Endocrine Precursor Lesions and Microadenomas of the Duodenum and Pancreas with and without MEN1: Criteria, Molecular Concepts and Clinical Significance

Abstract:

Proliferative changes in the neuroendocrine cells that precede neoplasia are of interest for the understanding of tumorigenesis and the early recognition of neuroendocrine tumors. This review focuses on precursor lesions of duodenal and pancreatic neuroendocrine tumors in multiple endocrine neoplasia type 1 (MEN1) and also discusses 2 new disease entities of pancreatic microadenomatosis. The gastrinomas observed in MEN1 are almost exclusively localized in the duodenum and are multicentric. It has been shown that, in contrast to sporadic duodenal gastrinomas, they are associated with hyperplastic gastrin cell lesions and tiny gastrin-producing microtumors less than 500 µm in diameter. In the pancreas, microadenomatosis (multiple tumors up to 5 mm in diameter) is a feature of MEN1. These microadenomas predominantly express glucagon and pancreatic polypeptide, but do not cause a hormonal syndrome. Approximately 50% of MEN1 minigastrinomas in the duodenum and almost all microadenomas in the pancreas show allelic deletion of the MEN1 gene and therefore may represent ‘initial’ neoplasms. In contrast, endocrine cell precursor lesions retain heterozygosity. Pancreatic microadenomatosis was also found unassociated with hereditary syndromes and 2
monohormonal types were identified: (1) glucagon-producing microadenomatosis and (2) insulin-producing microadenomatosis, both associated with macrotumors. Whether these types of microadenomatosis represent novel disease entities and how to diagnose and treat these patients remains to be clarified by further studies.

Stichworte: Endocrine tumor; Gastrinoma; MEN1 mutation; Insulinoma; Zollinger-Ellison syndrome

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