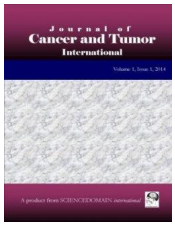

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Intracranial Hemangiopericytoma in a 15 Year-old Child: Neuroimaging Features and Management Therapy

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Intracranial hemangiopericytoma (HPR) is a highly vascular tumors which is exceptional in children. They may present management difficulties in some cases.
Clinical Presentation: The authors report the case of a 15 year-old female presented with symptoms of increased intracranial pressure. Physical examination featured a frontal syndrome, a Broca's aphasia, a grade 3 papilledema and a decreased visual acuity in her left eye. Radiological investigation including cranial CT scan and MRI displayed a large dural-based heterogeneous hemorrhagic tumor of the left frontal convexity. The patient underwent a total-gross resection of an extra-parenchymal hemorrhagic tumor through a large frontal craniotomy with no postoperative

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complications. Pathologic examination of the specimen and immuno-histochemistry concluded to an anaplastic HPR. Postoperative radiotherapy was performed without incidents. A local recurrence was diagnosed four months postoperatively and remained stable after additional radiotherapy.

Conclusion: Intracranial anaplastic HPR in children are rare. Imaging features are not specific and may mimics meningioma. Microneurosurgery with adjuvant radiotherapy is required in the management of these aggressive tumors.

Keywords: Hemangiopericytoma; child; surgery; radiotherapy; intracranial; MRI.

1. INTRODUCTION

Meningeal HPR is a highly cellular mesenchymal tumor which arises from pericytes and accounts for less than 1% of primary central nervous system tumors [1]. Intracranial HPR is uncommon in children, only a few cases have been reported in the litterature [2-8]. This tumor has a more aggressive course than meningiomas with variable clinical behavior and prognosis, depending on patients' age [9]. Management requires a multidisciplinary approach including surgery with often preoperative embolisation and complementary chemotherapy or radiotherapy. We present the case of an intracranial anaplastic HPR managed in our department emphasizing on its radiological and anatomopathological features.

2. CASE PRESENTATION

A 15 year-old female, was referred with a 2-month history of symptoms of increased intracranial pressure. Neurological examination featured a conscious right handed patient who was apathetic with reduced verbal fluency, a frontal syndrome, Broca's aphasia and right lively reflexes without motor deficit. Ophthalmologic examination showed bilateral grade 3 papilledema and decreased visual acuteness (5/10) in the left eye. No other abnormalities were present on clinical examination.

Computerized tomography (CT) scan showed a large multicystic mass, which measures 12 x 8 cm, with peripheral enhancement located in the left frontal parasagittal convexity (Fig. 1A). The mass showed mixed low and isoattenuation with moderate surrounding edema. Notable mass effect was associated, with left-to-right midline shift, subfalcine and uncal herniations.

On Magnetic resonance image (MRI), the mass appeared heterogeneous on T1 and T2 weighted-images (WI), with solid and cystic components. It showed a dural-base on the left-frontal convexity. The largest cystic component appeared hyperintense on T1-WI, hypointense on T2-WI in relation to hemorrhagic content. The solid component presented a narrow-based dural attachement and a "dural tail" sign. It appeared isointense to grey matter on T1 and T2-WI, enhanced markedly and heterogeneously after gadolinium administration and contained multiple signal intensity flow voids (Figs. 1B & 1C).

On diffusion-WI, the solid portion appeared hyperintense with a corresponding hypointensity on the ADC map; diffusion was restricted compared to normal white matter (Figs. 1D & E).

Perfusion-WI showed a fivefold increase in relative regional cerebral blood volume in the solid component (Fig. 1F). MR spectroscopic imaging was not performed because of the presence of the large hemorrhagic component.

The patient underwent a total-gross resection of the tumor through a large frontal craniotomy. The lesion was, indeed, extra-parenchymal and hemorrhagic with modest blood spoliation. Dura was thick, vascularized and invaded by the tumor. Arachnoid sheet was present in places between the tumor and the parenchyma. Duraplasty using the galea aponeurotica was performed at the end of the procedure. The postoperative course was uneventful and the patient was discharged on the 5th postoperative day.

