Case Report

A unique case of benign intracranial hemangioma mimicking malignant transformation

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ABSTRACT

Capillary hemangiomas are rare benign vascular lesions, commonly found on scalp, face, chest, or back of a neonate or infant. Hemangiomas of the central nervous system are very rare lesions. There are only a few cases of intracranial capillary hemangioma (ICH) arising in adults reported in the literature. We present a case of 59-year-old female with intermittent recurrent headache localized in the frontal area. Magnetic resonance imaging revealed left frontal extra-axial mass with peripheral enhancement. The patient underwent complete surgical resection of the tumor. Histopathology examination of the lesion revealed well defined vascular lesion composed of closely packed plump endothelial cells lining slit-like vascular channels containing scattered red blood cells. No evidence of infiltrative brain parenchyma was seen. Ki-67 proliferative index was low, less than 2%. The final diagnosis was confirmed to be ICH by histopathology and immunohistochemistry studies. The patient has remained healthy and free of disease 39 months since her initial surgery. ICH is a benign vascular lesion which rarely occurs in the central nervous system, particularly in the intracranial region. It can mimic malignant lesions on radiologic studies. Histopathology examination is the gold standard for diagnosis. If total resection is achieved, prognosis is generally good with no evidence of recurrence.

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1. Introduction

In 1867, Virchow first described hemangiomas [1]. Hemangiomas are benign vascular tumors, or tumor-like lesions. These lesions typically grow in the skin, soft tissue, face, scalp, and trunk. Neonates and infants are the most affected age group. Ten to twelve percent shows tendency to grow within the first year of life [2]. Hemangioma can rarely grow in adults, with a slight female predominance, which undergoes remarkable size change due to pregnancy and hormonal cycles’ response [3]. Clinically hemangioma goes through a proliferate phase before proceeding into an involutionary phase [4]. Most hemangiomas express spontaneous regression with age progression. Histologically, hemangiomas are classified into two main subtypes: capillary and cavernous. Capillary hemangiomas (CHs) are composed of lobules separated by variable degree of fibrous bands. These lobules are cellular due to the plump endothelial cells lining the vascular spaces and poorly defined capillary channels. Cavernous hemangiomas are large cystically dilated blood vessels with thin walls. In-
travascular thrombosis or calcification is frequent. Immuno-

histochemistry highlights the endothelial cells lining in clas-

sic slit-like spaces. CHs of the central nervous system are
dreaded as atypical or rare diagnosed pathology [2]. Although spinal nerve roots and
cauda equina are the favorable locations for CH, CH within the
brain is unusual. Review of the literature revealed that most
of the intracranial capillary hemangioma (ICH) was diagnosed
originally by brain computed tomography and magnetic reso-
nance imaging as meningioma prior to surgical resection. Due
to studies limitation and few reported cases about ICH, the
estimated rate, prevalence, and radiological findings might be
underestimated. In this paper, we present a case of a 59-year-
old female with left frontal extra-axial mass with peripheral
enhancement, confirmed histologically as an ICH.

2. Case presentation

A 59-year-old female was admitted to the emergency depart-
ment complaining of intermittent recurrent headache for the
past 3 months, localized in the frontal area. The headache
resolved spontaneously without medication. No associated
symptoms was identified. The patient demonstrated normal
vital signs and no focal neurologic deficit on physical exami-
nation. Magnetic resonance imaging of brain was performed
utilizing standard technique with gadolinium administration.
The study showed a peripherally located extra-axial mass le-
son in the left frontal region, measured 3.4 cm Antero-

posterior × 2.6 cm transverse × 1.4 cm craniocaudal. The le-
son caused significant vasogenic edema in the surround-
ing brain parenchyma. Postgadolinium images showed strong
predominantly peripheral enhancement within tumor mass
with probable central area of necrotic component identified
(Fig. 1A and B). Perfusion study showed significant increase
within the peripherally enhancing aspect of the tumor (Fig.
1C). The top differential diagnosis was meningioma with atyp-
tical features and significant surrounding edema, or astro-
cytoma. The patient underwent a navigation-guided brain
mass resection under general anesthesia. Solid round mass
was identified, closely related to the brain parenchymal tis-

ue, which was completely removed. Intraoperative bleed-
ing was successfully controlled. Postoperatively, the patient
was stable, doing well. Postoperative examination reveals fo-
cal hematoma without definite active tumor enhancement
detected (Fig. 1D). There was a significant decrease in the
surrounding vasogenic edema compared to the preoperative
image. The resected specimen was sent to anatomic pathol-
ology department for prober evaluation. Macroscopic exami-
nation revealed an oval mass, well-defined, partially capsul-
ated measured 3.5 × 2.5 × 1.5 cm, homogenous pink tan
cut-surface with hemorrhage cystic spaces. No area of necro-
sis was seen. Microscopic examination revealed a well de-
marcated vascular lesion (Fig. 2A) composed of thin vascu-
lar spaces lined by delicate plump endothelial cells. These
spaces are filled with red blood cells (Fig. 2B). No interven-
ing glial parenchymal tissue was seen. No evidence of nu-
clear atypia, mitotic figures, necrosis, and apoptosis was seen.
No evidence of intracytoplasmic hyaline body globules, ex-
tramedullary hematopoiesis was seen. Immunohistochem-

