Primary lymph node gastrinoma or occult duodenal microgastrinoma with lymph node metastases in a MEN1 patient: the need for a systematic search for the primary tumor.

Abstract:
Gastrinoma tissue has been found frequently in lymph nodes located near the duodenum without a known primary tumor. Therefore, it has been suggested that a primary lymph node gastrinoma exists. We report on a 38-year-old woman suffering from multiple endocrine neoplasia type 1 (MEN1) confirmed by menin gene mutation analysis. MEN1 disease started with primary hyperparathyroidism followed by Cushing disease, the detection of tumors of the pituitary, adrenal cortex, and the pancreas and also an elevated serum gastrin level. An octreotide scan revealed 4 tumors in the upper abdomen. A selective arterial calcium stimulation test located the source of the hypergastrinemia to the area of the gastroduodenal and the superior mesenteric arteries. Total pancreatoduodenectomy was performed and conventional histopathologic examination revealed a well-differentiated cystic neuroendocrine tumor of the pancreas expressing glucagon and accompanied by several microadenomas. In addition, 3 suprapancreatic lymph nodes with gastrin-positive endocrine tissue were found. None of the pancreatic microadenomas expressed gastrin and no duodenal endocrine tumor was found despite careful macroscopic
examination. Only after complete embedding of the duodenal and pancreatic tissue in 65 paraffin blocks, 2 microgastrinomas (0.45 and 0.8 mm in diameter) were identified in the duodenum. It is concluded that duodenal gastrinomas that give rise to lymph node metastases may be so tiny that they are easily overlooked in a routine examination and that systematic tissue monitoring is required to identify them.