Primary Paraganglioma of the Head of Pancreas: Contribution of Combinatorial Image Analyses to the Diagnosis of Disease

Key words: computed tomography, pancreas, paraganglioma

Introduction

The prevalence of pancreatic tumor including carcinoma is increasing, however, the precise diagnosis of pancreatic tumor remains difficult. Since excessive surgical resection of the pancreas often causes deficient secretion of pancreatic hormones and digestive enzymes such as insulin, amylase and lipase, accurate diagnosis is needed for proper surgical treatment of pancreatic tumor. Paraganglioma of the pancreas is a rare disease, and its ultrasonic, computed tomographic appearances have been evaluated in several cases (1–5). We herein report a patient with paraganglioma in the head of pancreas, who was diagnosed as the neuroendocrine tumor of pancreas before surgery. Combinatorial image analyses may be useful for the diagnosis of pancreatic paraganglioma.

Case Report

A 72-year-old woman showed abdominal discomfort of 2 months duration and was admitted to our hospital. She had no history of hypertension, diabetic mellitus or other diseases. There was no family history of pancreatic disease including pancreatic tumor. Her general physical examination was not contributory and there was no palpable abdominal mass. Laboratory data including tumor marker, hormones and catecholamine were within normal limits.

An ultrasonography demonstrated a mass of around 4 cm in diameter in the head of pancreas with a cystic component. An enhanced computed tomography (CT) showed an enhancement of parenchyma and a non-enhanced cyst in the tumor (Fig. 1A). A rich tumor stain was confirmed in the tumor in angiography. On endoscopic retrograde cholangiopancreatography (ERCP), the tumor displaced the main pancreatic duct inferiorly, but the pancreatic duct had neither obstruction nor irregularity. Based on these findings, we suspected a non-functional neuroendocrine tumor such as paraganglioma.

Surgical resection of the pancreas head was performed. Histology showed a classical zellballen pattern in the tumor (Fig. 1B). Immunohistochemistry revealed positive staining of neuron-specific enolase, S-100 and chromogranin A in the tumor cells. After surgery, she had no dysfunction in secretion of pancreatic hormones and enzymes.

Discussion

Paraganglioma arises from paraganglia and is essentially derived from neural cells (1). Primary pancreatic paraganglioma is a rare disease and difficult to be exactly distinguished from another tumor (1–5). Recently, the features of pancreatic paraganglioma has been reported. Paraganglioma

Figure 1. A) The appearance of enhanced computed tomography in the transverse section through the upper abdomen. CT shows a mass enhanced strongly with an area of low density in the pancreatic head. B) The microscopic view of paraganglioma of the pancreas head shows the zellballen pattern (HE stain, ×100).
of the pancreas, as well as other benign tumors, usually does not present any symptoms and is often found incidentally, in particular, there is no patient with the symptom of jaundice in contrast to carcinoma of the pancreas head (5). On image analyses, all patients with pancreatic paraganglioma had a cystic component and a parenchyma developing a rich blood supply from the surrounding vessels in contrast to carcinoma (4, 5). Ultrasonography and CT scan are useful for the detection of pancreatic tumor including paraganglioma. These examinations indicate that paragangliomas are characterized by a highly vascular and well-enhanced tumor with a cystic area (5). In the present case, the findings of the tumor were consistent with those of previous cases on image analyses. In addition, we confirmed the rich vascularity in the tumor by analysis of angiography. On the other hand, there is no finding of laboratory examination including hormone in patients with pancreatic paraganglioma as well as other retroperitoneal paragangliomas (5). Our patient also did not present any abnormal data on laboratory examination. In surgical treatment, the prognosis of simple excision of the tumor was equally well compared to radical surgery such as pancreatoduodenectomy. In our patient, combinatorial image analysis helped us to diagnose pancreatic paraganglioma and we could select the non-excessive surgical treatment without post-surgical functional disorder of pancreas.

In conclusion, we report a patient with paraganglioma in the head of pancreas considered by combinatorial image analysis. Although further study is needed, adequate combinatorial image analysis may contribute to the diagnosis of pancreatic paraganglioma.

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