Pheochromocytoma

- Revisiting radioresistance: a systematic review of outcomes of Stereotactic radiosurgery (SRS) in functional head and neck paragangliomas
- Pediatric and adolescent von Hippel-Lindau disease: tumor profiles, genotype-phenotype correlation and comparison with adults
- Resveratrol attenuates the CoCl₂-induced hypoxia damage by regulation of lysine β-hydroxybutyrylation in PC12 cells
- Factors that predict progression of von Hippel-Lindau disease-related malignancy: a longitudinal cohort study
- Von Hippel-Lindau Disease : A Comprehensive Review of Diagnosis, Genetics, Clinical Challenges, and Surveillance
- Overlapping presentations and diverse genetic defects characterize neuroendocrine neoplasms in a Mexican cohort
- Olfactory mucosa-mesenchymal stem cells with overexpressed Nrf2 modulate angiogenesis and exert anti-inflammation effect in an in vitro traumatic brain injury model
- Perioperative management of patients with glioblastoma copresenting with pheochromocytoma: illustrative case

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20% of pheochromocytomas is associated with von Hippel-Lindau disease. PCC occur in 7–20% of families with VHL

General information

Located in the adrenal gland. Maybe sporadic, or as part of a familial syndrome (von Hippel-Lindau disease, MEN 2A & 2B, & neurofibromatosis). Consider genetic testing if age at diagnosis is < 50 years for mutations of VHL and other genetic abnormalities (RET, SDHS, SDHB, SDHC [1]).

A pheochromocytoma (from Greek phaios “dark”, chroma “color”, kytos “cell”, -oma “tumor”) or phaeochromocytoma (PCC) is a neuroendocrine tumor of the medulla of the adrenal glands (originating in the chromaffin cells), or extra-adrenal chromaffin tissue that failed to involute after birth and secretes high amounts of catecholamines, mostly norepinephrine, plus epinephrine to a lesser extent.

Extra-adrenal paragangliomas (often described as extra-adrenal pheochromocytomas) are closely related, though less common, tumors that originate in the ganglia of the sympathetic nervous system and are named based upon the primary anatomical site of origin.

Case reports

A unique case of metastatic pheochromocytoma of the cervical spine treated with preoperative embolization and subsequent en bloc resection. A 65-year-old man with metastatic pheochromocytoma presented with two weeks of worsening neck pain, left arm and leg weakness and

paresthesia, and urinary incontinence. Magnetic resonance imaging showed a metastatic osseous lesion at C6 with severe stenosis and spinal cord compression. The patient underwent successful preoperative angiographic embolization with a liquid embolic agent followed by C5-C7 laminectomy, en bloc tumor resection, and C3-T2 posterior spinal fusion. Six weeks postoperatively, the patient reported improving strength and resolving neck pain and paresthesias. While there is no standard paradigm for the treatment of metastatic pheochromocytomas of the cervical spine, preoperative embolization may minimize intraoperative blood loss and hemodynamic instability during subsequent surgical resection [2].

1)
van Nederveen FH, Gaal J, Favier J, et al. An immunohistochemical procedure to detect patients with paraganglioma and phaeochromocytoma with germline SDHB, SDHC, or SDHD gene mutations: a retrospective and prospective analysis. Lancet Oncol. 2009; 10:764–771
2)
Singh A, Santangelo G, Ellens N, Kohli G, Pranaat R, Bender MT. Preoperative embolization and en bloc resection of a metastatic pheochromocytoma of the cervical spine. J Cerebrovasc Endovasc Neurosurg. 2024 Jun 20. doi: 10.7461/jcen.2024.E2023.04.005. Epub ahead of print. PMID: 38897596.