Endoscopic ultrasound findings in duodenal gangliocytic paraganglioma

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Received for publication March 11, 2004. Accepted June 30, 2004

Abstract

Gangliocytic paraganglioma (GP) is an uncommon benign neurogenic tumour of the digestive tract that is usually located in the descending duodenum. Due to the unusual location, such lesions are frequently overlooked on routine radiological examinations and initial endoscopy. With the recent development of endoscopic ultrasound, lesions in this location can be more easily detected and better viewed. We report an additional case of duodenal gangliocytic paraganglioma detected by endoscopic ultrasound, with the aim to further determine the endoscopic ultrasonographic imaging features of duodenal gangliocytic paraganglioma.

CASE PRESENTATION

A 39-year-old man presented with a four-month history of abdominal pain. Initial endoscopic examination at outside clinics revealed superficial gastritis. Owing to the persistent complaints, duodenoscopy was performed and a duodenal submucosal tumour with a smooth surface and intact mucosa over the second portion of the duodenum was found. A fine needle aspiration revealed chronic duodenitis. After being transferred to the Chang Gung Memorial Hospital in Taiwan, endoscopic ultrasonography (Olympus, Japan; 12 MHz to 20 MHz) was arranged and revealed a hypoechoic, heterogenous 1.75 cm tumour with smooth margins (Figure 1), originating from the submucosal layer of the duodenum. There was no neighbouring lymphoadenopathy. The lesion was excised locally and postoperative recovery was uneventful. The patient remained well at a follow-up of 36 months after surgery. Endoscopic follow-up showed no obvious evidence of recurrence. Microscopically, the tumour showed the typical, distinctive morphology of GP: a neuroendocrine-like component, ganglion cells and spindle-shaped Schwannian cells (Figure 2). The endocrine cell components had a carcinoid-like appearance and exhibited immunoreactivity for neuron-specific enolase, synaptophysin and chromogranin. The isolated ganglion cells were immunoreactive for neuron-specific enolase and the spindle Schwann cells were immunopositive for S-100 protein. The distinct morphological features, in conjunction with the immunohistochemical stains, were diagnostic for this entity and differentiated it from other tumours such as schwannoma, gastrointestinal stromal tumours and malignant lymphoma, which are more commonly seen in this location.

DISCUSSION

GP is a very rare tumour of the gastrointestinal tract, most often arising from the second portion of the duodenum (1). Although several theories attempt to explain this rare histological combination of neuroendocrine components, the exact
The pathogenesis of GP remains somewhat elusive. Immunocytochemical studies have led to more convincing ideas. It has been suggested that the tumour arises from an embryonic celiac ganglion; from pluripotent stem cells located at the base of intestinal glands; or that the lesion is due to hamartomatous proliferation of endodermally derived epithelial cells originating from the ventral primordium of the pancreas, and neuroectodermally derived ganglion and Schwannian spindle cells. Burke and Helwig proposed that GP is a hamartoma or choristoma arising from ectopic pancreas tissue, because pancreatic-like tissue has been identified, and immunoreactive human pancreatic polypeptide or somatostatin is occasionally detected in GP.

Clinically, GP is approximately twice as common in male patients as in female patients and, when present, is typically found in middle-aged (50 to 60 years of age) patients. Gastrointestinal tract hemorrhage and abdominal pain are among the most common manifestations of this lesion. Although there have been rare case reports of malignant paragangliomas with lymph node metastases, paragangliomas are clinically a benign lesion. It is usually sufficient to locally excise the tumour with excellent results. Scheithauer et al reported a series of 11 cases with duodenal GP. Three of the 11 in their series underwent endoscopic removal smoothly, and the remaining seven cases underwent surgical resection. All the patients were well on long-term follow-up. Radical surgery like Whipple’s procedure is usually not recommended. GP involving the ampulla still can be smoothly and locally excised; Sando et al successfully conducted the resection of an ampullary GP via an anterior duodenotomy employing Kocherization of the duodenum, mobilization of the hepatic colonic flexure, and cannulation of the common bile and pancreatic ducts. Complete local excision with sphincteroplasty was successfully performed. In extreme cases, such as GP of the papilla of Vater with regional lymph node metastasis, the pancreato-duodenectomy with lymph node dissection for en bloc resection of tumour is justified. Immunohistochemistry of duodenal GP typically shows positive epithelial cell staining for somatostatin, pancreatic polypeptide and other neuroendocrine markers. Uncommonly, Evana et al reported a case of duodenal GP presenting as an ampullary tumour with obstructive jaundice and the histologically confirmed typical features of a duodenal GP, but immunohistochemistry demonstrated negative staining for somatostatin and other polypeptides, which contradicts most other reported cases.

A number of other neoplasms that arise in the duodenal submucosa should be considered in the differential diagnosis of GP. Endoscopic ultrasound may supply abundant information for the initial differential diagnosis. Leiomyomas or leiomyosarcomas usually appear to arise from the muscular propria, and larger leiomyosarcomas are commonly associated with areas of cystic internal hemorrhage or necrosis. Hemangiomas and hamartomas tend to show a heterogeneous pattern of soft tissue. Choledochal and duplication cysts are mainly cystic in nature. Lipomas may show similar echogenicity of surrounding fat. Isolated lymphomas of the duodenum are quite unusual and the imaging features of this entity probably are more protean. The endoscopic ultrasonography of this case revealed a 1.75 cm tumour originating from the submucosal layer of the duodenum (Figure 1).

**CONCLUSION**

A case of duodenal GP was detected by endoscopic ultrasound and was successfully surgically treated with excellent results. Although endoscopic ultrasound is instrumental in making the initial differential diagnosis, histological examination is
mandatory to confirm the final diagnosis. With awareness of this disease entity and advanced endoscopic ultrasound examination, an accurate diagnosis might be achieved earlier and the treatment then can be well planned.

REFERENCES