Pathologic gross examination of a piecemeal lesion displayed elastic whitish portions with reddish hemorrhagic foci. The smears featured a very cellular tumor, with round to oval cells. Cells were arranged around thin-walled vascular spaces lined by a non-neoplastic endothelium. They were large, with finely granular cytoplasms and round-shaped hyperchromatic nuclei. Mitotic atypia with a high mitotic index, vascular thrombosis, hemorrhage, hemosiderin deposits and necrosis areas were noted.

Immuno-histochemistry showed diffuse positivity for vimentin, CD99, and bcl-2 and variable positivity for CD34 (Fig. 2). The diagnosis of anaplasic hemangiopericytoma was made.

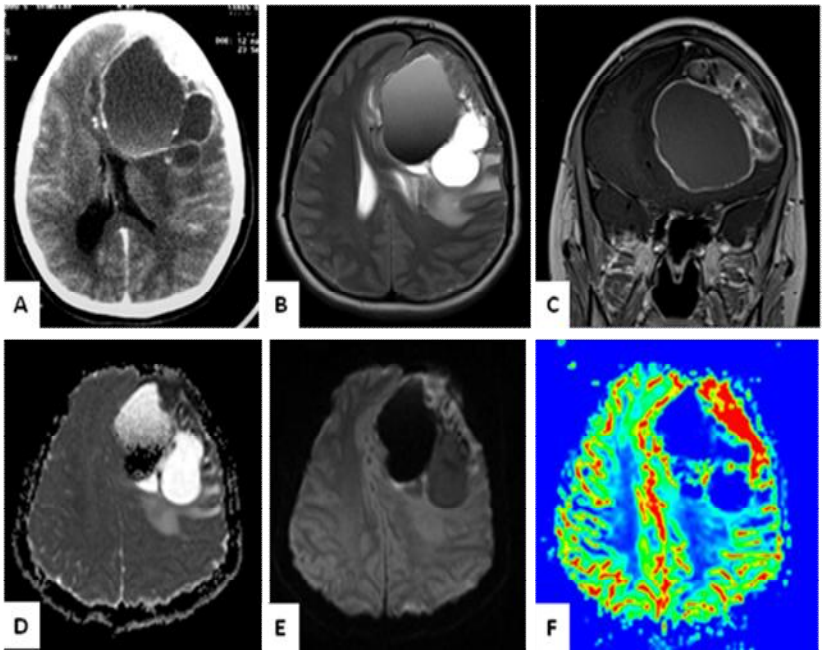

Fig. 1. A, cranial axial computed tomography with contrast enhancement shows a left frontal large multicystic mass with peripheral enhancement. B, A, axial T2-WI displays an heterogeneous signal of this lesion. The largest cystic component appeared hypointense whereas the solid component appeared isointense to grey matter. C, post-enhanced coronal T1-WI revealing an intense and heterogeneous contrast enhancement of the solid component of this intracranial mass. D & E, diffusion-WI shows a hyperintense signal with a corresponding hypointensity on the ADC of the solid portion. F, perfusion-WI showed an increase in relative regional cerebral blood volume in this solid component

