

Cerebellopontine angle hemangiopericytoma

J. Sales-Llopis; P. González-López

Neurosurgery Department, University General Hospital of Alicante, Foundation for the Promotion of Health and Biomedical Research in the Valencian Region (FISABIO), Alicante, Spain

Hemangiopericytomas (HPCs) in the [cerebellopontine angle](#) (CPA) is exceptional ^{1) 2) 3) 4) 5) 6) 7) 8)}, but given the considerable overlap of clinical, radiological and pathological features between [cerebellopontine angle meningiomas](#), [solitary fibrous tumors](#) (SFTs) and HPCs, and the very aggressive behavior of HPC compared to its counterparts; the differentiation becomes of the utmost importance.

Differential diagnosis

Since HPCs are difficult to differentiate radiographically from meningiomas, SFTs, and schwannomas; some authors such as Salunke et al. ⁹⁾ insist on the importance of preoperative planning in suspicious cases of CPA dura based masses and [cerebellopontine angle lesions](#) showing disproportionate perilesional edema, narrow base of attachment, or multilobulated cross-leaf growth.

Other authors have proposed subtle imaging characteristics that may help to distinguish HPCs from meningiomas, such as the absence of calcification or bony hyperostosis, or the comparison of the apparent diffusion coefficient values in peritumoral edema ^{10) 11)}.

[Positron emission tomography](#) has emerged as a potentially useful diagnostic tool for differentiating HPCs from meningiomas, but its high cost and availability do not permit routine use in the initial radiological investigation of CPA dura-based lesions. Consequently, the most reliable tool in HPC diagnosis remains as accurate immunohistochemical workup, and although psammoma body and nuclear pseudo inclusions can be found in meningiomas, epithelial membrane antigen negativity and [CD34/CD99](#) positivity assists in the differentiation ¹²⁾.

Solitary fibrous tumor is a rare mesenchymal tumor with a propensity to recur. The most affected area is the cerebellopontine angle. Immunohistochemistry should be used to differentiate solitary fibrous tumor from other tumors ¹³⁾.

Treatment

Its tight and strategical location make surgery in the CPA very challenging and require appropriate pre-surgical planning. This planning relies on correct pre-surgical diagnosis of the tumor as well as on means to reduce intraoperative hemorrhage. This is of even greater importance when encountering the rare hypervascular tumors of the CPA, namely hemangioblastoma, hemangiopericytoma and paraganglioma, as misdiagnosis of these tumors presurgically might have detrimental outcomes. To this end, angiography is a crucial diagnostic and therapeutic tool, helping in narrowing both the pre-

surgical differential diagnosis as well as the intraoperative bleeding. In addition, due to the high surgical risks associated with resection of vascular tumors in the CPA, non-invasive treatments, such as surgical radiosurgery (SRS), may have a crucial role ¹⁴⁾.

Case reports

2016

A 42-year-old man presented with a three-month history of progressively worsening vertigo and difficulty in walking. On admission, his neurological examination revealed a right peripheral facial palsy, right abducens palsy and left hemiparesis, suggesting the diagnosis of [Millard Gubler syndrome](#). Computed tomography and magnetic resonance imaging demonstrated a homogeneously enhancing dura based lesion of the right CPA causing major brain stem compression. There was no widening of the ipsilateral internal auditory canal. A standard retrosigmoid craniotomy was performed to access the right CPA. Exposure of the lesion revealed a well-encapsulated, gray, fibrous lesion, which appeared to originate from the tentorium. Gross total resection was achieved and confirmed radiologically. The microscopic features and the immunohistochemical profile confirmed the diagnosis of a HPC, and adjuvant radiation therapy was administered. Ten years later, the patient presented with a severe neurological deficit due to a local recurrence, but at that time refused any second intervention. He died three months later. HPC can locate within the CPA and present as a Millard-Gubler syndrome. The diagnosis should be kept in mind in case of a CPA dura based tumor. Radical surgery plus radiation therapy can maximize the recurrence-free survival and close follow-up remains mandatory to spot recurrences early ¹⁵⁾.

