



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

Gamma Knife for Intracranial Hemangioblastomas in von Hippel-Lindau Patients. When and how?

María Elena Kusak, Nuria Martínez, Jorge Gutiérrez, José María de Campos, Germán Rey, Roberto Martínez Unidad Gamma

Hospital Ruber Internacional; Neurooncología Familiar, Fundación Jiménez Díaz, UAM, Madrid, Spain

Introduction: Hemangioblastomas (HGBs) are a distinctive feature of von Hippel-Lindau (vHL) disease, with successive surgical treatments being a major cause of morbidity and mortality. Radiosurgery has become an option in their treatment.

Objective: The analysis of our results and those published of Gamma Knife (GK) Radiosurgery for intracranial (IC) HGBs, focusing on vHL's patients. Methods: Between 1994 and 2010, 18 treatments in 14 patients (7 males/7 females) with a total of 40 HGBs have been performed. Fourteen treatments have a complete follow-up. Mean age was 37.4 years. The mean marginal dose was 13.9 Gy, with a mean prescription isodose of 59.4% and a mean treated volume of 4.4 cm³. Six patients had a vHL diagnosis. Results: The mean follow up time has been 4 years. The local volumetric control was obtained in all but three patients. In all vHL patients other location HGBs appeared during follow up.

Conclusions: Gamma Knife radiosurgery is an effective option to surgery in the treatment for vHL patient's hemangioblastomas. Due to the steep dose decay and minimal peripheral irradiation of healthy tissues, it should be the preferred radiotherapeutic technique. Having in mind the genetic condition of this disease, where the potential oncogenic effect of radiotherapy should be taken into account, any therapeutic decision must be evaluated individually. This treatment must be used in patients with lesions with evident growth or with progressive symptoms, when surgery is not a safe option in a vHL experienced neurosurgical unit.