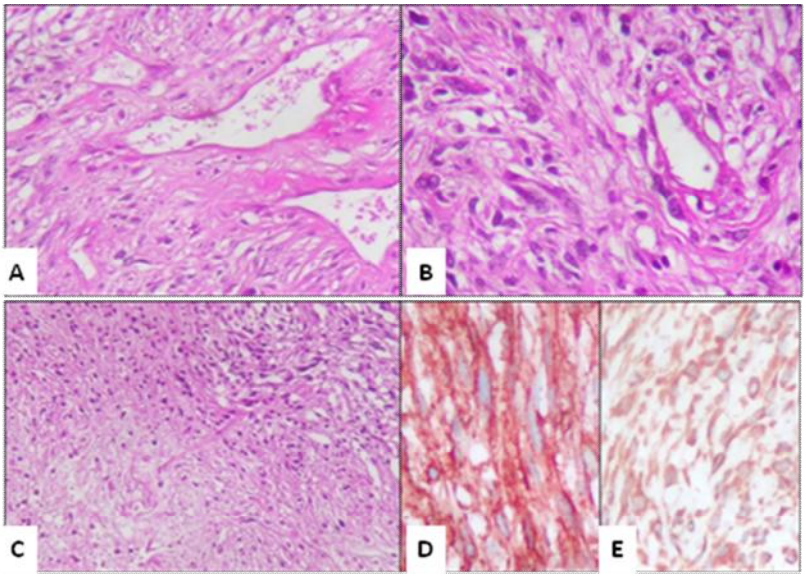

Fig. 2. Histopathological and immunohistochemical evidence displaying a highly cellular and vascularized mesenchymal tumor with dilated, staghorn-type vessels (A), High nuclear atypia (B) and necrosis (C). Ki67 index was 25%. The tumor cells were immune-positive for vimentin (D) and CD 34 (E)

One month postoperatively, the child started whole-brain radiotherapy. She received 54 Gy without incidents. On the fifth postoperative month, physical examination showed improvement of Broca's aphasia but persistent decreased visual acuity in her left eye. However, MRI revealed a local recurrence (Fig. 3B). Therefore, an additional radiation dose of 10 Gy was delivered. After 1-year follow-up, the recurrence remained unchanged on MRI (Fig. 3C) and No metastases were found on body scan.

3. DISCUSSION

HPRs were first described by Stout and Murrey in 1942 [3]. Intially, they were considered variants of angioblastic meningiomas, hemangiopericytic type. The 2007 WHO classification divided HPRs into two categories: grade II HPR, the more common type, and grade III HPR or anaplastic HPR [10].

HPRs are quite uncommon in all age groups, representing less than 1% of all CNS tumors, with only 10% of cases occurring in children [1]. They represent less than 3% of dural-based lesions [1]. The peak age ranges within the third and fourth decades [10] with a slight male predominance. HPR is extremely rare among children and most of them occur within the first year of life [3-6, 11]. This infantile form of HPR is associated with a more benign course. The behavior of HPR in children older than 1 year is similar to that of adult HPR [9].

Intracranial HPRs are almost invariably solitary, attached to the cranial dura and to the venous sinuses, rarely the skull base dura. HPR may develop into the parenchyma, the ventricles, the sella, the pineal gland or the orbit [12-16]. The spectrum of clinical signs and symptoms mirrors that described for meningiomas.

Neuro-imaging features of intracranial HPRs are non-specific but show differences with regard to meningiomas. Indeed, intracranial HPRs are multilobulated, and their location is similar to that of meningiomas. However, HPRs present a narrow base of dural attachment in one third of cases, which is not typically seen in meningiomas [17]. They usually are large, even in children, with a mean diameter of 75 mm [18]. On CT-scan, HPRs are heterogeneous, hyperdense and typically show heterogeneous enhancement. They also show bony erosion, frequently seen in most anaplastic form but they lack calcifications and hyperostosis, usually observed in meningiomas [17,19,20]. MRI findings show that intracranial HPR are heterogeneous, isointense on T1 and T2-WI with signal flow voids and enhance heterogeneously [17,19,20]. In some cases, the serpentine signal intensity voids in vessels are numerous and large, giving an AVM-like appearance on MRI, which is suggestive of HPR [18]. According to Zhou et al. [21], pronounced lobulation, necrosis, cystic changes and damage of adjacent skull on MRI findings are more commen in anaplastic form of HPR. On magnetic resonance spectroscopy, Barba et al. noted that elevated myoinositol using short TE (20 msec) may help differentiate HPR from meningioma [22].

