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Meningeal Hemangiopericytoma: A Rare Intracranial Tumor—About a Case

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Abstract

Introduction: Hemangiopericytoma is a very rare vascular tumor, difficult to diagnose, often confused with a meningioma. Imaging is not very specific and the diagnostic certainty remains histological. **Observation:** We report a case of meningeal hemangiopericytoma in a 48-year-old patient with a history of active smoking and whose sister died of glioblastoma. He was brought to the emergency room for a fall, confusion, and tonic-clonic seizures. The diagnosis was suspected on imaging (CT and MRI, in front of a syndrome of right temporal intraventricular mass measured at 37 mm in diameter, centered on the choroid plexus, with perilesional vasogenic edema and posterior white commissure. There was homogeneous enhancement of the lesion after gadolinium injection. The diagnosis was confirmed by immunohistochemical study, which showed diffuse expression of STAT6 and partial labeling for CD34, without significant expression of SSTR2. Expression of neurofilament protein was also found in the brain parenchyma. **Conclusion:** Hemangiopericytoma is a rare tumor, characterized by its malignant potential, its high rate of recurrence and distant metastasis. The management is based on a total microsurgical resection followed by radiotherapy.

Keywords

Intracranial Meningeal Hemangiopericytoma, Surgery, Metastasis, Radiotherapy

1. Introduction

Hemangiopericytoma (HP) is a rare mesenchymal tumor that develops from Zimmermann's pericytes around capillaries and venules and can, therefore, occur

anywhere in the body, especially the retroperitoneum and musculoskeletal system. It represents less than 1% of intracerebral tumors. Its histogenesis was for a long time the subject of controversy, and it was only in 1993, based on ultrastructural and immunohistochemical findings, that hemangiopericytoma was classified by the WHO as a "non-meningothelial malignant mesenchymal tumor" [1]. Its appearance on computed tomography (CT) or magnetic resonance imaging (MRI) can be misleading and may lead to a false diagnosis of meningioma. Hemangiopericytoma is characterized by its malignant potential and its high rate of recurrence and distant metastasis, justifying a large surgical excision and complementary radiotherapy [2].

We report a case of intracranial HP with review of the literature.

2. Patient and Observation

The patient was 48 years old, with a history of active smoking and a sister who died of a glioblastoma. He was brought to the emergency room for a fall, confusion, and tonic-clonic convulsions. The physical examination revealed a confused patient with a Glasgow score of 14, intermediate and reactive pupils, osteo-tendinous reflexes difficult to find. There was no Babinski. Heart sounds were regular without murmurs heard, vesicular murmurs were bilateral and symmetrical without noises, calves were soft and not painful. The abdomen was soft, depressed, with no palpable mass. Vitals showed a temperature of 36.5˚C, blood pressure 147/99 mmhg, pulse 68 bpm, SpO_2 92%. Biological tests showed D-dimer levels of 1840, CRP cold, troponin negative. Blood count and renal function were normal. Liver and pancreatic function tests were normal.

Cardiac ultrasound was unremarkable. The cerebral computed tomography (CT) showed a supra tentorial and/or intra axial tumor mass syndrome measuring 45 mm in diameter centered on the right ventricular junction, pushing forward the choroid plexus, with a centimetric calcium contingent at its superior pole. Exclusion hydrocephalus by trapping of the temporal horn of the right lateral ventricle, adjacent diffuse peri-lesional edema radiating in a right parieto-temporal pocket, right-left transfalcoral involvement with midline deviation evaluated at 12 mm. There was also a right frontal subcutaneous hematoma and normal trophicity for age (**Figure 1**). There was no medial temporal involvement, no intra- or peri-cerebral hematoma, and no meningeal hemorrhage. On brain MRI, there was a right temporal intraventricular nodular lesion measured at 37 mm in diameter, centered on the choroid plexus, perilesional vasogenic edema and posterior white commissure. There was homogeneous lesion enhancement after gadolinium injection. There was no pathologic contrast of the supra- or infra-tentorial brain parenchyma (**Figure 2**).

In view of these radiological findings, the diagnosis of choroid plexus papilloma or intraventricular meningioma was evoked.

