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Recurrent Tympanic Paragangliomas: An Overview of Institutional Experience

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BACKGROUND

Tympanic paragangliomas (TPGLs) are rare and mostly slow growing head and neck tumors of the lateral skull base. TPGLs are neuroendocrine tumors arising from neural crest-derived cell clusters. Recurrence rates in the literature have been reported at 0 to 15%, depending on cohort size and mean follow up time.

METHODS

Single-institution chart review of tertiary care hospital from 2009-2025.

RESULTS

Among the cohort of 43 patients diagnosed with TPGLs from 2009 to 2025, all patients had a unilateral TPGL, 40 (91%) were women and 8 had recurrent TPGLs (18%). The primary TPGLs were diagnosed from 1994 to 2019, and the recurrent TPGLs were diagnosed between 2010 and 2024. The mean age at the primary TPGL diagnosis was 51.5 years (range 18 to 76 years). The mean age at the recurrence was 62.5 years (range 19 to 83 years). The mean interval of recurrence was 11 years (range 1-20 years). Among this cohort, one patient with recurrent TPGL had an additional pheochromocytoma diagnosed at the same time as the TPGL, and one patient without recurrent TPGL had multiple additional HNPGLs including a facial nerve, bilateral carotid and bilateral vagal PGLs. All patients underwent surgery for their primary TPGLs. For recurrent TPGLs, six patients (75%) had surgical resection while two patients opted for observation. Among the six patients who underwent surgical resection, three required tympanoplasty due to adherent tumor, two required an extensive skull base approach due to expansile tumor growth requiring an infratemporal fossa approach with a fat graft, and one underwent a radical mastoidectomy with closure of the external auditory canal due to extensive recurrent tumor and cholesteatoma. Among the eight patients with recurrent TPGLs, four had genetic testing and one had SDHD mutation. Two recurrent TPGLs were considered biochemically active with either elevated metanephrines or catecholamines. Surgery remained the definitive treatment for both primary and recurrent TPGLs.

CONCLUSIONS

The current practice at our institution has evolved to have all patients with TPGLs go for genetic and biochemical evaluation. We also have weekly multidisciplinary meetings to discuss the management. Given the high rate (18%) and long interval (mean 11 years) of recurrence, we believe that TPGLs require long-term follow-up.

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