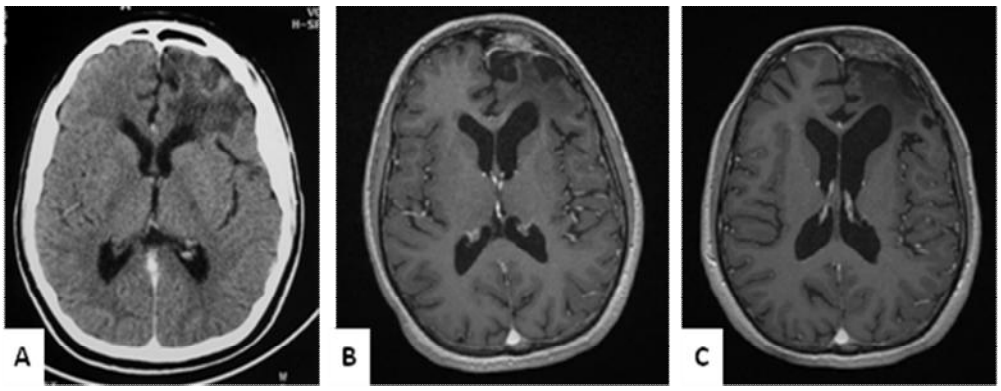

Fig. 3. A, cranial axial CT scan with contrast enhancement on the first postoperative month shows a total goss resction of the HPR. B, post-enhanced axial T1-WI revealing a local recurrence of the tumor 3 months after radiotherapy. C, after an additional radiation dose of 10 Gy The recurrence remained unchanged on the 1-year follow-up post-enhanced axial T1-WI

Tumors showing increased mitotic activity and/or necrosis as well as hemorrhage have been defined as anaplastic grade III HPR [10,11] and have been reported to be associated with malignant behavior [23]. Histological distinction between the benign and the malignant forms of HPR in children cannot be made [11]. Meningeal HPR shows diffuse positivity for vimentin, CD99, and bcl-2 [24] and more variable positivity for CD34 (33–100%) which is generally patchy as opposed to the diffuse positivity typical of solitary fibrous tumor [25].

Treatment strategies for the management of intracranial HPR in children remain non-uniform. Surgical resection is the treatment of choice for intracranial HPR [26]. However, it has been proven very difficult to cure the disease with surgical resection alone [27,28].

Gross-total resection at first operation was strongly correlated with prolongation of time to recurrence and extension of survival duration [29,30]. HPR are hypervascular tumors causing intra-operative hemorrhage [27,31], as in our case. Surgical mortality rate is about 9–24%. Hence, some authors recommend prior embolization [32].

The effect of adjuvant radiotherapy in the treatment of HPR is discussed. Several studies have shown that postoperative radiotherapy decreases the local recurrence rate [28,33,34] and improved survival outcome [34]. However, these results are not accepted by all authors. Indeed, HPR may be a relatively radioresistant lesion and the addition of postoperative radiation does not confer a survival benefit compared with gross-total resection [30].

Stereotactic radiosurgery may be an effective therapeutic mean to treat recurrent intracranial HPR in adult after surgical resection followed or not by radiotherapy [27,35]. Given the rarity of HPR in children, using stereotactic radiosurgery has not yet been worked out.

The role of chemotherapy is more debatable [9]. For adult HPR, chemotherapy efficacy is limited [36]. However, the use of chemotherapy for congenital and infantile HPR has proven to be successful in many cases combined with surgery [7,8,11].

Local aggressiveness, high rate of recurrence and extra-neural metastasis are the characteristics of intracranial HPR [8]. Anaplastic HPR recurs locally earlier than low-grade HPR. Consequently, clinical control and annual MRI follow up are necessary.

