

Spinal Hemangioblastomas in von Hippel-Lindau Disease: Clinical and Radiological Characteristics

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Background: Hemangioblastomas in the central nervous system (CNS) are common manifestations in Von Hippel-Lindau disease (VHL). Clinical follow-up and regular Magnetic Resonance imaging (MRI) of the CNS are mandatory. Periodical surveillance should lead to timely intervention to prevent irreversible morbidity. Asymptomatic spinal hemangioblastomas may be present over a long period and progression may be slow. However, the relation between symptoms and radiological findings of spinal hemangioblastomas remains unclear.

Aim of the Study: To investigate the relation between MRI findings of spinal hemangioblastomas and symptoms with a special focus on peritumoural edema and spinal cysts.

Method: VHL patients under surveillance at the UMCG for at least two years between 2000-2017 were eligible for evaluation. Symptoms defined as hypesthesia, weakness, ataxia and hyperreflexia were retrospectively scored from the medical charts. MRIs were re-scored for the presence of edema (T2) and cysts. The association between symptoms and the presence of peritumoural edema was evaluated using descriptive statistics and 2-sided Fisher's Exact Tests.

Results: Forty-three (23 male, 20 female, mean age 34 ± 16 y) patients were evaluated, 28 patients (65%) showed in total 78 spinal hemangioblastomas. Eighteen patients with a spinal hemangioblastoma experienced symptoms. Ten patients had spinal cysts and 9 patients showed peritumoural edema on the T2 MRI. Nine patients with spinal cysts and eight patients with peritumoural edema had symptoms. Both peritumoural edema and spinal cysts were associated with symptoms ($p=0.009$ respectively $p=0.041$). 11/28 patients underwent interventions (surgical resection, arterial embolization or radiotherapy) during study period.

Conclusions: The presence of peritumoural edema and/or spinal cysts on MRI in VHL-patients with spinal hemangioblastomas is associated with symptoms and may alert the clinician, to intensify the radiological and neurological follow up in order to prevent irreversible morbidity.