Chapter 4

HEAD AND NECK PARAGANGLIOMAS: IMAGING FINDINGS

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Introduction

Head and neck paragangliomas (HNPs) are hypervascular, slow-growing, mostly benign neoplasms that arise from paraganglionic cells of the autonomic nervous system (1). It is estimated to make up 3% of all paragangliomas (PGLs), 0.6% of all head and neck cancers and 0.03% of all tumours (1,2). HNPs can be of sympathetic or parasympathetic paraganglionic origin. The majority of HNPs originate from parasympathetic paraganglia, are usually biochemically silent and less than 4% of them secrete catecholamines (3).

Etiology and Genetic

HNPs are particularly high risk in women aged 50-70 years who are exposed to chronic hypoxia living at high altitudes. They can be sporadic or inherited. About 30- 40% of HNPs are hereditary, usually develops in the carotid body, and up to 80% is multifocal, tending to occur earlier than sporadic paragangliomas (2,4-6).

Hereditary HNPs are mainly associated with germline mutations in the enzyme succinate dehydrogenase (SDH), a multiprotein complex consisting of SDHB, SDHC, SDHD, SDHA and SDHAF2. SDH subunit mutation is frequently associated with bilateral or multifocal parasympathetic HNPs (4-6).

While sporadic HNPs are usually benign, the risk of malignancy increases in the presence of SDHB mutation. Malignancy is most commonly found in vagal PGLs (10-19%) and metastases are often to cervical lymph nodes. Distant metastases are very rare, such as in the lungs, liver and bones (1,5). There are no known histopathological criteria defined for malignancy. Malignant paraganglioma is

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