4. CONCLUSION

Management of intracranial HPR in children older than 1 year does not differ from adult HPR. Imaging diagnosis of HPR is not always straightforward, because it mimics meningioma. Anaplastic HPR are recognized for their aggressive clinical behavior, high recurrence rates, Microneurosurgery with adjuvant radiotherapy is the main treatment. Long-term follow-up is mandatory because this tumor had tendency for recurrence and metastasis even after complete macroscopic resection.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Alen JF, Lobato RD, Gomez PA, Boto GR, Lagares A, Ramos A, et al. Intracranial hemangiopericytoma: study of 12 cases. Acta Neurochir (Wien). 2001;143(6):575-86.
2. Huisman TAGM, Brandner S, Niggli F, Kacl G, Willi UV, Martin E. Meningeal hemangiopericytoma in childhood. Eur Radiol. 2000;10:1073-5.
3. Cole JC, Naul LG. Intracranial infantile hemangiopericytoma. Pediatr Radiol. 2000;30(4):271-3.
4. Herzog CE, Leeds NE, Bruner JM, Baumgartner JE. Intracranial hemangiopericytomas in children. Pediatr Neurosurg. 1995;22(5):274-9.
5. Aouad N, Vital C, Rivel J, Ramsoubramanian K, Santosh S, Chwdry O. Giant supratentoriel meningeal hemangiopericytoma in a new born. Acta Neurochir (Wien). 1991;112(3-4):154-6.
6. Laviv Y, Michowitz S, Schwartz M. Neonatal intracranial hemangiopericytoma:

a 7-year follow-up. Acta Neurochir (Wien). 2011;154:637-8.
7. Sulit DJ, Tom WL, Willert JR, Nespeca MP, Friedlander SF. Multicentric infantile hemangiopericytoma: case report and review of the literature. J Pediatr Hematol Oncol. 2011;33(4):e166-70.
8. Fernandez-Pineda I, Parida L, Jenkins JJ, Davidoff AM, Rao BN, Rodriguez-Galindo C. Childhood hemangiopericytoma: review of St Jude Children's Research Hospital. J Pediatr Hematol Oncol. 2011;33(5):356-9.
9. Rodriguez-Galindo C, Ramsey K, Jenkins JJ, Kaste SC, Merchant TE, Rao BN, et al. hemangiopericytoma in children and infants. Cancer. 2000;88:198-204.
10. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, et al. WHO classification of tumours of the central nervous system. Acta Neuropathol. 2007;114:97-109.
11. Kerl K, Strater R, Hasselblatt M, Brentrup A, Fruhwald MC. Role of neoadjuvant chemotherapy in congenital intracranial haemangiopericytoma. Pediatr Blood Cancer. 2011;56(1):161-3.
12. Shetty PM, Moiyadi AV, Sridhar E. Primary CNS hemangiopericytoma presenting as an intraparenchymal mass-case report and review of literature. Clin Neurol Neurosurg. 2010;112(3):261-4.
13. Sumi K, Watanabe T, Ohta T, Fukushima T, Kano T, Yoshino A, et al. Hemangiopericytoma arising in the body of the lateral ventricle. Acta Neurochir (Wien). 2010;152(1):145-9.
14. Jalali R, Srinivas C, Nadkarni TD, Rajasekharan P. Suprasellar haemangiopericytoma-challenges in diagnosis and treatment. Acta Neurochir (Wien). 2008;150(1):67-71.
15. Jian BJ, Han SJ, Yang I, Waldron JS, Tihan T, Parsa AT. Surgical resection and adjuvant radiotherapy for a large pineal hemangiopericytoma. J Clin Neurosci. 2010;17(9):1209-11.
16. Shinder R, Jackson TL, Araujo D, Prieto VG, Guadagnolo BA, Esmaeli B. Preoperative radiation therapy in the management of recurrent orbital hemangiopericytoma. Ophthal Plast Reconstr Surg. 2011;27(5):e126-8.
17. Chiechi MV, Smirniotopoulos JG, Mena H. Intracranial hemangiopericytomas: MR and CT features. AJNR Am J Neuroradiol. 1996;17(7):1365-71.
18. Chen Q, Chen XZ, Wang JM, Li SW, Jiang T, P. DJ. Intracranial meningeal hemangiopericytomas in children and adolescents: CT and MR imaging findings. AJNR Am J Neuroradiol. 2012;33(1):195-9.
19. Akiyama M, Sakai H, Onoue H, Miyazaki Y, Abe T. Imaging intracranial haemangiopericytomas: study of seven cases. Neuroradiology. 2004;46(3):194-7.
20. Sibtain NA, Butt S, Connor SE. Imaging features of central nervous system haemangiopericytomas. Eur Radiol. 2007;17(7):1685-93.
21. Zhou JL, Liu JL, Zhang JMZ. Thirty-nine cases of intracranial hemangiopericytoma and anaplastic hemangiopericytoma: a retrospective review of MRI features and pathological findings. Eur J Radiol. 2012;81(11):3504-10.
22. Barba I, Moreno A, Martinez-Pérez I, Tate AR, Cabañas ME, Baquero M, et al. Magnetic resonance spectroscopy of brain hemangiopericytomas: high myoinositol concentrations and discrimination from meningiomas. J Neurosurg. 2001;94(1):55-60.
23. Mena H, Ribas JL, Pezeshkpour GH, Cowan DN, Parisi JE. Hemangiopericytoma of the central nervous system: a review of 94 cases. Hum Pathol. 1991;22(1):84-91.
24. Rajaram V, Brat DJ, Perry A. Anaplastic meningioma versus meningeal hemangiopericytoma:Immunohistochemical and genetic markers. Hum Pathol. 2004;35(11):1413-8.
25. Hayashi Y, Uchiyama N, Nakada M, Iwato M, Kita D, Higashi R, et al. A reevaluation of the primary diagnosis of hemangiopericytoma and the clinical importance of differential diagnosis from solitary fibrous tumor of the central nervous system. Clin Neurol Neurosurg. 2009;111(1):34-8.
26. Wu W, Shi JX, Cheng HL, Wang HD, Hang CH, Shi QL, et al. Hemangiopericytomas in the central nervous system. J Clin Neurosci. 2009;16(4):519-23.
27. Payne BR, Prasad D, Steiner M, Steiner L. Gamma surgery for hemangiopericytomas. Acta Neurochir (Wien). 2000;142(5):527-36; discussion 36-7.
28. Soyuer S, Chang EL, Selek U, McCutcheon IE, Maor MH. Intracranial meningeal hemangiopericytoma: the role of radiotherapy: report of 29 cases and

