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Titel des Beitrags: Insulinomatosis: a multicentric insulinoma disease that frequently causes early recurrent hyperinsulinemic hypoglycemia.

Abstract:
BACKGROUND: Multicentric insulinoma disease was characterized with regard to its histopathology, multiple endocrine neoplasia type 1 (MEN1) status, precursor lesions, and the risk of hyperinsulinemic hypoglycemia recurrence. METHODS: Fourteen patients with multicentric insulinoma disease were compared with 267 patients with sporadic and familial insulinomas. The tumors were classified according to the World Health Organization (WHO) criteria. The MEN1 status was defined clinically and by germline mutation analysis. Detection of the MEN1 gene locus was performed using fluorescence in situ hybridization. The surgical interventions and the duration of disease-free survival were recorded. RESULTS: Fourteen patients (5%) without evidence of MEN1 showed 53 macrotumors and 285 microtumors expressing exclusively insulin. In addition, they had small proliferative insulin-expressing monohormonal endocrine cell clusters (IMECCs). No allelic loss of the MEN1 locus was detected in 64 tumors. All but one patient had benign disease. Recurrent hypoglycemia occurred in 6/14 patients (11 recurrences; mean time to relapse 8.4 y). Thirteen patients with MEN1 (4.6%) showed 41 insulinomas
and 133 tumors expressing islet hormones other than insulin. IMECCs were not detected. Allelic loss of the MEN1 locus was found in 17/19 insulinomas. Recurrent hypoglycemia occurred in 4/13 patients (4 recurrences; mean time to relapse 14.5 y). Solitary insulinomas were found in 254/281 patients (90.4%). IMECCs were absent. There was no recurrent hypoglycemia in 84 patients with benign insulinomas. CONCLUSIONS: Insulinomatosis is characterized by the synchronous and metachronous occurrence of insulinomas, multiple insulinoma precursor lesions, and rare development of metastases, but common recurrent hypoglycemia. This disease differs from solitary sporadic and MEN1-associated insulinomas.