9º Simpósio Médico Internacional de VHL
9th International Medical Symposium on VHL
III Encontro de Famílias com a Síndrome de VHL
3rd VHL Family Meeting

Rio de Janeiro • October 2010

ABSTRACT

Management of Brainstem and spinal cord Hemangioblastomas in Patients with von Hippel-Lindau Disease

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Introduction: Spinal cord hemangioblastomas are associated with von Hippel-Lindau disease (VHL) in more than 75% of cases. Brainstem hemangioblastomas are present in up to 20% of VHL patients, and are almost pathognomonic of the disease. Management of these tumors is controversial, as patients are not affected from isolated hemangioblastomas, but by a genetic multineoplastic condition. The aim of this paper is to present the surgical results of spinal and brainstem hemangioblastomas in a VHL referral center.

Material and methods: We reviewed a series of 12 patients (age range 15-66) who underwent 14 surgical procedures to remove 20 hemangioblastomas: 4 in the brainstem, 4 in bulbo-medullary junction, and 12 in the spinal cord. The indication for surgery was established by initiation of symptoms or evident growth of tumor.

Results: Preoperative sensory deficit was present in 10 patients (76.9%), followed by motor deficit in 7 (53.8%). Complete resection was possible in all cases. In the postoperative functional assesment, improvement was obtained in 1 (7.7%), clinical stability in 12 (84.6%) and clinical deterioration in 1, from functional grades I to II (7.7%). Early in the postoperative assesment, a functional deterioration occurred in 4 (30.8%) patients, all fully recovered after 3 months, excepting the abovementioned case.

Conclusions: Complete microsurgical resection of spinal cord and brainstem hemangioblastomas in VHL patients is achieved with good results and at very low rate of complications, when performed by surgeons particularly involved in the management of patients with VHL disease