Hereditary neuroendocrine tumors of the gastroenteropancreatic system.

Abstract:
Approximately 5-10% of neuroendocrine tumors (NETs) of the gastroenteropancreatic system (GEP) have a hereditary background. The known inherited syndromes include multiple endocrine neoplasia type 1, neurofibromatosis type 1, von Hippel-Lindau disease, and the tuberous sclerosis complex. This review discusses for each of these syndromes the: (1) involved genes and specific types of mutations, (2) disease prevalence and penetrance, (3) affected neuroendocrine tissues and related clinical syndromes, (4) special morphological features of NETs and their putative precursor lesions. In addition, GEP-NETs clustering in individual families or associated with other malignancies without known genetic background are discussed.

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