The histological study showed a lesion of diffuse architecture, more or less fasciculated, formed by tumor cells of medium size, oval, slightly anisokaryotic,

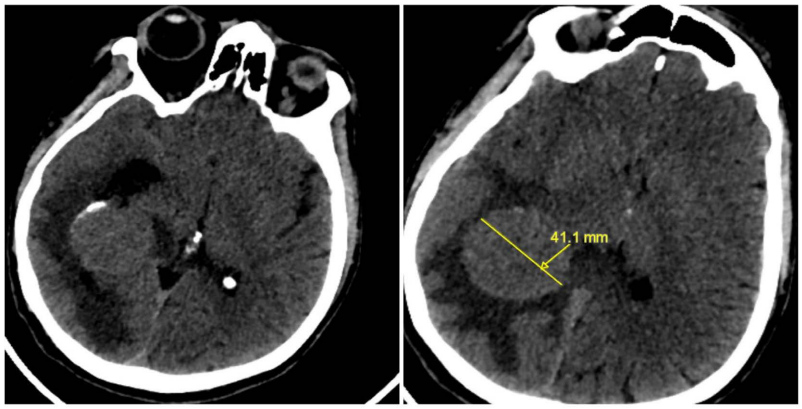

Figure 1. Axial slice brain CT of our patient, showing a supra tentorial tumor mass syndrome measuring 45 mm in diameter centered on the right ventricular junction, with centimetric calcium contingent at its superior pole. Exclusion hydrocephalus by trapping of the temporal horn of the right lateral ventricle, adjacent diffuse peri-lesional edema radiating in right parieto-temporal immersion sleeve, right-left transfalcoral involvement with midline deviation evaluated at 12 mm.

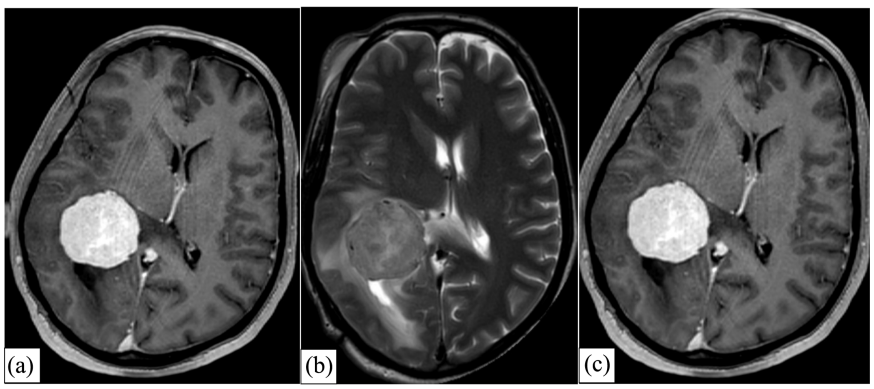

Figure 2. Axial section brain MRI of our patient, showing a right temporal intraventricular nodular lesion measured at 37 mm in diameter, centered on the choroid plexus, perilesional vasogenic edema and of the posterior white commissure. There was homogeneous lesion enhancement after gadolinium injection. There was no pathologic contrast of the brain parenchyma. (a): T1 SE, (b): T2 SE, (c): T1 + Gado.

with pale eosinophilic cytoplasm, containing some collagenous fibers and an abundant vascular network, with gaping lumens and thrombosed walls. The lesion is the site of hemorrhagic changes and presents some aspects of coagulation necrosis, evoking an ischemic necrosis, without any obvious tumor necrosis area.

The mitotic index is evaluated at one mitosis per 10 fields at high magnification (1.6 mm^2).

The brain parenchyma was infiltrated by the lesion, but the interface between the tumor and the parenchyma remained clear and fairly well limited.

The immunohistochemical study showed diffuse expression of STAT6 and partial marking for CD34, without significant expression of SSTR2. Neurofilament proteins were expressed in the brain parenchyma.

Together, these findings led to the diagnosis of grade II meningeal hemangi-

opericytoma according to the WHO 2016 classification and grade I according to the Marseille classification.

In view of the mass effect, the patient was offered rapid surgical management. The procedure, the after-effects and complications as well as the therapeutic alternatives were explained to the patient who consented to the procedure.

During the operation, a complete macroscopic excision of the lesion was performed, with coagulation and securing of the diseased choroid plexuses which were removed. The patient was discharged from surgery without sequelae with disappearance of the intracranial hypertension syndrome. He did not have any more epileptic seizures, he was put on KEPPRA 500 mg morning and evening, there was no corticotherapy. The case was discussed in a multidisciplinary meeting with a proposal for additional treatment by adjuvant radiotherapy. A dosimetric scan was performed and merged with an MRI to adapt the volume to be treated.

The follow-up at three years was favorable, the successive imaging examinations of control at 3 years of the intervention did not show any tumor recurrence.

The patient's informed consent was obtained for the publication of this case.