2015

Hemangiopericytoma of the cerebellopontine angle: case report and review of literature ¹⁶⁾.

2012

A case of cellular solitary fibrous tumor in a 22-year-old man. Neuroimaging revealed a right cerebellopontine angle tumor. Most of the tumor was cellular although some less cellular areas were seen. Sinusoidally dilated large vessels, including staghorn type, were seen. Nuclear pleomorphism and increased mitotic activity (5 mitosis/10 high power field) were regarded as evidence of anaplasia. Diffuse CD34 immunoreactivity and focal positivity for Factor XIIIa were seen in the tumor, which was negative for EMA and S100. The tumor also displayed rich reticulin network. Solitary fibrous tumor at cerebellopontine angle is rare, and 20 such cases (five reported as hemangiopericytoma) have been reported in the English literature ¹⁷⁾.

2011

Recurrent cerebellopontine angle haemangiopericytoma 5 years after stereotactic radiosurgery ¹⁸⁾.

2009

A 37-year-old Asian woman presented with a 7-month history of right ear and mandible numbness, as well as subjective hearing loss involving the right ear. Magnetic resonance imaging demonstrated the presence of a homogeneously enhancing extraaxial lesion in the right CPA, radiographically suggestive of an acoustic schwannoma. The lesion proved to be an intracranial HPC on histologic sections. Review of the neurosurgical literature yielded only one prior detailed account of HPC confined to the CPA. The patient underwent right retrosigmoid craniotomy for gross total resection of the mass, followed by stereotactic radiotherapy several weeks postoperatively.

Given the fundamentally different treatment approach for HPCs over other more common CPA tumors, it is imperative that the treating surgeon consider this rare diagnosis when evaluating patients with lesions localized to this area. Specifically, gross total resection, followed by adjuvant SRT, provides patients with the highest probability for disease-free survival, based on current evidence in the neurosurgical literature ¹⁹⁾.

1995

A case that during its 9-year-long history has repeatedly been misdiagnosed due to the misleading clinical and histopathological findings. The patient has been treated, in chronological order, for cerebrovascular disease, acoustic Schwannoma, glomus tympanicum tumor (chemodectoma) and finally turned out to have an intracranial hemangiopericytoma that originated from the area of the glomus tympanicum and eventually widely metastasized within and outside the intracranial compartment. The proper diagnosis was reached with the help of detailed immunohistochemical analysis. The subunit A of Factor XIII (FXIIIa) can be demonstrated on formaldehyde-fixed paraffin embedded sections. It has recently been shown that FXIIIa reactivity is characteristic and hence diagnostic of a subpopulation of cells within systemic and intracranial (central) hemangiopericytomas (HPCs). Since it is consistently missing from all cell components of ordinary meningiomas, glomus tumors (chemodectomas) and a host of other soft tissue tumors, its presence or absence is a helpful sign in various differential diagnostic dilemmas ²⁰⁾.

¹⁾ ²⁰⁾

Molnar P, Nemes Z. Hemangiopericytoma of the cerebello-pontine angle. Diagnostic pitfalls and the diagnostic value of the subunit A of factor XIII as a tumor marker. Clin Neuropathol. 1995 Jan-Feb;14(1):19-24. PubMed PMID: 7729075.

²⁾

Mallucci CL, Ward V, Carney AS, O'Donoghue GM, Robertson I. Clinical features and outcomes in patients with non-acoustic cerebellopontine angle tumours. J Neurol Neurosurg Psychiatry. 1999 Jun;66(6):768-71. PubMed PMID: 10329752; PubMed Central PMCID: PMC1736400.

³⁾

Alén JF, Lobato RD, Gómez PA, Boto GR, Lagares A, Ramos A, Ricoy JR. Intracranial hemangiopericytoma: study of 12 cases. Acta Neurochir (Wien). 2001;143(6):575-86. PubMed PMID: 11534674.

