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Long-term Follow-Up of Pheochromocytoma/ Paraganglioma (PPGL) after first diagnosis: a retrospective single-center study of 173 patients

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Background: Pheochromocytoma (Pheo)/Paraganglioma (PGL), together PPGL, are rare but life-threatening tumors, with tumor relapse (TR) rates after initial surgery of 16.5%, and genetic mutations in about 30-40% of patients. Long-term follow-up (FU) and genetic testing in all patients are recommended. Questions remain, however, as to the most suitable duration of FU, the prognostic value of genetic diagnoses and of clinical, biochemical or radiological findings during FU, of the incidence of TR after recommended 10 years FU of low-risk sporadic Pheos, or of TR on morbidity and mortality.

Objective, patients, and methods: We retrospectively determined the overall survival (OAS), progression-free survival (PFS) and tumor-recurrence (TR) in 173 patients with PPGL initially operated and followed-up at a single tertiary referral center from 1988 to 2020 by Kaplan-Meier Estimates, and assessed age, sex, hormonal activity, tumor size, and metastatic spread at first diagnosis, pathohistological PASS score, mutation-positive PPGLs (MP-PPGL), TR, and comorbidities (associated with increased cardiovascular risk) as to independent prognostic markers thereof by multivariate Cox-regression. Certificats of the Austrian death registry were also obtained.

Results: In 8 (5%) PPGLs there was no FU in the charts, but death certified occurring 65±28 (mean±SD, median 2.7) months after first diagnosis. Mean (±SD) age of the remaining 165 (43.9% female) patients (94.5 Pheos, 3.0% multiple PGLs, 2.5% head-and-neck PGLs) was 49(±16) years, mean (range) FU 90(3-537) months. 93 (54%) patients underwent genetic testing, 37 (40%, 21% of entire cohort) had MP-PPGL (mean±SD age 33.8±13.2 yrs, OAS 391 months). 5 of these 37 (13.5%), and 7 of 57 (12%) mutation-negative (MN-PPGL, mean±SD age 53.3±14.8 years, $p<0.01$ vs. MP-PPGL, OAS 190 months) died during FU ($p=0.03$). Male sex, higher age, presence of comorbidities, and primary metastatic disease were independent negative prognostic factors for OAS by univariate, but only age and metastatic disease remained significant by multivariate analysis. As to PFS, higher age and no hormonal activity were negatively associated by univariate, but not by multivariate analysis, as were all other tested parameters.

Conclusions: The cohort of our study is the third-largest original report on long-term FU and prognostic markers in PPGL. While others (Ayola-Ramirez et al, JECM 2011) in 371 patients found tumor size and sympathetic PGL independently associated with OAS, we did not. Patients may have harbored more aggressive tumors in that study, as higher primary metastatic spread (25% of Pheos, 60% of sympathetic PGL) was identified. We identified only age and metastatic disease as independent predictors of OAS, in line with data from Korea (Kim J H, et al., Endocrinol Metab. [Seoul]) in 1048 patients (9% with primary metastatis) and with a metaanalysis comprising 703 PPGLs (Crona J, Endocr Relat Cancer, 2019).

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