Intracranial Capillary Hemangioma – a Case Report and Literature Review

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Case report

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Abstract

Background

Capillary hemangiomas of the central nervous system are exceedingly rare. These are benign tumour-like lesions that usually occur in the skin and soft tissues of mainly children. We report the case of an extra-axial capillary hemangioma mimicking a tentorial meningioma.

Case presentation

A 46-year old female patient presented with headaches. Imaging revealed an avidly enhancing tentorial lesion with a wide dural base, features suggestive of a meningioma. Intra-operatively, an extra-axial heavily vascular tumour was found and was radically resected en bloc. Histology revealed thin-walled capillary-sized vessels arranged in closely packed lobules, and a diagnosis of capillary hemangioma was made. At one year follow up, the patient was asymptomatic and no tumour recurrence was noted.

Conclusions

A literature review showed that capillary hemangiomas may be misdiagnosed as other extra-axial dural-based lesions such as meningiomas, schwannomas, hemangiopericytomas and inflammatory granulomatous lesions. Diagnosis should be considered especially in radiologically atypical cases. Complete excision is curative. Stereotactic radiosurgery on its own or in conjunction with surgery also achieves good results while partial excision or biopsy alone often leads to recurrence.

Background

Capillary hemangiomas are benign tumour-like lesions usually occurring in the skin and mucosal tissues. They present most commonly as a strawberry naevus during infancy with spontaneous regression usually by age 5. In adults, they may present as lesions usually of the face or scalp. There are only rare accounts of these lesions appearing in the central or peripheral nervous system, most often seen in the spinal roots or cauda equina. Intracranial capillary hemangiomas are extremely rare.

We present a case of tentorial base capillary hemangioma initially thought to be a meningioma, but subsequently confirmed histologically as a capillary hemangioma. We review the literature on intracranial capillary hemangiomas, their presentations and outcome. We also discuss how these lesions differ from other vascular lesions of the CNS from a radiological and histological perspective.

Case Presentation

A 46-year old woman presented with a 3-month history of right sided headaches, drowsiness and feeling drained. The headaches were constant and dull, with no obvious exacerbating or relieving factors, with no associated nausea, vomiting or visual problems. She had a past medical history of hypertension managed with a single medication alone. Neurological examination was unremarkable.

MRI examination revealed a well-defined right-sided tentorium base tumour, measuring 3.7 x 3.1 x 3 cm, and enhancing heterogeneously (see Fig. 1). It extended on either side of the tentorium with the larger component being in the supratentorial subtemporal region. Flow voids were visible, indicating the vascular nature of the lesion. There was moderate surrounding brain oedema. This lesion was initially felt to be a meningioma given the extra-axial nature and wide-based dural attachment. She underwent a right temporal craniotomy and microscopic subtemporal approach to expose the tumour. The lesion was noted to be vascular in nature, encapsulated by dura, and was excised en bloc. The tentorium was opened and the subtentorial nodule was resected. Histological examination revealed thin-walled capillary-sized blood vessels arranged in closely packed lobules in a sinusoidal pattern (see Fig. 3). The stroma was rich in smooth muscle cells, with no mitotic or necrotic features. Ki-67 proliferation fraction was extremely low indicating a low-grade lesion. CD34 and CD31 immunopreparation highlighted vascular endothelium only. Epithelial membrane antigen (EMA) and STAT-6 were negative excluding meningioma and hemangiopericytoma respectively. Diagnosis was made of a capillary hemangioma. Post-operative MRI showed complete excision, and no recurrence was noted on repeat imaging one year later (see Fig. 2). She had no post-surgical complications, was able to return to work and remained well at one-year follow up.

Discussion

Hemangiomas refer to benign vascular tumour-like lesions found within skin and mucus membranes. Traditionally they are used to refer to neoplastic lesions alone, however non-neoplastic vascular malformations such as cavernous hemangiomas have also been included in this definition. They are classified into capillary, cavernous, mixed and cellular (1). Capillary hemangiomas are characterised by nodules of capillaries lined by flattened endothelium (2). These lesions are most often found within the skin and subcutaneous soft tissue, and very few reports describe their occurrence in the central nervous system (2). Due to the rarity of this type of lesion, there is insufficient data analysing their growth rate and natural history in the CNS, and no guidelines on their management (3).

