Multiple hemangioblastoma

Hemangioblastomas may arise sporadically in a solitary form or associated with Von Hippel-Lindau disease with multiple tumors. Surgery is the mainstay treatment, but management is challenging in case of recurrent and/or multiple tumors. VHL protein is defective in both forms of hemangioblastoma, leading to the accumulation of hypoxia-inducible factor, stimulating angiogenesis via VEGF and PDGF mainly [1].

Ryba et al. reported a case of multiple hemangioblastomas involving two radiologically silent lesions only detected intraoperatively by Indocyanine green fluorescence. A 26-year-old woman presented with a cystic cerebellar tumor on the tentorial surface of the left cerebellar hemisphere on MRI. A left paramedian suboccipital approach was performed to remove the mural nodule with the aid of ICG injection. The first injection applied just prior to removing the nodule, highlighted the tumor and vessels. After resection, two new lesions, invisible on the preoperative MRI, surprisingly enhanced on fluorescent imaging 35 minutes after the ICG bolus. Both silent lesions were removed. Histological analysis of all three lesions revealed they were positive for HB. The main goal of this report is to hypothesize possible explanations about the mechanism that led to the behavior of the two silent lesions. Intraoperative ICG video angiography was useful to understand the 3D angioarchitecture and HB flow patterns to perform a safe and complete resection in this case. Understanding the HB ultrastructure and pathophysiological mechanisms, in conjunction with the properties of ICG, may expand potential applications for their diagnosis and future treatments [2].

Migliorini et al. reported a 37-year-old woman's case with recurrent and rapidly progressive VHL-associated hemangioblastomas, causing severe disability. She was treated 24 months with pazopanib, a multi tyrosine kinase inhibitor (TKI) targeting VEGF and PDGF-β pathways. Despite moderate radiological changes, progressive improvement in her clinical condition persisting over 3 years was observed. Inhibiting angiogenesis is a therapeutic option that may improve the quality of life and the autonomy of VHL patients disabled with multiple hemangioblastomas [3].

[1], [3]
Migliorini D, Haller S, Merkler D, Pugliesi-Rinaldi A, Koka A, Schaller K, Leemann B, Dietrich PY. Recurrent multiple CNS hemangioblastomas with VHL disease treated with pazopanib: a case report and literature review. CNS Oncol. 2015;4(6):387-92. doi: 10.2217/cns.15.22. Epub 2015 Oct 26. PMID: 26497655; PMCID: PMC6083944.

[2]
Ryba AS, Sales-Llopis J, Wolfsberger S, Laakso A, Daniel RT, González-López P. Utility of indocyanine green in the detection of radiologically silent hemangioblastomas: case report. J Neurosurg. 2021 Feb 12:1-7. doi: 10.3171/2020.8.JNS202176. Epub ahead of print. PMID: 33578384.