Syndromic versus non-syndromic sporadic gastrin-producing neuroendocrine tumors of the duodenum: comparison of pathological features and biological behavior.

Abstract:
Sporadic gastrin-producing neuroendocrine tumors of the duodenum present either with the Zollinger-Ellison syndrome (ZES) or with unspecific symptoms. While syndromic gastrin-producing neuroendocrine tumors often show metastases at the time of diagnosis, those without a syndrome do not. The aim of the study was to search for clinicopathological features that may distinguish the two categories of gastrin-producing duodenal tumors. In a retrospective study, we analyzed the clinical and pathological data in a series of 41 patients with syndromic (i.e., gastrinomas) or non-syndromic duodenal gastrin-producing neuroendocrine tumors (ns-gas-NETs). Twenty-four (59 %) of the 41 patients had tumors that were associated with a ZES and were classified as gastrinomas. These tumors showed a higher Ki-67 index than that of the ns-gas-NETs (1.74 vs. 0.85 %, p = 0.012). In addition, they had more lymph node metastases (75 vs. 6 %, p=III (75 vs. 6 %; p< 0.001) than their non-syndromic counterparts. Gastrinomas were removed surgically, ns-gas-NETs endoscopically. We did not observe any significant differences in overall survival or recurrence of disease. Duodenal gastrinomas show no clear morphological features that
distinguish them from their non-syndromic counterparts. However, the patients with gastrinomas present in a more advanced stage of disease and need surgical treatment, while non-syndromic gastrin-producing duodenal NETs may be cured by complete endoscopical removal.