We carried out a literature search using the terms (capillary hemangioma) AND (intracranial OR brain OR nervous system) to include case reports, case series, reviews and meta-analyses, which produced a result of 281 articles. Abstracts were analysed and 27 of these papers were included as pertaining to intracranial capillary hemangiomas, which included a total of 32 diagnosed cases of which 17 in paediatric patients, and 15 in adults (see Tables 1 and 2). There was a wide age range from 0 to 79 years. A majority of patients (N = 17, 53%) were children below age 15, including 7 patients (22.9%) being less than a year old. A majority of these lesions were extra-axial and occurring in variable locations, most commonly the cavernous sinus (N = 6, 18.8%), followed by skull base and convexity regions. Three patients had multiple intracranial lesions (4–6), of which one was a child with widespread neurocutaneous capillary hemangiomas (4). Only 4 cases of capillary hemangiomas were intra-axial (4–8). Lesions diagnosed in newborns were giant in a majority (N = 3, 60%) of cases of newborn capillary hemangiomas, and presented with features of raised intracranial pressure (ICP) (9–11). Only one other patient aged 3 was diagnosed with a giant lesion (12). In older children and in adults, headache and

associated symptoms of raised ICP were the commonest presenting feature, followed by seizures and cranial nerve palsies. Of the 30 patients whose treatment was discussed in the paper, a majority (N = 17, 56.7%) were managed with complete resection of the tumour, usually en bloc. Five patients had partial resection with or without stereotactic radiosurgery (SRS), two had SRS alone and five had biopsy alone (see Table 3). No patient with complete excision or stereotactic radiosurgery had recurrence following a year. On the other hand, 60% of patients who underwent biopsy alone or partial resection with at least one year of follow up showed recurrence.

Propranolol, a medication used first line for cutaneous capillary hemangiomas has also been applied to intracranial capillary hemangioma in a single case alone (9). In this case, the patient, a neonate with a presumed giant posterior fossa capillary hemangioma based on detailed radiology assessment alone, was treated with propranolol for 6 months (9). Complete tumour regression was seen at one year post-treatment. Limited evidence suggests that complete surgical resection or SRS alone or in combination with surgery may be curative, however medical treatments including propranolol should also be further evaluated for their role in treating intracranial capillary hemangiomas.

Benign capillary hemangiomas are radiologically and histologically distinct from most other vascular tumours occurring in the CNS, although their clinical presentation may be similar. Cavernous hemangiomas consist of enlarged, dilated vessels, unlike the tightly packed capillary-sized vessels seen with capillary hemangiomas. They are often thrombosed with perivascular hemosiderin deposition and calcification. Radiologically, they occur intra-axially and display a characteristic popcorn like appearance on MRI (6). Clearer evidence exists from systematic reviews and meta-analyses as to their natural history and management compared to capillary hemangiomas (13). Capillary telangiectasia of the CNS is also a type of capillary ectasia, however this is characterised histologically by neural parenchyma interspersed between ectatic vessels rather than the closely packed and lobular pattern seen with capillary hemangiomas. Radiologically they are intraparenchymal, usually found in the pons, lack clear demarcation and display a characteristic “brush” or “stipple”- like enhancement (14). They are most often asymptomatic and may be managed conservatively in a large majority of cases.

It is often difficult to radiologically distinguish these lesions from meningiomas, hemangiopericytomas and other vascular dural-based lesions of the CNS. In our literature review, we found most lesions were initially misdiagnosed as meningiomas from the radiological appearance. Hemangiopericytomas may also appear radiologically similar to capillary hemangiomas with their extra-axial location, dural attachment and vivid enhancement with intralesional signal voids, however they are different histological entities. They arise from fibroblast cells, are highly mitotic and cellular on histology, and classically express STAT-6 antibodies. Clinically, there is a risk of local invasion of bone and distant metastases to liver, lung or bone (15).

Conclusions

This case highlights a rare differential for an extra-axial dural-based lesion affecting mainly children, but also adults with a wide age range. They may often be misdiagnosed as a meningioma, hemangiopericytoma or another vascular lesion. Careful radiological and histological examination is important especially in the case of radiologically atypical tumours, so that they may be managed appropriately. Complete surgical resection is often achievable en bloc, with no future recurrence. However stereotactic radiosurgery and systemic steroids have also been found to ameliorate the lesion in a small group of patients.

Systematic reviews or case control studies evaluating patient outcomes for this rare condition will help establish guidelines for treatment regimens in these patients. Longer term follow up, especially in inoperable cases will help elucidate the natural history of these lesions.