⁴⁾ ¹²⁾ ¹⁹⁾

Tashjian VS, Khanlou N, Vinters HV, Canalis RF, Becker DP. Hemangiopericytoma of the cerebellopontine angle: a case report and review of the literature. Surg Neurol. 2009 Sep;72(3):290-5. doi: 10.1016/j.surneu.2008.06.016. Review. PubMed PMID: 18786704.

⁵⁾ ¹⁸⁾

Cho JM, Kim SH, Kim SH, Lee KS, Chang JH. Recurred cerebellopontine angle haemangiopericytoma 5

years after stereotactic radiosurgery. Clin Neurol Neurosurg. 2011 Dec;113(10):931-3. doi: 10.1016/j.clineuro.2011.06.013. PubMed PMID: 21802837.

6) 17)

Zeng J, Ogera P, Benardete EA, Nicastri AD, Rao C. Cellular solitary fibrous tumor (hemangiopericytoma) with anaplasia at cerebellopontine angle—a case report. Pathol Res Pract. 2012 Aug 15;208(8):493-6. doi: 10.1016/j.prp.2012.04.008. PubMed PMID: 22784469.

7) 9) 10)

Salunke P, Futane S, Gupta K, Vasishta RK. Cerebello-pontine angle hemangiopericytoma: an orphan differential diagnosis. Clin Neurol Neurosurg. 2013 Jul;115(7):1184-6. doi: 10.1016/j.clineuro.2012.10.020. PubMed PMID: 23182178.

8)

Teoh JW, Goh BS, Shahizon Azura MM, Siti Aishah MA, Nor Hafliza MS. An unexpected lesion in cerebellopontine angle: hemangiopericytoma. Med J Malaysia. 2014 Jun;69(3):146-7. PubMed PMID: 25326360.

11)

Liu L, Yin B, Geng DY, Li Y, Zhang BY, Peng WJ. Comparison of ADC values of intracranial hemangiopericytomas and angiomatous and anaplastic meningiomas. J Neuroradiol. 2014 Jul;41(3):188-94. doi: 10.1016/j.neurad.2013.07.002. PubMed PMID: 24524869.

13)

Chen H, Zeng XW, Wu JS, Dou YF, Wang Y, Zhong P, Xu R, Jiang CC, Wang XQ. Solitary fibrous tumor of the central nervous system: a clinicopathologic study of 24 cases. Acta Neurochir (Wien). 2012 Feb;154(2):237-48; discussion 248. doi: 10.1007/s00701-011-1160-9. PubMed PMID: 21938461.

14)

Laviv Y, Thomas A, Kasper EM. Hypervascular lesions of the cerebellopontine angle: the relevance of angiography as a diagnostic and therapeutic tool and the role of stereotactic radiosurgery in management. A comprehensive review. World Neurosurg. 2016 Dec 31. pii: S1878-8750(16)31406-1. doi: 10.1016/j.wneu.2016.12.091. [Epub ahead of print] Review. PubMed PMID: 28049034.

15)

Ben Nsir A, Badri M, Kassas AZ, Hammouda KB, Jemel H. Hemangiopericytoma of the Cerebellopontine Angle: A Wolf in Sheep's Clothing. Brain Tumor Res Treat. 2016 Apr;4(1):8-12. doi: 10.14791/btrt.2016.4.1.8. PubMed PMID: 27195255; PubMed Central PMCID: PMC4868819.

16)

Najib J, Aniba K, Laghmari M, Lmejjati M, Ghannane H, Benali SA, Ennadam H, Jalal H, idrissi C. [Hemangiopericytoma of the cerebellopontine angle: case report and review of literature]. Pan Afr Med J. 2015 Jan 22;20:61. doi: 10.11604/pamj.2015.20.61.4198. Review. French. PubMed PMID: 26090019; PubMed Central PMCID: PMC4450031.

From:

<https://neurosurgerywiki.com/wiki/> - **Neurosurgery Wiki**

Permanent link:

https://neurosurgerywiki.com/wiki/doku.php?id=cerebellopontine_angle_hemangiopericytoma

Last update: **2024/06/07 02:58**

