CASE REPORT

Ganglioneuroma of the Duodenum
Report of a Case and Literature Review

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Neurogenic tumors of the gastrointestinal tract are very rare, especially duodenal paragangliomas. Since 1957, when Dahl et al (1) reported the first case, up to the present, a total 74 cases (2) of paraganglioma of the duodenum have been reported. The term paraganglioma is used in the broader sense and, according to the histological composition, it may be called a ganglioneuroma, nonchromaffin paraganglioma, gangliocytic paraganglioma, paraganglioma, and paraganglioneuroma (3). Here we report a case of ganglioneuroma of the duodenum, only the fourth case since the first reported by Dahl et al (1).

CASE REPORT

Mr. Lee, a 43-year-old male, was admitted to the hospital because of upper gastrointestinal bleeding. He has been experiencing dull abdominal pain over the epigastric region for about one month; the pain usually occurred before meals and was relieved postprandially or after medication with antacids. One day before admission, the patient began to pass tarry stool associated with dizziness and general weakness. Except for his alcohol history, there was no history of taking ulcerogenic drugs, no previous episode of gastrointestinal bleeding, no blood dyscrasia, and no history of peptic ulcer. On admission, pulse rate was 84/min and regular, blood pressure 120/84 mm Hg, physical examination was essentially normal except for an anemic appearance. Laboratory data were: Hgb 8.8 g/dl, Hct 27.5%, WBC 6.1 x 10³/mm³, platelet 232 x 10³/mm³, stool occult blood +++, electrolytes and biochemistry were all within normal range.

Upper gastrointestinal endoscopic examination disclosed a polypoid mass about 3.0 cm in diameter with ulceration at the plateau located in the second portion of the duodenum (Figure 1) near the ampulla of Vater. Biopsy through the endoscope showed hyperplastic glands and hemorrhage of the duodenal mucosa. A barium meal of the hypotonic duodenogram was compatible with the endoscopic findings. The bleeding ceased spontaneously, but because of the obscure nature of this lesion, we decided to explore and resect the lesion. Surgery revealed a solitary submucosal tumor mass, measuring 3.0 x 2.5 cm, with ulceration, located just proximal to the ampulla of Vater. Pathologic examination showed ganglioneuroma of the duodenum with mature ganglion cells and neurofibers (Figure 2) noted beneath the duodenal mucosa (Figure 3).

DISCUSSION

Ganglioneuroma is a well-differentiated benign tumor composed of mature nerve cells and varying numbers of nerve fibers (1, 4, 5). The adrenal medulla is the most common origin, but it may occur along any portion of the sympathetic nerve chains. It very rarely occurs in the gastrointestinal tract and is almost exclusively located in the second part of the duodenum with a predilection for the region of the ampulla of Vater. Dahl et al (1) reported the first case of ganglioneuroma of the duodenum. Taylor and Helwig in 1962 (6) reported nine cases of polypoid tumor in the duodenum. The tumor was composed of spindle cells, epithelioid cells, and some ganglionlike cells. The spindle cell is a Schwann component. The epithelioid cell was argyrophilic without argentachromaffinity, and the nests of epithelioid cells are surrounded by a network of capillaries that were indistinguishable from Zellballen of the carotid body. These tumors were named nonchromaffin paragangliomas. Kepes et al in 1970 (7) reported two cases of tumors composed of epithelioid cells, spindle cells, and well-developed ganglion cells, which he named gangliocytic paragan-
DUODENAL GANGLIONEUROMA

To the present, a total of 74 cases of paraganglioma of the duodenum have been reported (2) and the variable histologic composition has led to varying terminology: ganglioneuroma (1, 4, 5), nonchromaffin paraganglioma (6, 8, 9) gangliocytic paraganglioma (7, 10–13) paraganglioma (14–16) and paraganglioneuroma (17). This is the fourth reported case of ganglioneuroma; the previous three cases were reported by Dahl et al (1) Gemer et al (4) and Goldman et al (5).

Most of the patients with duodenal paraganglioma were in the sixth decade of life (range 15–80 with mean age of 51.4) and males predominated 1.8:1 (2). The most common complaint was gastrointestinal bleeding. The tumor is polypoid in appearance with a size range of 0.5–10 cm in diameter, with erosion or ulceration in the top portion of the mass that is the source of the bleeding. Most of the tumors were found in the second portion of the duodenum with a predilection for the ampulla of Vater.

Fig 1. Ulcerative tumor mass located in the second portion of the duodenum.

Fig 2. Mature ganglion cells and neurofibers with positive NS100 (400×).