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ABSTRACT

Relief of Intractable Nausea after Resection of Brainstem Hemangioblastoma in Patients with von Hippel-Lindau Disease: a Clinical Series

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Symptoms of hemangioblastomas in VHL depend on location, and the common predilection for cerebellum and brainstem lead to issues with balance, strength, coordination, and cranial nerve palsies. We present five patients who suffered intractable nausea from hemangioblastoma located in or near the distal medulla. All had VHL, and 4/5 had a family history for that condition. Age ranged from 19 to 48 years at time of surgery, 4/5 patients were female, and 2/5 had an associated cyst. Relief in all cases occurred within one week after tumor removal. The physiological basis of the nausea is hypothesized to be disruption of function of the area postrema. This circumventricular organ is situated just cranial to the obex in the floor of the distal fourth ventricle, and induces nausea and vomiting when visceral afferents or afferents from extramedullary brain centers are stimulated or when chemical changes in blood activate chemoreceptors there. In all patients here reported, tumor either sat immediately adjacent to or within the area postrema, or tumor cyst compressed it. Etiologies of the nausea include direct disruption or compression of the area postrema, or shifts in perfusion of this region caused by tumor hypervascularity and AV shunting. Nausea in patients with VHL can be caused by other conditions associated with the disease (e.g., pancreatic cysts or tumors), but this symptom should trigger a search for hemangioblastoma of the distal medulla, which can easily be shown by MRI. As such patients are resistant to anti-emetic medications, surgery can offer much benefit.