review of the literature. Cancer. 2004;100(7):1491-7.
29. Kim JH, Jung HW, Kim YS, Kim CJ, Hwang SK, Paek SH, et al. Meningeal hemangiopericytomas: long-term outcome and biological behavior. Surg Neurol. 2003;59(1):47-53; discussion -4.
30. Rutkowski MJ, Sughrue ME, Kane AJ, Aranda D, Mills SA, Barani IJ, et al. Predictors of mortality following treatment of intracranial hemangiopericytoma. J Neurosurg. 2010;113(2):333-9.
31. Sheehan J, Kondziolka D, Flickinger J, Lunsford LD. Radiosurgery for treatment of recurrent intracranial hemangiopericytomas. Neurosurgery. 2002;51(4):905-10; discussion 10-1.
32. Fountas KN, Kapsalaki E, Kassam M, Feltes CH, Dimopoulos VG, Robinson JS, et al. Management of intracranial meningeal hemangiopericytomas: outcome and experience. Neurosurg Rev. 2006;29(2):145-53.
33. Dufour H, Metellus P, Fuentes S, Murracciole X, Regis J, Figarella-Branger D, et al. Meningeal hemangiopericytoma: a retrospective study of 21 patients with special review of postoperative external radiotherapy. Neurosurgery. 2001;48(4): 756-62. Discussion 62-3.
34. Ghia AJ, Allen PK, Mahajan A, Penas-Prado M, McCutcheon IE, D. BP. Intracranial hemangiopericytoma and the role of radiation therapy: a population based analysis. Neurosurgery. 2013;72(2):203-9.
35. Kim JW, Kim DG, Chung HT, Paek SH, Kim YH, Han JH, et al. Gamma Knife stereotactic radiosurgery for intracranial hemangiopericytomas. J Neurooncol. 2010;99(1):115-22.
36. Chamberlain MC, Glantz MJ. Sequential salvage chemotherapy for recurrent intracranial hemangiopericytoma. Neurosurgery. 2008;63(4):720-6; author reply 6-7.

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