3. Discussion

Hemangiopericytoma is a rare vascular tumor that represents less than 1% of all intracranial tumors and 2% to 4% of meningeal tumors [3] [4] [5]. Unlike meningiomas, hemangiopericytoma has a predilection for the male gender, occurs in 56% - 75% between 38 and 42 years of age, and most often affects the supratentorial stage [5] [6] [7], which was the case of our patient. The distribution of hemangiopericytomas is similar to that of meningiomas [8]: they occur mainly in the supratentorial region, 77% in the series reported by Jeong-Hoon *et al.* [7], 66% in that of Dufour *et al.* [6].

Initially classified as subtypes of meningioma by Bayley *et al.* [9], called "angioblastic meningiomas" or "hemangiopericytic meningiomas". This classification was controversial because of the different origin of these two tumors: HP originate from the pericytes of the capillaries surrounding the meninges, whereas meningiomas develop from arachnoid cells [10]. It was not until 1942 that the term HP was introduced by Stout and Murray [11], who published the first nine cases of HP in the literature. Since 1993, the World Health Organization (WHO) classifies hemangiopericytoma in the group of "non-meningothelial malignant mesenchymal tumors". Clinically, the initial symptomatology varies depending on the location and size of the tumor. Supratentorial involvement is mainly manifested by headache, and less frequently by sensory-motor deficit, visual disturbances, or comital seizures, which was the case in our patient. Tumors of the posterior fossa are mainly responsible for balance disorders, which was the case in our patient. Parasellar involvement is evoked in front of a syndrome of the cavernous cavity [10] [12]. Cerebral imaging (CT or MRI) allows to describe the aspect and the characteristics of the tumor evoking a HP without

however being able to eliminate the differential diagnosis of meningioma. Angiography seems to be interesting because of the hypervascular nature of HP [13]. Immunohistochemistry confirms the diagnosis. HP expresses only mesenchymal markers. Tumor cells are labeled with antibodies to CD 34, but are negative for antibodies to factor VIII and protein S-100 [14].

Treatment is based on surgical resection followed by radiation therapy. Surgical resection should be as large as possible. However, the highly hemorrhagic nature of the tumor may pose major problems of hemostasis and make the exeresis delicate. In order to reduce tumor vascularization, preoperative embolization is sometimes performed with variable results depending on the series [12] [15].

The role of radiotherapy is currently unanimously accepted: it prolongs the remission period, reduces the rate of recurrence and consequently increases the survival time [1]. The response to radiotherapy is dose-dependent: only doses higher than 50 Gy are associated with a statistically significant reduction in the local recurrence rate [1] [11]. This radiotherapy should be started immediately, even if the tumor removal is considered complete. Jeong H *et al.* [16] report 5-year remission rates of 100% in patients who had radiotherapy after surgery and 70.3% in patients who had surgery alone. For Guthrie *et al.* [12], radiotherapy after the first operation prolongs the period of remission from 34 to 75 months and prolongs survival from 62 to 92 months. Dufour *et al.* [10] report recurrence rates of 12% and zero mortality in the group of irradiated patients, whereas in the group of non-irradiated patients the recurrence and mortality rates were 88% and 55% respectively. The evolution of HP is marked by the frequency of local and distant recurrences. The rate of local recurrence varies according to the series from 26% to 86% and depends mainly on the quality of the exeresis and the practice of postoperative radiotherapy [10] [12] [16].

Distant extension can occur along the neuraxis, by fluid or by hematogenous route. The most frequent metastatic sites are bone, lung and liver. The probability of metastasis increases with time, 13% at 5 years, 33% at 10 years, and 64% at 15 years [10] [16].

4. Conclusion

Meningeal hemangiopericytoma is a rare tumor, often mistaken for a meningioma. However, it is important to evoke the diagnosis preoperatively, given the risks of bleeding during surgery. In the absence of specific tumor characteristics, radiological diagnosis is difficult. The diagnostic certainty remains histological, mainly based on immunohistochemical study. Treatment is based on complete excision and radiotherapy at an effective dose, which significantly reduces the recurrence rate or even leads to a complete cure, as in the case of our patient. A prolonged follow-up of these patients is necessary, considering the frequency of recurrence and the sometimes late occurrence of metastases.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

References

[1] Ksira, I., Berhouma, M., Jemel, H., Khouja, N. and Khaldi, M. (2006) Hémangiopéricytome sellaire: À propos d'un cas. *Neurochirurgie*, **52**, 123-127. https://doi.org/10.1016/S0028-3770(06)71207-3

