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Head and Neck Paragangliomas: Overview of Institutional Experience

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BACKGROUND

Paragangliomas of head and neck (HNPPGLs) are rare and mostly slow growing tumors arising from neural crest-derived cell clusters and have high rate of genetic mutation, up to 40% germline mutations and 30% somatic mutations.

METHODS

Retrospective review of institutional experience with clinical evaluation and management of HNPPGLs.

RESULTS

Among 174 patients with HNPPGLs diagnosed from 1981 to 2023, 116 (67%) were women. The mean age at first tumor diagnosis was 50 years, ranging 11 to 82 years. Among 242 tumors identified for the entire cohort; carotid body tumors were the most common type (101), followed by jugular (59) and tympanic (27). Most patients had one tumor; however, many had multiple tumors (ranging 2 to 8), especially those with genetic mutations. Multiple tumors could present at the time of detection or developed over the years, sometimes over a decade apart. Among 98 patients (56% of cohort) with genetic screening, only 2 had additional somatic testing. Among 60 patients (61% of tested or 34.5% of cohort) with genetic mutations, SHDx mutations accounted for the vast majority, including SDHB (25), SDHD (21), SDHC (7), and SDHA (2). A few other known PPGL related mutations were identified, including VHL (1), MEN1 (1) and TMEM127 (1). HNPPGLs tend to be less biochemically active compared to non-HNPPGL. 118 patients (67.8% of cohort) had at least one biochemical evaluation of metanephrines or catecholamines. If biochemistries were elevated, patients were blocked with alpha antagonists prior to any procedures. CT and MRI were the most common imaging modalities. Functional scans with somatostatin analogs were used to evaluate multifocal and metastatic disease. Among the entire cohort, 120 patients had surgery, 22 had radiation therapy, and 5 had MIBG as treatment. Surgery remained the most definitive and commonly used treatment for local disease control, though radiation with stereotactic radiosurgery (SRS) or conventional external beam radiation (EBRT) was used in surgically challenging or inoperable cases. The current practice at our institution has evolved to refer all patients with suspected HNPPGLs for genetic screening and biochemical evaluation. We also have weekly multidisciplinary meetings to discuss the care.

CONCLUSIONS

Our experience highlights the need for referral for genetic testing and biochemical evaluation and for a team based approach to improve the clinical outcomes of patients with HNPPGLs.

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