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Use of Somatostatin Analog Therapy in Patients with Advanced Pheochromocytoma/Paraganglioma and Somatostatin-Receptor Avid Disease

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BACKGROUND: Neuroendocrine tumors (NETs) commonly express somatostatin receptors (SSRs), as assessed by SSR scintigraphy or somatostatin analog (SSA) radiolabelled PET imaging. As a result, SSAs are used in the standard management of advanced NETs and have demonstrated anti-tumor benefit in prospective trials. Importantly, pheochromocytoma/paraganglioma NETs (PPGL) may also express SSRs at a level comparable to other NETs. However, the potential benefit of SSAs in PPGL remains largely unknown, with an existing literature largely limited to individual case reports. Our objective was to compare the clinical courses of a series of advanced PPGL patients with SSR avid disease treated with or without a SSA.

METHODS: Retrospective analysis of advanced PPGL patients enrolled in a single-institution database (N = 76). Major clinical events of interest included the initiation of systemic therapy, radiation therapy, radiopharmaceutical therapy, surgical debulking, symptomatic deterioration, or death. The Kaplan-Meier method was used to estimate clinical progression-free survival (cPFS), defined as the time from SSR imaging to the first major clinical event following the initial treatment.

RESULTS: Twenty patients underwent SSR imaging (N = 14 with SSR avid disease, N = 6 with SSR non-avid disease). Of the 14 patients with SSR avidity, 8 received subsequent SSA therapy. Four of 8 patients received SSA monotherapy,

while the remaining 4 received SSAs in combination with alternative systemic therapies. Seven of 14 patients developed clinical progression during study follow-up. The median cPFS was 24.7 months (IQR 7.1 – 24.7) and 20.8 months (IQR 15.5 – NR) for patients receiving and not receiving SSA therapy, respectively.

CONCLUSION: A significant portion of advanced PPGL patients demonstrate SSR avid disease and may be treated with SSA therapy, either alone or in combination with other systemic therapies. SSA therapy may provide a delay in clinical progression. This will be prospectively evaluated in a planned randomized trial.