Declarations

Ethics approval and consent to participate

Not applicable

Consent for publication

Written consent given by patient

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study

Competing interests

None

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None

Author's contributions

AG: Drafted manuscript

HC: Assisted in drafting manuscript

JJ: Edited radiology images and guided radiology-related discussion

EA: Edited and supervised drafting of manuscript

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References

1. Le Bihannic A, Michot C, Heckly A, Loget P, Beucher A, Brassier G, et al. Capillary haemangioma arising from the anterior choroidal artery. *Child's Nerv Syst.* 2005;
2. Abe M, Misago N, Tanaka S, Masuoka J, Tabuchi K. Capillary hemangioma of the central nervous system: a comparative study with lobular capillary hemangioma of the skin. *Acta Neuropathol* [Internet]. 2005 [cited 2021 Jan 9];109(2):151–8. Available from: <https://pubmed.ncbi.nlm.nih.gov/15365728/>
3. Santoro G, Piccirilli M, Chiarella V, Greco N, Berra LV, Santoro A. Intracranial capillary hemangiomas: literature review in pediatric and adult population [Internet]. *Neurosurgical Review.* Springer Science and Business Media Deutschland GmbH; 2020 [cited 2021 Jan 9]. p. 1–9. Available from: <https://doi.org/10.1007/s10143-020-01419-8>
4. Uyama A, Kawamura A, Akiyama H, Nakamizo S, Yamamoto K, Nagashima T, et al. A case of cerebellar capillary hemangioma with multiple cysts. *Pediatr Neurosurg.* 2008;
5. Maurer GD, Schittenhelm J, Ernemann U, Kempf VAJ, Ritz R, Weller M, et al. Intracranial hemangiomas in a patient with POEMS syndrome. *J Neurol.* 2010;
6. Younas F, Durrani Q, Shahzad MA, Mushtaq A, Imlay S. Multiple intracranial capillary hemangiomas and transient cerebrovascular insufficiency. *Neurol Sci.* 2011;
7. Koga Y, Hamada S, Saito H, Akai T, Kuroda S. Intracranial, Intra-parenchymal Capillary Hemangioma – Case Report –. *NMC Case Rep J.* 2020;
8. John SG, Pillai U, Lacasse A. Intracranial capillary hemangioma mimicking a dissociative disorder. *Clin Pract.* 2012;
9. Cavaleiro S, Do Amaral Campos HG, Da Costa MDS. A case of giant fetal intracranial capillary hemangioma cured with propranolol. *J Neurosurg Pediatr.* 2016;
10. Jalloh I, Dean AF, O'Donovan DG, Cross J, Garnett MR, Santarius T. Giant intracranial hemangioma in a neonate. *Acta Neurochir (Wien).* 2014;
11. Daenekindt T, Weyns F, Kuan HK, Peuskens D, Engelborghs K, Wuyts J. Giant intracranial capillary hemangioma associated with enlarged head circumference in a newborn: Case report. *J Neurosurg Pediatr.* 2008;
12. Zheng SP, Ju Y, You C. Giant intracranial capillary hemangioma in a 3-year-old child: Case report and literature review. *Clin Neurol Neurosurg.* 2012;
13. Akers A, Al-Shahi Salman R, Awad IA, Dahlem K, Flemming K, Hart B, et al. Synopsis of guidelines for the clinical management of cerebral cavernous malformations: Consensus recommendations based on systematic literature review by the angioma alliance scientific advisory board clinical experts panel. *Clin Neurosurg.* 2017;
14. Wong M, Shatzkes D. Brain capillary telangiectasia. *Appl Radiol.* 2013;

15. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvett A, et al. The 2007 WHO classification of tumours of the central nervous system. *Acta Neuropathologica*. 2007.

Tables

Table 1

Reported cases of paediatric capillary hemangioma along with their demographics, location in brain and presentation

