Neurocirugia. 2018;29(Espec Congr):194

Neurocirugía

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V0391 - INTRAMEDULLARY CERVICAL HEMANGIOBLASTOMA IN A PATIENT WITH VON HIPPEL-LINDAU DISEASE. SURGICAL GROSS REMOVAL WITH EXCELLENT CLINICAL RESULT

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Resumen

Objectives: Intramedullary hemangioblastomas (HMBs) are hypervascular lesions which are common on von Hippel-Lindau disease. Spinal HMBs are rare – comprise 1.6-6.4% of spinal cord tumours – and occur mainly intramedullary. Motor and sensitive deficits, syringomyelia and scoliosis may be seen on presentation. Surgical resection is challenging since they are highly haemorrhagic and the strategy to aim a complete resection resembles Arteriovenous Malformations – arterial feeders may be occluded before tumour extraction.

Methods: A 26-year-old female with von Hippel-Lindau disease, previously operated to posterior fossa for removing two HMBs, presented with cervical pain with irradiation to left arm. Later, she developed a motor deficit on elbow extension. A cervical MRI was performed showing an intramedullary hyperintense lesion on T1 sequences with 13 × 15 mm with a left paramedian location to the level of C5-C6. It was associated with a syringomyelic cavity extending superiorly to C1 level and inferiorly to D1. She had no other motor or sensitive deficits.

Results: A surgical video is presented. A laminotomy was performed centred to C4-C6 levels. A gross total removal was achieved after coagulating vascular feeders and venous drainage. Dura-máter was closed in a water-tight fashion and a laminoplasty was made to prevent instability. Patient was admitted on Intensive Care Unit for vigilance with no ventilatory assistance requirement. Post-operatively, she had no new neurological deficits besides those presented on pre-op. She was discharged on fifth day and is on an ambulatory rehabilitation program.

Conclusions: HMBs are benign neoplasms which occur mainly on cerebellum. They can occur spontaneously but are commonly associated to von Hippel-Lindau disease. Radiological findings may suggest the diagnosis – lesions are small, superficial and well demarcated, hyperintense in T1, usually associated to large syrinx cavities. Early diagnosis and surgical treatment may prevent persistent neurological deficit which may occur progressively or suddenly since these tumours are prone to bleeding.