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**Primary lymph node gastrinoma in jejunal mesentery**

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Primary gastrinomas have been reported in lymph nodes within the gastrinoma triangle. We report a 56-year-old woman with possible primary lymph node gastrinoma in the jejunal mesentery. Six months after excision of the tumor, she is asymptomatic and serum gastrin level is normal. [Indian J Gastroenterol 2005;24:266-267]

Sporadic gastrinomas are found predominantly to the right of the superior mesenteric artery (SMA) in an area called the gastrinoma triangle. They are frequently extra-pancreatic, multiple, and have a high incidence of lymph node involvement within the triangle. These tumors can be identified and excised for cure in a high percentage of cases.1 Gastrinomas found to the left of the SMA are often within the pancreatic substance, large, infiltrative, and frequently metastatic to the liver at the time of initial diagnosis.2,3

A 56-year-old woman presented with 6-year history of upper abdominal discomfort and loose stools. She had undergone appendicectomy 4 years earlier. Physical examination was unremarkable. She was evaluated for acid-peptic disease and small-bowel diarrhea. At gastroscopy inflammatory changes in the distal esophagus and multiple small ulcers in the second part of the duodenum were seen. Basal serum gastrin level was elevated (960 pg/mL; normal 0-90). She also had elevated parathormone (361 pg/mL; normal 8-74), mildly elevated prolactin (39.3 ng/mL; normal 5-25) and low serum calcium (8.8 mg/dL; normal >10.4). Plain skull radiograph was unremarkable. CT showed a 2-cm well-enhancing nodule in the jejunal mesentery. 111-indium octreotide scan showed increased uptake in the jejunal mesentery (Fig) and left parathyroid region. Ultrasoundography of the thyroid and parathyroid was unremarkable. In view of low serum calcium and negative family history, MEN 1 syndrome was considered unlikely and elevated parathormone was interpreted as a physiological compensatory response.

She was started on calcium supplements. Excision of the gastrinoma was done under general anesthesia. The nodule was located 20 cm from the duodeno-jejunal flexure in the jejunal mesentery free from the bowel wall and mesenteric vessels. Intraoperative ultrasonography of the duodenum and pancreas was normal. Histology revealed a lymph node with its architecture effaced by a tumor. Tumor cells were positive for synaptophysin and chromogranin.

Postoperative basal and secretin-stimulated gastrin levels were normal. The patient was symptom-free 6 months later, and basal gastrin levels and serum parathormone were normal.

The gastrinoma triangle is defined as the confluence of the cystic and common bile duct superiorly, second and third portions of the duodenum inferiorly, and the neck and body of the pancreas medially, both dorsally and ventrally. The area of the triangle corresponds to an area of pancreatic development, where the ventral pancreatic bud rotates 180° around the primitive foregut to fuse with the dorsal pancreatic bud.

The existence of primary lymph node gastrinomas is widely debated. There is no embryological basis for the presence of neuroectodermal cells within lymphatic tissue. The current opinion is that the putative stem cell of their genesis is of ventral pancreatic bud origin and that these cells migrate during the posterior rotation of the ventral pancreatic bud to become incorporated in embryonic lymph tissue, later coalescing to form lymph nodes.4 One study reported that chromogranin and synaptophysin-positive cells can be found in abdominal lymph nodes of patients without gastrinomas.5 About 20% of the lymph nodes in the gastrinoma triangle showed synaptophysin-positive cells and 15% had chromogranin in an autopsies series without Zollinger-
Ellison syndrome (ZES). This histologic study supports the theory that entrapment of neuroendocrine cells during development could lead to development of primary lymph node gastrinomas.

In our patient it is possible that a microscopic primary tumor in an unknown location might have been overlooked; somatostatin receptor scintigraphy, the most sensitive imaging modality available to localize gastrinomas or their metastases, frequently (~50%) misses primary gastrinomas less than 1 cm in diameter and therefore might only localize the accompanying positive lymph nodes. We are of the opinion that she had a primary lymph node gastrinoma as six years is too protracted a period for a primary to remain microscopic. Data suggest that patients with tumors to the left of the SMA have shorter duration of symptoms and are often large at presentation. Long duration of symptoms is also in variance with the observed biologic behavior of non ventral pancreatic bud tumors.

The puzzle of the origin of sporadic gastrinomas still seems to be undeciphered. The identification of a possible primary lymph node gastrinoma outside the gastrinoma triangle is at conflict with current theories.

References

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