Case 15855

Haemangioblastoma (central nervous system)
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Section: Neuroradiology
Area of Interest: Neuroradiology brain
Procedure: Education
Imaging Technique: MR
Special Focus: Pathology Case Type: Clinical Cases
Authors: Andres Diaz, MD. Lina Caro, MD. Marco Charry, MD. Angel Donato, MD.
Patient: 36 years, female

Clinical History:

A 35-year-old man with a clinical course that began 2 months before, with global headache associated with a rotating environment sensation that did not improve with the intake of common analgesics. 8 days before he reported an increase in headache associated with emesis.

Imaging Findings:

MRI of the posterior fossa shows a cystic mass in the right cerebellar hemisphere hypointense on T1 and hyperintense on T2 compared to adjacent brain, with an eccentric solid component which enhances vividly, with no evidence of enhancement of the cyst wall.

Discussion:

Haemangioblastomas are benign neoplasms that originate in the central nervous system, are highly vascular and constitute between 1.6-5.8% of intracranial tumours [1]. The majority are of infratentorial location, mainly affecting the cerebellum [1]. They are usually associated with von Hippel Lindau syndrome (VHL) in 25% [1, 2]. This is an autosomal dominant disorder due to the mutation of the suppressor gene located on chromosome 3p25-26 causing this disease in the central nervous system (CNS), retinal, renal cell carcinoma, visceral cysts, adrenal pheochromocytoma and papillary cystadenoma of the epididymis [2, 3, 4], or may be sporadic in 75%. [2] The anatomopathological study of haemangioblastomas can help us to understand part of the symptomatology due to its expansion of the solid component and the resulting compression of the brainstem and together with the adequate clinical evaluation it is possible to differentiate from VHL and confirm the definitive diagnosis [1]. The haemangioblastomas are derived from embryonic haemangioblasts that contain endothelial and stromal elements. The neoplastic component is the stromal element. The endothelial component, which gives rise to the vasculature of the tumour, is highly permeable, which leads to the formation of a peritumoral cyst comprising ultrafiltered plasma [2]. Microscopically, they are well-defined lesions, although not encapsulated, that can occasionally invade the surrounding tissue [4]. Clinical manifestations are usually signs of endocranial hypertension, epilepsy and hemiparesis in 40.7% of patients and hydrocephalus in 70.4% of patients, accompanied by headache, blurred vision, and occasional vomiting [1, 2].
Diagnosis through imaging techniques is difficult. Studies such as MRI show hypointense T1 and hyperintense T2 cystic lesions with the presence of a mural nodule that shows heterogeneous enhancement with the contrast medium [2, 3]. They are located more frequently in the cerebellum and are less often suspected when they are in a supratentorial location as differential diagnosis infections such as neurocysticercosis, toxoplasmosis, and metastasis [2, 3, 5]. Therefore, brainstem haemangioblastomas present a unique challenge because of their eloquent location and vascularisation. The optimal treatment strategies are the successful total excision of the haemangioblastoma; the radically curable criterion is the surgical removal of the haemangioblastoma in a one-stage or a second-stage operation [2, 3].

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Haemangioblastoma, Posterior fossa metastases, Pilocytic astrocytoma, Glioblastoma (GBM), Medulloblastoma

**Final Diagnosis:** Haemangioblastoma

**References:**


Mai- Lan Ho, MD, Ronald L. Eisenberg MD,JD (2014) Adult and General Brain. Neuroradiology signs pag 32
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Figure 4

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