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Multiple Endocrine Neoplasia (MEN1) Patients Need Close Monitoring and Aggressive Management

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Background: Multiple Endocrine Neoplasia (MEN1) patients develop primary hyperparathyroidism, pituitary adenomas and pancreatic neuroendocrine tumors (PNETs; functioning or non-functioning). The major source of disease-specific mortality is liver metastasis from PNETs. In 2006 we showed that early diagnosis and intervention could increase overall disease free survival. To further expand on this work we have revisited our experience with patients with MEN1 and PNET who underwent at least one pancreatic operation.

Methods: All patients had either radiographic and ± histologic PNET in the setting of genetically or clinically confirmed MEN1. Patients who had pancreatic resection were included in the study.

Results: 171 cases had a PNET out of a total of 487 patients with confirmed MEN1. 103 patients (52 males: 51 females) met the inclusion criteria. Median age was 43.08 years old, (SD:13.5). The most common mutation identified was

210_211delCC in 5 cases. Mutations in menin exon numbers 2 and 10 were the most frequent (19 and 15 cases respectively). Nonsense mutations occurred in 24 cases. The type of PNETs identified included nonfunctioning (NF, 45.6%), insulinoma (20.5%), glucagonoma (2.9%), PPoma (2.9%), and VIPoma (1.0%). In addition, 27.2% of patients presented clinically with synchrononous duoduodenal gastrinoma together with NF PNET. Disease status at presentation was local-regional (LR) in 91.3% and distant metastasis (DM) in 8.7%. The median OS for LR patients was 22.27 years and for DM patients was 12.80 years ($p < 0.001$). OS was statistically different between different types of PNETs [Median OS for Insulinoma, Glucagonoma, non-functioning tumors were 44.04, 19.55, 22.27 years respectively ($p=0.027$)].

Conclusions: Overall survival of PNETs is related to the functional status of the tumor and the aggressiveness of the disease at presentation. PNETs in patients with MEN1 should be diagnosed as early as possible and managed with surgical resection in appropriate candidates.

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