Case Report

Falcine meningioma in Von Hippel–Lindau disease: An unusual association

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Abstract

**Background:** Von Hippel–Lindau (VHL) disease is an autosomal dominant condition characterized by formation of multiple benign and malignant tumors. In this disease supratentorial lesions are rare and no falcine meningioma has been previously reported. Differential diagnosis is very difficult and the histopathological examination is the definitive method for diagnosis.

**Case Description:** A patient with VHL underwent a suboccipital craniotomy for removal of cerebellar hemangioblastoma and after 2 years magnetic resonance imaging (MRI) showed an iperintense solid mass located at posterior part of the falx. Histological diagnosis revealed meningioma.

**Conclusion:** The only case in the literature of falcine meningioma in a patient with Von Hippel–Lindau disease, discovered during radiological follow-up, is described and a surgical management is proposed.

**Key Words:** Falcine meningioma, hemangioblastoma, Von Hippel-Lindau disease

INTRODUCTION

Von Hippel–Lindau (VHL) disease is a rare condition caused by genetic mutations on chromosome 3,[1,6,12,11] transmitted in an autosomal dominant fashion with near complete penetrance and characterized by formation of multiple benign and malignant tumors, as well as cysts in multiple organs.[7] Affected patients frequently develop retinal and central nervous system hemangioblastomas (HB), clear cell renal cell carcinomas (RCC), pheochromocytomas, pancreatic neuroendocrine tumors, and endolymphatic sac tumors (ELSTs).[13] In VHL disease supratentorial lesions, usually HB, are rare and meningiomas are extremely rare.[11] In literature no case of falcine meningioma in VHL has been previously reported. We describe the first case of meningioma located at the third posterior of the falx in a patient with VHL.

CASE REPORT

A 55-year-old Caucasian female patient underwent a suboccipital craniotomy for removal of left cerebellar...
hemisphere HB. Abdominal computed tomography revealed a pheochromocytoma that was excised. Genetic analysis showed the presence of a VHL gene mutation and an evaluation of the family history demonstrated VHL disease in two of the patient’s siblings. Thus this patient’s condition had been diagnosed as VHL disease. Follow-up magnetic resonance imaging (MRI) performed 1 year after the first operation had shown no evidence of recurrence or abnormal findings in the supratentorial region [Figure 1]. However, MRI performed 2 years after the first operation showed a solid mass with strong enhancement in the right cerebellar hemisphere as recurrence and an iiperintense solid mass located at posterior part of the falk [Figure 2]. MRI performed at 4 years [Figure 3] revealed slow growth of both lesions that were asymptomatic. A frameless stereotactiic biopsy with the Leksell Model G stereotactic system (Elekta, Inc., Norcross, GA) of falicne lesion was performed. The intra and postoperative histological diagnosis was meningothelial meningioma of World Health Organization Grade 1 [Figure 4]. There were no postoperative complications. The patient decided for clinical and radiological follow‑up, temporarily delaying a following definitive surgical or radiotherapical treatment.

DISCUSSION

Melmon and Rosen[14] first proposed clinical criteria for the diagnosis of VHL, recognizing cerebellar HB, the hallmark of Lindau’s tumor. Later Lamiell[10] redefined the clinical diagnosis of VHL including an expanded list of associated visceral tumors, as ELST, RCC, pheochromocytoma, paraganglioma, neuroendocrine neoplasm, and/or multiple cysts of the pancreas. The diagnosis of VHL is made if a patient has at least two central nervous system HB or at least one central nervous system HB and one of the visceral lesions described above or at least one of the visceral lesions previously described above, and a pathogenic mutation in VHL gene or a first-degree relative with VHL.[1] The incidence of VHL ranges from 1 in 40,000 live births and typically it first manifests in the second decade of life.[3] Because of the phenomenon of genetic
In 1995, Bleggi-Torres first reported a case of meningioma in VHL, and in the pertinent literature only two other cases have been previously described [Table 1]. Because of a very difficult preoperative radiological differential diagnosis, the histopathological examination after surgical resection, if indicated, is the definitive method for diagnosis.[19] The main differential diagnoses of these lesions include meningial HB, hemangiopericytoma, and metastatic RCC. Epithelial membrane antigen (EMA) immunostaining can help distinguish supratentorial meningial HB from meningioma.[13] Furthermore, HB tumors are generally negative for glial fibrillary acidic protein and positive for neuron-specific enolase.[17] Finally, vimentin is positive in HB tumors, supporting the hypothesis that HBs originate from the mesenchyme.[3] Recently, inhibin-α has been described as a useful marker for distinguishing HB from angiomatous meningioma.[9] Immunohistochemically, RCC stain positive for EMA whereas HB are EMA negative.[1] Because of the extreme rarity of falcal meningiomas in VHL, treatment modality has never been analyzed. In our opinion, in asymptomatic lesions stereotactic biopsy is suggested to design the next therapeutic step, while in symptomatic or hemorrhagic lesions or asymptomatic lesions that show significant growth during radiological follow-up, maximal resection is recommended to prevent recurrence and postoperative hemorrhage. Radiotherapy is useful for multiple, subtotally resected, and recurrent tumors.

**CONCLUSIONS**

We report a rare case of asymptomatic slow-growing meningioma located at the third posterior of the falx in a patient with VHL, detected 2 years after surgical resection of a cerebellar HB, in which no surgical or radiotherapical treatment has been previously performed. In VHL, supratentorial meningial masses are extremely rare and no other case of falcal involvement has been previously reported. Pathological and immunohistochemical techniques are helpful for differential diagnosis. Regular follow-up is mandatory and surgical resection is the definitive treatment for these lesions.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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**Table 1: Review of meningiomas in VHL**

| Authors          | Sex/Age Location | Histological diagnosis                  |
|------------------|------------------|----------------------------------------|
| Bleggi-Torres    | M/34 Right frontal convexity | Atypical meningioma |
| Governale        | M/37 Right frontal convexity | Fibrous meningioma |
| Kanno            | M/66 Posterior fossa | Meningothelial meningioma |
| Present case     | F/55 Posterior part of the falx | Meningothelial meningioma |