealed lateral displacement of the stomach, and ultrasound showed a cystic lesion approximately 7.4 × 5.6 cm in size, with a solid component (approximately 5 cm) within the pancreas. Upper gastrointestinal endoscopy was normal. Abdominal computed tomography (CT)-scan revealed one cystic mass, measuring 5.7 × 8 × 5.8 cm, within the pancreatic body (Fig. 1). The differential diagnosis included pancreatic mucinous cystic adenoma, carcinoma, or solid pseudopapillary tumor.

Because the mass lesion was large, surgical exploration was indicated, and the patient consented. During surgical exploration, one cystic tumor was identified over the pancreatic body; it had a soft consistency and was well-circumscribed, without infiltration into or adhesion to the surrounding tissues. No evidence of enlarged lymph nodes or carcinomatosis was found. The patient underwent distal pancreatectomy and splenectomy without complications, and the postoperative course was uneventful. No signs of tumor recurrence were observed after 1 year and 6 months of follow-up.

Fig. 1. Computed tomography of the abdomen. The (A) transverse and (B) sagittal views revealed one large cystic mass (T), measuring 5.7 × 8 × 5.8 cm over the pancreatic body without contrast enhancement. No associated lymphadenopathy was observed.

Fig. 2. Analysis of the resected specimen. Distal pancreas (P) with spleen (S) and one well-delineated yellowish-tan soft tissue mass (T) measuring 7.5 × 6 × 4.9 cm in size was observed (A) anteriorly and (B) posteriorly. (C) Cross-section of the tumor revealed a xanthomatous capsule (X).
Macroscopically, a well-delineated, yellowish tan soft tissue was observed over the pancreatic body (Fig. 2). Microscopic examinations of the tumor revealed typical characteristics of neurofibroma with focal xanthomatous features without cellular atypia (Fig. 3). Nerve fibers were detected using Bodian stain (Fig. 4), and the tumor cells were immunoreactive to S-100 (Fig. 5). There was no evidence of malignancy, and the lesion was compatible with neurofibroma.

3. Discussion

Although solitary neurofibroma is rare in cases without NF-1, some studies have reported sporadic cases in the skin, appendix, stomach, and jejunum.6,7 Solitary sporadic neurofibroma of the pancreas is uncommon.8 PubMed and Medline searches up to 2008 detected a limited number of related studies, all of which were case reports (Table 1).6,7,9
Neurofibroma of the pancreas was more commonly observed in middle age and was associated with upper quadrantal abdominal pain; one case reported symptoms similar to acute pancreatitis with irregular obstruction of the main pancreatic duct and a narrowed intrapancreatic common bile duct upon ERCP (endoscopic retrograde cholangiopancreatography) examination. However, in our patient, no sign of pancreatic duct obstruction was observed.

Another difference from the previously reported cases was that our patient was female. Despite the large tumor size, no significant nutritional wasting or weight loss was detected. In all cases, pancreatic carcinoma could not be excluded until final pathological examination was undertaken.

Neurofibroma is a neurogenic tumor, which arises from the nerve sheath. It is a well-demarcated, intraneural or diffusely infiltrative extraneural tumor consisting of a mixture of cell types and elements of the peripheral nerve, including axons, fibroblasts, and Schwann cells. Typically, neurofibromas grow slowly and have minimal potential for malignant transformation unless they are associated with NF-1 (formerly known as von Recklinghausen’s disease).

Macroscopic appearance of neurofibromas was well-delineated with or without the capsule; they are not sharply separated from the surrounding tissue. The cut surface is primarily gray-white, often with whorls and spirals. In the event of high lipid content, the tumor appears more yellowish, with a xanthomatous capsule, as in our case.

Highly collagenized stroma with little myxoid material and spindle cells are found upon microscopic analysis of neurofibromas. In addition, interlacing bundles of elongated cells with wavy nuclei were observed in our case. Also, mucinous, edematous, or xanthomatous changes in the stroma may occur, and mitotic figures are rare. Furthermore, when elevated mitoses or overly expressed cell proliferation markers are observed, the potential for malignant transformation should be suspected.

Some studies have tried to differentiate benign from malignant pancreatic tumors using CT imaging and found that neurofibromas of the pancreas tended to have marked and homogeneous hypodensity, 20-25 HU at baseline, with mild contrast enhancement. Ladouce et al reported three cases of solitary neurofibroma and concluded that it was difficult to have a diagnosis without histopathological analysis.

When patients are diagnosed with neurofibroma, physicians must be aware of possible associations with NF-1. With the benign nature of neurofibroma and the minimal risk of recurrence, surgical excision seems to be curative. However, the small number of cases hinders the discussion of proper treatment modality.

References