SNI
SURGICAL NEUROLOGY INTERNATIONAL
SNI: Unique Case Observations
OPEN ACCESS
For entire Editorial Board visit : http://www.surgicalneurologyint.com
Editor: Ather Enam, M.D., Aga Khan University, Karachi, Sindh, Pakistan

Case Report

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

Domenico Murrone, Bruno Romanelli, Nicola Montemurro, Domenico Chirchiglia[1], Aldo Ierardi

Unit of Neurosurgery, "Di Venere" City Hospital, Bari, [1]Department of Neurosurgery, University "Magna Graecia", Catanzaro, Italy

E-mail: *Domenico Murrone - doflamingo82@gmail.com; Bruno Romanelli - brunoromanelli@alice.it; Nicola Montemurro - montemurronic@hotmail.com; Domenico Chirchiglia - chirchiglia@unicz.it; Aldo Ierardi - aldo.ierardi@gmail.com
*Corresponding author

Received: 25 October 17 Accepted: 19 December 17 Published: 14 February 18

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

Access this article online
Website: www.surgicalneurologyint.com
DOI: 10.4103/sni.sni_398_17
Quick Response Code:

INTRODUCTION

Von Hippel–Lindau (VHL) disease is a rare condition caused by genetic mutations on chromosome 3,[1,6,12,13] 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

How to cite this article: Murrone D, Romanelli B, Montemurro N, Chirchiglia D, Ierardi A. Falcine meningioma in Von Hippel–Lindau disease: An unusual association. Surg Neurol Int 2018;9:37.
http://surgicalneurologyint.com/Falcine-meningioma-in-Von-Hippel–Lindau-disease:-An-unusual-association/

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 iperintense solid mass located at posterior part of the falx [Figure 2]. MRI performed at 4 years [Figure 3] revealed slow growth of both lesions that were asymptomatic. A frameless stereotactic biopsy with the Leksell Model G stereotactic system (Elekta, Inc., Norcross, GA) of falcine 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.

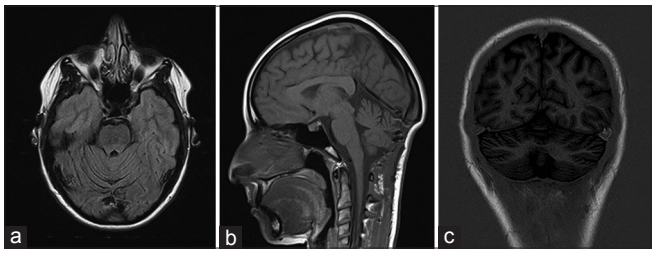

Figure 1: One year postoperative axial (a), sagittal (b), a coronal (c) T1-weighted MR images showing no evidence of recurrence or abnormal findings in the supratentorial region

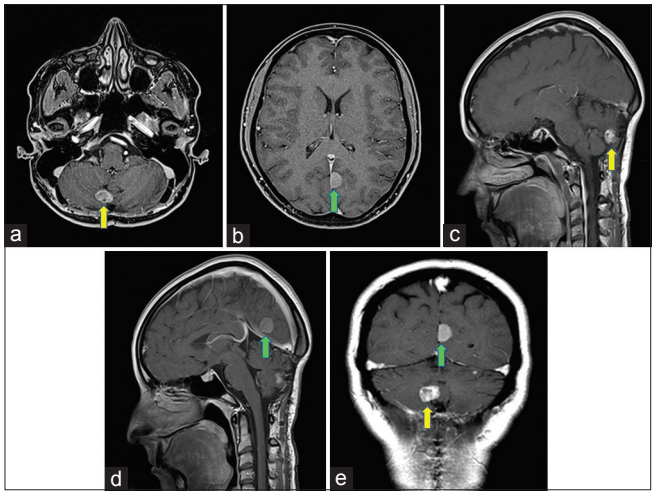

Figure 3: Four years postoperative axial (a and b), sagittal (c and d), a coronal (e) T1-weighted MR images with gadolinium demonstrating further slow growth of both lesions described in Figure 2

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

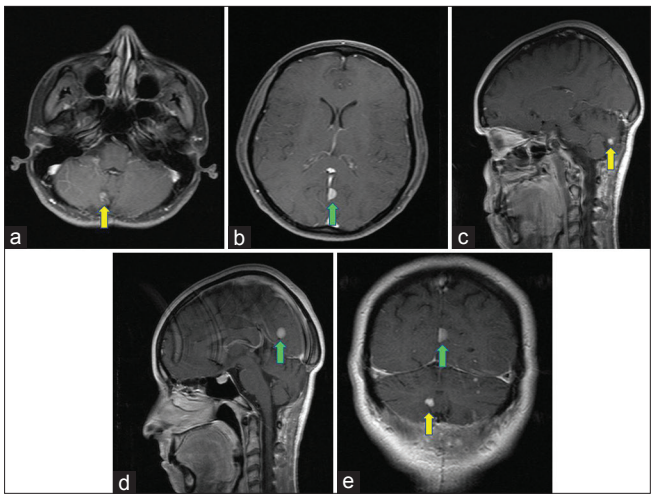

Figure 2: Two years postoperative axial (a and b), sagittal (c and d), a coronal (e) T1-weighted MR images with gadolinium detecting a solid mass with strong enhancement in the right cerebellar hemisphere (*yellow arrow*) and an hyperintense extra-axial solid mass located at posterior part of the falx (*green arrow*)