Case number	Age (in years)	Sex	Intracranial location	Intra-axial (IA) or extra-axial (EA)	Presentation	Authors
1	Neonate	M	Posterior fossa - giant	EA	Raised ICP	Cavalheiro., 2016
2	Neonate	M	Skull base – middle cranial fossa	EA	Raised ICP	Jalloh et al., 2014
3	Neonate	M	Skull base – middle cranial fossa	EA	Raised ICP	Daenekindt et al., 2008
4	Neonate	M	Skull base – anterior cranial fossa	EA	Raised ICP	Le Bihannic et al., 2005
5	Neonate	M	Posterior fossa - giant	EA	Raised ICP	Haine et al., 2017
6	3 month	M	Skull base – cerebellopontine angle	EA	Hypotonia	Karikari et al., 2006
7	4 month	F	Cerebellar + multiple hemangiomas in other organs	IA	Raised ICP	Uyama et al., 2008
8	1	M	Convexity - frontal	EA	Seizures	Willing et al., 1993
9	3	F	Skull base – middle cranial fossa	EA	Raised ICP	Zheng et al.m 2012
10	8	M	Skull base – middle cranial fossa	EA	Raised ICP	Watanabe et al., 2001
11	10	M	Convexity – parietal	EA	Raised ICP	Dadoub et al., 2016
12	10	M	Tentorial	EA	Raised ICP	Grabb, 2016
13	10	F	Midline – superior sagittal sinus	EA	Raised ICP	Brotchi et al., 2005
14	14	M	Sylvian fissure	EA	Diplopia	Grabb, 2016
15	14	M	Skull base – cerebello-pontine angle	EA	Facial palsy, deafness, dysphagia, vertigo	Morace et al., 2012

Case number	Age (in years)	Sex	Intracranial location	Intra-axial (IA) or extra-axial (EA)	Presentation	Authors
16	15	F	Parietal	IA	Headache	Koga et al., 2020
17	15	F	Cavernous sinus	EA	Diplopia	Tsao et al., 2003

Table 2
Reported cases of adult capillary hemangioma along with their demographics, location in brain and presentation

Case number	Age (in years)	Sex	Intracranial location	Intra-axial (IA) or extra-axial (EA)	Presentation	Authors
1	19	F	Cavernous sinus	EA	Diplopia	Tsao et al., 2003
2	23	F	Cavernous sinus	EA	Diplopia	Massman et al., 2021
3	26	F	Skull base – middle cranial fossa	EA	Raised ICP	Smith and Skelton, 2007
4	26	F	Cavernous sinus	EA	Pituitary dysfunction	Morace et al., 2012
5	31	F	Tentorial	EA	Raised ICP	Simon et al., 2005
6	35	F	Convexity - occipital	EA	Raised ICP	Srinivasan et al., 2016
7	36	F	Skull base – Meckel's cave	EA	Facial pain	Santoro et al., 2020
8	42	M	Convexity - temporoparietal	EA	Dysphasia	Morace et al., 2012
9	44	F	Multiple	EA	Diplopia	Maurer et al., 2009
10	59	F	Convexity – frontal	EA	Raised ICP	Almaghrabi et al., 2018
11	59	M	Convexity - temporoparietal	IA	Falls, dissociative disorder	John et al., 2012
12	59	F	Skull base – infundibular recess	EA	Headache	Lee et al., 2010
13	61	F	Cavernous sinus	EA	Visual impairment	Morace et al., 2012
14	65	F	Cavernous sinus	EA	Diplopia, periorbital pain	Nawashiro et al., 2011
15	69	M	Multiple	IA	Seizures	Younas et al., 2011

Table 3
Management approach and reported outcomes in 29 reported cases of capillary hemangioma

Management approach	No. of cases	Outcome
Complete excision	17	No recurrence at ≥ 1 year in 13 cases No recurrence at < 1 year in 3 cases No outcome data for 1 case
Partial excision + SRS	2	No residual or recurrence at ≥ 1 year
SRS alone	2	No residual or recurrence at ≥ 1 year
Biopsy + dexamethasone	1	No residual or recurrence at < 1 year
Biopsy or partial excision alone	6	Increased lesion size in 3 cases (2 cases > 1 year, 1 case < 1 year) Stable disease at ≤ 1 year in 2 cases No follow up in 1 case
Propranolol	1	No residual or recurrence at ≥ 1 year

Figures

Figure 1

Pre-operative MRI images, T1 with gadolinium contrast axial (a) and coronal (b) views; T2 FLAIR axial (c) and T2 gradient echo coronal (d) views.

Figure 2

Post-operative MRI images, T1 with gadolinium contrast axial (a) and coronal (b) views. 1-year follow-up MRI images, T1 with gadolinium contrast axial (c) and coronal (d) views.

Figure 3

Histological images showing thin-walled capillary sized vessels arranged in closely packed lobules in a sinusoidal pattern with a stroma rich in smooth muscle cells, features suggestive of a capillary hemangioma.