

Intracranial hemangiopericytoma in a 15 year-old child: neuroimaging features and management therapy.

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 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.

Key words: Hemangiopericytoma, child, surgery, radiotherapy, intracranial, MRI

23 **Introduction:**

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

33

34 **Case presentation:**

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

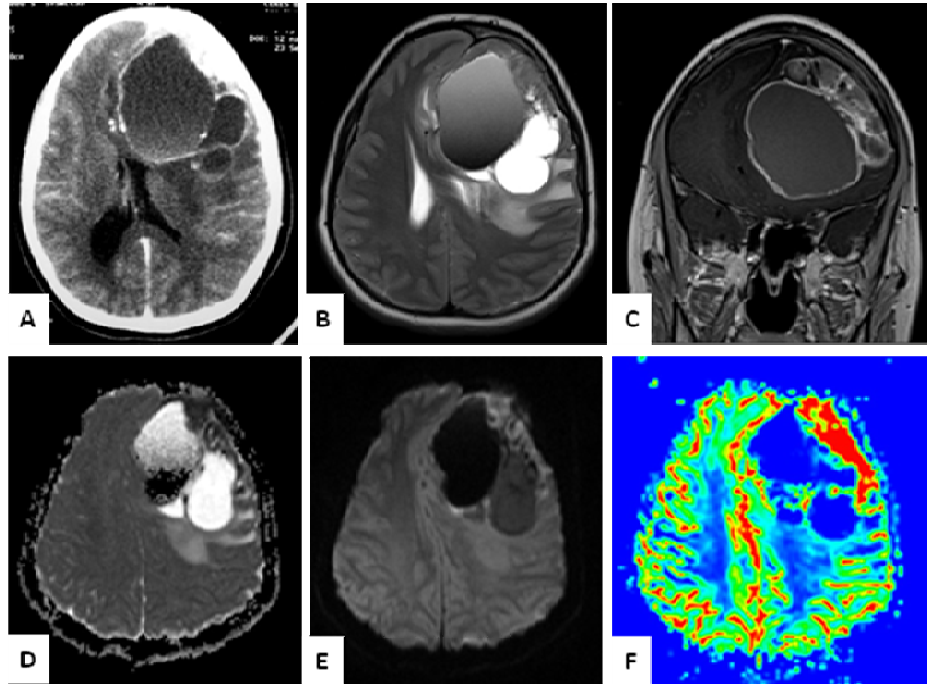
41 Computerized tomography (CT) scan showed a large multicystic mass, which
42 measures 12 x 8 cm, with peripheral enhancement located in the left frontal
43 parasagittal convexity (figure n°1A). The mass showed mixed low and isoattenuation
44 with moderate surrounding edema. Notable mass effect was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

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

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

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

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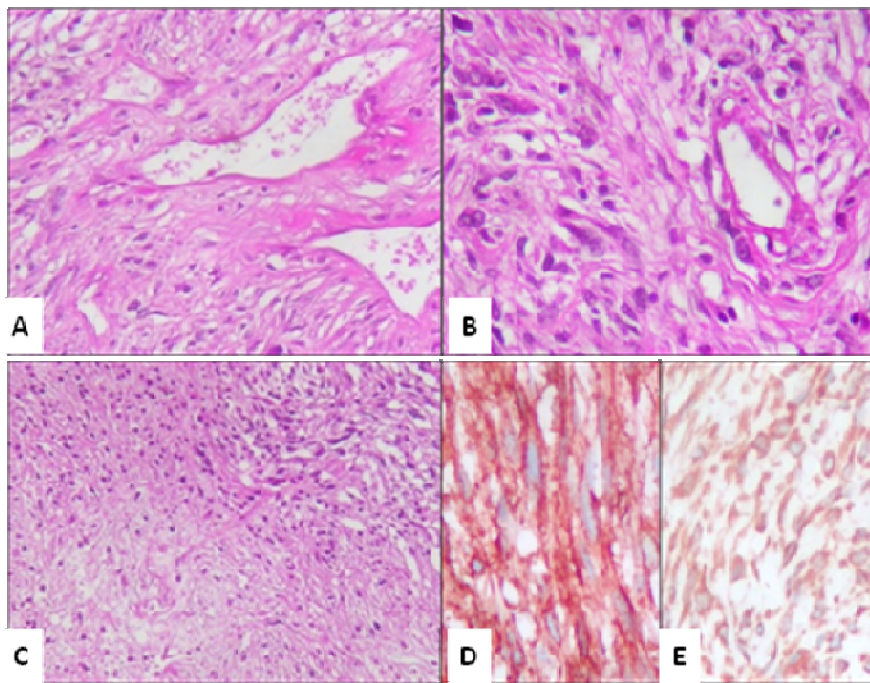
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62 Figure 1: A, cranial axial computed tomography with contrast enhancement shows a a left frontal large
 63 multicystic mass with peripheral enhancement. B, A, axial T2-WI displays an heterogeneous signal of
 64 this lesion. The largest cystic component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