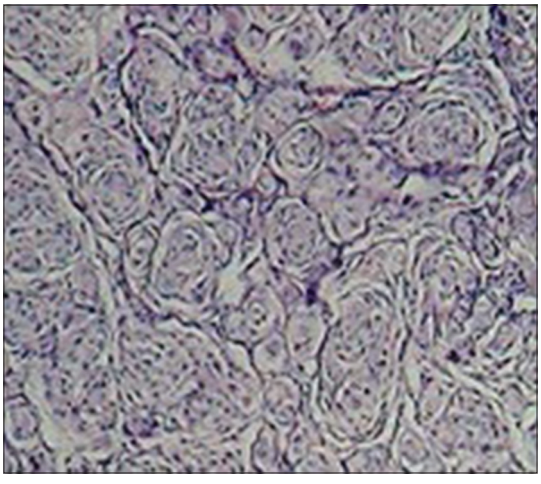

Figure 4: Histological images of meningothelial meningioma showing syncytial clusters of meningothelial cells

Table 1: Review of meningiomas in VHL

Authors	Sex/Age	Location	Histological diagnosis
Bleggi-Torres[1]	M/34	Right frontal convexity	Atypical meningioma
Governale[2]	M/37	Right frontal convexity	Fibrous meningioma
Kanno[3]	M/66	Posterior fossa	Meningothelial meningioma
Present case	F/55	Posterior part of the falx	Meningothelial meningioma

anticipation, evidence of progressively earlier age of onset and more severe presentation in successive generations have been reported.[6,16] Life expectancy in VHL remains the lowest among common inherited tumor syndromes, with a male life expectancy (59.4 years) significantly higher than female (48.4 years) and with HB or RCC as the major cause of mortality.[13] In 1995, Bleggi-Torres[2] first reported a case of meningioma in VHL, and in the pertinent literature only other two cases[4,8] 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.[7,9] The main differential diagnoses of these lesions include meningeal HB, hemangiopericytoma, and metastatic RCC. Epithelial membrane antigen (EMA) immunostaining can help distinguish supratentorial meningeal HB from meningioma.[15] 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.[5] Because of the extreme rarity of falcine 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 meningeal masses are extremely rare and no other case of falcine 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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Binderup ML, Bisgaard ML, Harbud V, Moller HU, Gimsing S, Friis-Hansen L, *et al.* Von Hippel-Lindau disease (vHL). National clinical guideline for diagnosis and surveillance in Denmark. 3rd edition. Dan Med J 2013;60:B4763.
2. Bleggi-Torres LF, De Noranha L, Fillus Neto J, Telles JE, Madalozzo LE. Von Hippel-Lindau's disease: Report of three cases and review of the literature. Arq Neuropsiquiatr 1995;53:782-8.
3. Chittiboina P, Lonser RR. Von Hippel-Lindau disease. Handb Clin Neurol 2015;132:139-56.
4. Governale LS, Vortmeyer AO, Zhuang Z, Oldfield EH. Fibrous meningioma in a patient with von Hippel-Lindau disease: A genetic analysis. J Neurosurg 2001;95:1045-9.
5. Ishwar S, Taniguchi RM, Vogel FS. Multiple supratentorial hemangioblastomas. Case study and ultrastructural characteristics. J Neurosurg 1971;35:396-405.
6. Kaelin WG, Jr. Molecular basis of the VHL hereditary cancer syndrome. Nat Rev Cancer 2002;2:673-82.
7. Kaloostian P, Taylor C. Supratentorial dural-based haemangioblastoma in a native American patient without Von Hippel Lindau Syndrome. J Surg Case Rep 2012;2012:11.
8. Kanno H, Yamamoto I, Yoshida M, Kitamura H. Meningioma showing VHL gene inactivation in a patient with von Hippel-Lindau disease. Neurology 2003;60:1197-9.
9. Kim H, Park IS, Jo KW. Meningeal supratentorial hemangioblastoma in a patient with von Hippel-Lindau disease mimicking angioblastic meningioma. J Korean Neurosurg Soc 2013;54:415-9.
10. Lamiell JM. Army clinical investigation of von Hippel-Lindau disease, 1977-2000. Mil Med 2001;166:839-42.
11. Lee KR, Kishore PR, Wulfsberg E, Kepes JJ. Supratentorial leptomeningeal hemangioblastoma. Neurology 1978;28:727-30.
12. Lonser RR, Glenn GM, Walther M, Chew EY, Libutti SK, Linehan WM, *et al.* von Hippel-Lindau disease. Lancet 2003;361:2059-67.
13. Maddock IR, Moran A, Maher ER, Teare MD, Norman A, Payne SJ, *et al.* A genetic register for von Hippel-Lindau disease. J Med Genet 1996;33:120-7.
14. Melmon KL, Rosen SW. Lindau's disease. Review of the literature and study of a large kindred. Am J Med 1964;36:595-617.
15. Murali R, Jones WI, Ma Wyatt J. A 57-year-old man with a dural-based parietal lobe tumor. Brain Pathol 2007;17:460-3, 474.
16. Ning XH, Zhang N, Li T, Wu PJ, Wang X, Li XY, *et al.* Telomere shortening is associated with genetic anticipation in Chinese Von Hippel-Lindau disease families. Cancer Res 2014;74:3802-9.
17. Takei H, Bhattacharjee MB, Rivera A, Dancer Y, Powell SZ. New immunohistochemical markers in the evaluation of central nervous system tumors: A review of 7 selected adult and pediatric brain tumors. Arch Pathol Lab Med 2007;131:234-41.