[2] Errachdi, A., Asabbane, A., Epala, B.N., Hemmich, M., Kabbali, N., Diakité, A., *et al.* (2014) L'hémangiopéricytome méningé: Une tumeur intracrânienne rare—à propos d'un cas. *The Pan African Medical Journal*, **17**, Article No. 223. https://doi.org/10.11604/pamj.2014.17.223.2920

[3] Alen, J.F., Lobato, R.D., Gomez, P.A., Boto, G.R., Lagares, A., Ramos, A. and Ricoy, J.R. (2001) Intracranial Hemangiopericytoma: Study of 12 Cases. *Acta Neurochirurgica*, **143**, 575-586. https://doi.org/10.1007/s007010170062

[4] Ecker, R.D., Marsh, W.R., Pollock, B.E., Kurtkayayapicier, O., Mcclelland, R., Schetthauer, B.W., *et al.* (2003) Hemangiopericytoma in the Central Nervous System: Treatment, Pathological Features, and Long-Term Followup in 38 Patients. *Journal of Neurosurgery*, **98**, 1182-1187. https://doi.org/10.3171/jns.2003.98.6.1182

[5] Jalali, R., Srinivas, C., Nadkarni, T.D. and Rajasekharan, P. (2008) Suprasellar Haemangiopericytoma-Challenges in Diagnosis and Treatment. *Acta Neurochirurgica*, **150**, 67-67. https://doi.org/10.1007/s00701-007-1474-9

[6] Dufour, H., Metellus, P., Fuentes, S., Murracciole, X., Regis, J., Figarella-Branger, D., *et al.* (2001) Meningeal Hemangiopericytoma. A Retrospective Study of 21 Patients with Special Review of Postoperative External Radiotherapy. *Neurosurgery*, **48**, 756-763.

[7] Kim, J.H., Jung, H.W., Kim, Y.S., Kim, C.J., Hwang, S.K., Paek, S.H., *et al.* (2003) Meningeal Hemangiopericytomas: Long-Term Outcome and Biological Behaviour. *Surgical Neurology*, **59**, 47-54. https://doi.org/10.1016/S0090-3019(02)00917-5

[8] Garbi, A., Ousehal, A., Kissani, N., Lakhdar, A., Azhari, A. and Kadiri, R. (2001) Hémangiopéricytome sellaire: A propos d'un cas. *Journal of Neuroradiology*, **28**, 195-199.

[9] Bayley, P., Cushing, H. and Eisenhardt, L. (1928) Angioblastic Meningiomas. *Archives of Pathology*, **6**, 953-990.

[10] Dufour, H., Bouillot, P., Figarella-branger, D., Nodoye, N., Regis, J., Njeebugha, T. and Grisoli, F. (1998) Hémangiopéricytomes Méningés: Revue rétrospective de 20 cas. *Neurochirurgie*, **44**, 5-18.

[11] Stout, A.P. and Murray, M.R. (1942) Hemangopericytoma: A Vascular Tumor Featuring Zimmerman's Pericytes. *Annals of Surgery*, **116**, 26-33. https://doi.org/10.1097/00000658-194207000-00004

[12] Guthrie, B.L., Ebersold, M.J., Scheithauser, B.W. and Shaw, E.G. (1989) Meningeal Hemangiopericytoma: Histopathological Features, Treatment, and Long-Term Follow-up of 44 Cases. *Neurosurgery*, **25**, 514-522.

[13] Akiyama, M., Sakai, H., Onoue, H., Miyazaki, Y. and Abe, T. (2004) Imaging Intracranial Haemangiopericytomas: Study of Seven Cases. *Neuroradiology*, **46**, 194-197.

[14] Bouvier-Labit, C., Liprandi, A., Piercecchi, M.D., HosseiniH, Henin, D. and Figarella-Branger, D. (2000) Apport de l'immunohistochimie et de la microscopie électro-

nique dans le diagnostic des hémangiopéricytomes méningés: À propos de 15 cas. *Annales de Pathologie*, **20**, 492-498.

[15] Sheehan, J., Kondziolka, D., Flickinger, J. and Lunsford, L.D. (2002) Radiosurgery for Treatment of Recurrent Intracranial Hemangiopericytomas. *Neurosurgery*, **51**, 905-910.

[16] Kim, J.H., Jung, H.W., Kim, Y.S., Kim, C.J., Hwang, S.K., Paek, S.H., Kim, D.G. and DukKwun, B. (2003) Meningeal Hemangiopericytomas: Long-Term Outcome and Biological Behaviour. *Surgical Neurology*, **59**, 47-53. https://doi.org/10.1016/S0090-3019(02)00917-5