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

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

83 Immuno-histochemistry showed diffuse positivity for vimentin, CD99, and bcl-2 and
84 variable positivity for CD34 (figure n°2). The diagnosis of anaplastic
85 hemangiopericytoma was made.

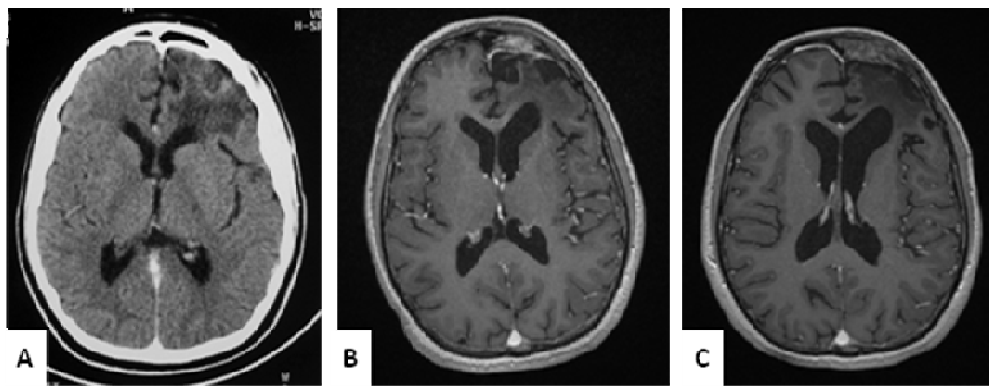


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87 Figure n°2: Histopathological and immunohistochemical evidence displaying a highly cellular and
88 vascularized mesenchymal tumor with dilated, staghorn-type vessels (A), High nuclear atypia (B) and
89 necrosis (C). Ki67 index was 25%. The tumor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

91

92 One month postoperatively, the child started whole-brain radiotherapy. She received
93 54 Gy without incidents. On the fifth postoperative month, physical examination
94 showed improvement of Broca's aphasia but persistent decreased visual acuity in her
95 left eye. However, MRI revealed a local recurrence (figure n°3B). Therefore, an
96 additional radiation dose of 10 Gy was delivered. After 1-year follow-up, the
97 recurrence remained unchanged on MRI (figure n°3C) and No metastases were
98 found on body scan.



99
100 Figure 3: A, cranial axial CT scan with contrast enhancement on the first postoperative month shows a
101 total gross resection of the HPR. B, post-enhanced axial T1-WI revealing a local recurrence of the tumor
102 3 months after radiotherapy. C, after an additional radiation dose of 10 Gy The recurrence remained
103 unchanged on the 1-year follow-up post-enhanced axial T1-WI.

104

105 Discussion:

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

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

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

121 **Neuro-imaging features of intracranial HPRs are non-specific but show differences**
122 **with regard to meningiomas.** Indeed, intracranial HPRs are multilobulated, and their

123 location is similar to that of meningiomas. However, HPRs present a narrow base of
124 dural attachment in one third of cases, which is not typically seen in meningiomas
125 (17). They usually are large, even in children, with a mean diameter of 75 mm (18).
126 On CT-scan, HPRs are heterogeneous, hyperdense and typically show
127 heterogeneous enhancement. They also show bony erosion, frequently seen in most
128 anaplastic form but they lack calcifications and hyperostosis, usually observed in
129 meningiomas (17, 19, 20). MRI findings show that intracranial HPR are
130 heterogeneous, isointense on T1 and T2-WI with signal flow voids and enhance
131 heterogeneously (17, 19, 20). In some cases, the serpentine signal intensity voids in
132 vessels are numerous and large, giving an AVM-like appearance on MRI, which is
133 suggestive of HPR (18). According to Zhou et al., pronounced lobulation, necrosis,
134 cystic properties and damage of adjacent skull on MRI findings are more common in

anaplastic form of HPR (21). On magnetic resonance spectroscopy, Barba et al. noted that elevated myoinositol using short TE (20 msec) may help differentiate HPR from meningioma (22).

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 the treatment of HPR is discussed. Several studies have shown that 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.

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.

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