Somatostatin-producing neuroendocrine tumors of the duodenum and pancreas: incidence, types, biological behavior, association with inherited syndromes, and functional activity.

Abstract:
Somatostatin-producing neuroendocrine tumors (SOM-NETs) of the duodenum and pancreas appear to be heterogeneous. To determine their clinicopathological profiles, respective data were analyzed on a series of 82 duodenal and 541 pancreatic NETs. In addition, the clinical records of 821 patients with duodenal or pancreatic NETs were reviewed for evidence of a somatostatinoma syndrome. Predominant or exclusive expression of somatostatin was found in 21 (26%) duodenal and 21 (4%) pancreatic NETs. They were classified as sporadic (n=31) or neurofibromatosis type 1 (NF1)-associated duodenal NETs (n=3), gangliocytic paragangliomas (GCPGs; n=6), or poorly differentiated neuroendocrine carcinomas (pdNECs; n=2). In addition, five duodenal and four pancreatic SOM-NETs were found in five patients with multiple endocrine neoplasia type 1 (MEN1). Metastases occurred in 13 (43%) patients with sporadic or NF1-associated SOM-NETs, but in none of the duodenal or pancreatic MEN1-associated SOM-NETs or GCPGs. Sporadic advanced (stage IV) SOM-NETs were more commonly
detected in the pancreas than in the duodenum. None of the patients (including the 821 patients for whom only the clinical records were reviewed) fulfilled the criteria of a somatostatinoma syndrome. Our data show that somatostatin expression is not only seen in sporadic NETs but may also occur in GCPGs, pdNECs, and hereditary NETs. Surgical treatment is effective in most duodenal and many pancreatic SOM-NETs. MEN1-associated SOM-NETs and GCPGs follow a benign course, while somatostatin-producing pdNECs are aggressive neoplasms. The occurrence of the so-called somatostatinoma syndrome appears to be extremely uncommon.