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Post-Operative Biochemical Surveillance Thresholds Can be Used to Monitor for Sympathetic Pheochromocytoma/Paraganglioma Recurrence and Metastasis

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BACKGROUND

Sympathetic pheochromocytomas and paragangliomas (PPGLs) are rare neuroendocrine tumors associated with excess catecholamine production. Routine biochemical and imaging surveillance to monitor for recurrence and metastasis is recommended. However, there is limited data describing optimal surveillance approaches and post-operative biochemical thresholds for detecting recurrences or metastases. This study sought to correlate biochemical and imaging surveillance with recurrences or metastases.

METHODS

Patients who underwent surgery for PPGLs at a tertiary-care cancer hospital between 2000-2021 were retrospectively reviewed. Patients with <3 years of post-operative biochemical surveillance or with early locoregional recurrence (LRR) or metastasis at presentation were excluded. Surveillance catecholamine values and imaging were compared between patients with and without LRR/metastasis.

RESULTS

Eighty patients treated for PPGLs met inclusion criteria. Median follow-up was 72 months (IQR 48.5-98.5). Seventy-one patients (89%) had genetic testing, with 40 patients (50%) having a PPGL-susceptibility mutation. Thirteen patients (16%) developed recurrence (local n=8, contralateral de novo n=2, metastasis n=3). LRR/metastases were predominately from pheochromocytomas (n=12); one paraganglioma developed diffuse metastases. Nine of 13 patients with LRR/metastasis had an identified PPGL-susceptibility mutation (RET 6, VHL 2, SDHD 1). Patients with LRR/metastasis were younger at presentation than those without LRR/metastasis (22 years vs 48 years, p=0.025). No difference in gender, tumor size, or surgical approach was observed between groups. All patients with LRR/metastasis had evidence of biochemical excess preoperatively. Median time to identification of LRR or metastasis was 38 months (IQR 20-71.5). Biochemical surveillance was the most commonly used surveillance modality (91% of patients at 1 year). Only 45 patients (56%) had both biochemical and imaging surveillance at 1 year. Nine LRR/metastases were initially identified by biochemical surveillance versus 4 LRR/metastases identified by imaging first.

All 4 recurrences/metastases identified by imaging first were patients who had undergone cortical-sparing adrenalectomy in the context of a PPGL-susceptibilty mutation. Eleven patients with LRR/metastasis had normetanephrine levels ≥2 times the upper limit of normal (ULN). Although patients without LRR/metastasis intermittently had minimally-elevated metanephrine levels, none exceeded twice the ULN.

CONCLUSIONS

Routine postoperative biochemical surveillance in patients with functional PPGL may be adequate to detect development of LRR/metastasis. Normetanephrine levels ≥2 times the ULN are suggestive of LRR/metastasis. Elevated post-operative metanephrine levels are observed but did not exceed ≥2 times the ULN in patients without LRR/metastasis. Surveillance metanephrines were commonly the sole modality needed in identifying LRR/metastasis, except for in cases of cortical-sparing adrenalectomy. This data may be helpful in determining optimal modalities for long-term surveillance.

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