The Molecular Subtypes of Pheochromocytoma and Paragangliomas (PPGLs): The Cancer Genome Atlas Transcriptome Data Derived Taxonomic Scheme and 2017 WHO Classification System

All Day Room: MS Community, Learning Center Digital Education Exhibit

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TEACHING POINTS
Pheochromocytomas and paragangliomas (PPGLs) are tumors arising from adrenal and extra-adrenal chromaffin cells. Currently associated with germline and/or somatic mutations in more than 20 genes. Up to 40% are associated with inherited germline mutations, most common genes include SDHB, SDHD, VHL, RET and NF1. Genotype/phenotype correlations exist between germline mutation, tumor location and biochemical secretion. Four main molecular subtypes are identified: 1. Pseudohypoxia, 2. Kinase-signaling, 3. Wnt-signaling and 4. Cortical admixture. SDHB mutations are associated with increased risk of metastatic disease. Understanding tumor biology has expanded the scope of personalized care and treatment in PPGLs.

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