try studies revealed diffuse positive antigenicity to endo-
theelial cells markers including CD31 (Fig. 2C), CD34 (Fig. 2D),
factor VIII, and vimentin, while negative for epithelial mem-
brane antigen, S-100, human melanoma black, CD10, smooth
muscle actin, desmin, D2-40, alpha-fetoprotein, neuron-
specific enolase, glial fibrillary acidic protein, and signal trans-
ducer and activator of transcription 6. Ki-67 proliferative index
was less than 2%. Histopathology and immunohistochemical
studies were consistent with the diagnosis of ICH. The patient
had a continuous follow-up after the surgery for 40 months
duration, and she is healthy without any complications and/or
recurrence (Fig. 1E).

3. Discussion

CHs are seen in 1–2.6% of live births [2]. These tumors demon-
strate female predominance, and can undergo hormonal re-
ponse changes. The pathogenesis of hemangiomas is not
fully understood. No single theory can explain the predilec-
tion of hemangioma for infants, females, responses to hormonal
levels, and spontaneous involution. Most theories suggested
the origin of angioblasts, trophoblasts, along with defect in
the cytokine regulatory pathways that can initiate the pro-
cess of angiogenesis of hemangiomas [5]. Only few reports and
studies are found in the literature about CH arising primarily
in the brain. PubMed search until March 2018 reveals only 19
studies reporting 29 cases of ICH, confirmed by histopathology
examination. Majority of ICH was diagnosed in infants and
young adults with the age range from 2 weeks to 69 years old.
These cases were seen in 14 male and 15 female patients [2].
ICH can arise in the cerebral lobe [6], sagittal sinus [7], cere-
bellum [6], sellar region [8], cavernous sinus [9], fourth ven-
tricles [10], and anterior choroidal artery [11]. Clinical signs
and symptoms are variable and depend on lesion location,
ranging from asymptomatic to headache, seizure, and cranial
nerve palsy [2]. Radiological differential diagnoses in most of
the reported cases were meningioma, astrocytoma, and any
the tumors with high-grade features. Most of them were di-
agnosed as ICH postoperatively by histopathology examina-
tion. Postgadolinium studies reveal the peripheral enhance-
ment mimicking other tumors. Therefore, it is difficult to dis-
tinguish ICH preoperatively. Generally, most of the previous
cases, including this case were treated with complete surgical
resection. Treatment for ICH includes surgical resection,
embolization, laser treatment, β-blockers such as propranolo,
corticosteroids, interferon. However, these different modal-
ities are not clearly standard in the literature [12]. When gross
total resection of the lesion cannot be achieved, the patient
should be observed frequently, with consideration of adjuvant
radiotherapy [3]. Histopathology examination is the gold stan-
dard for diagnosis. Hematoxylin and eosin stain can easily
highlight the delicate vascular channels lined by endothelial
cells. Most cases in the literature were described as capillary
type intracranial hemangioma. However, one case reported a
mixture of capillary and cavernous intracranial hemangioma
[13].

The differential diagnoses include hemangioblastoma, hemangioendothelioma, and hemangiopericytoma. Heman-
Fig. 1 – Intracranial capillary hemangioma with avid peripheral enhancement. (A) Sagittal and (B) coronal magnetic resonance imaging (MRI) view showing extra-axial mass lesion in the left frontal region with strong predominant peripheral enhancement. (C) Perfusion study showed significant increased perfusion within the peripherally enhancing aspect of the tumor. (D) Postoperative examination reveals focal hematoma without definite active tumor enhancement. (E) Axial MRI scan performed on the patient ~39 months following initial surgery, no recurrence of the tumor was detected.
Intracranial capillary hemangioma. (A) Low-power examination reveals a well-demarcated lesion, partially capsulated vascular lesion (hematoxylin and eosin (H&E); 4x). (B) High-power examination shows vascular spaces lined by plump endothelial cells containing red blood cells (H&E; 40x). (C) Immunohistochemistry stain reveals positivity for CD31 in the vascular spaces (40x). (D) Immunohistochemistry shows positivity for CD34 in the vascular spaces (40x).

glioblastoma is a slow growing and indolent tumor, that arise commonly in young to middle-age, typically in the posterior fossa. They can be either sporadic or associated with von Hippel–Lindau disease associated. They typically present with cyst with mural enhancing nodule and are composed of neoplastic cells showing nuclear pleomorphic changes, lipidized stromal cells, and highly vascular background. Foci of extramedullary erythropoiesis may be seen. These features were not detected in our case. Hemangioendothelioma was excluded from our differential. It shows well-differentiated nests and cords of cells with abundant eosinophilic cytoplasm and prominent intracytoplasmic lumina, commonly seen in epithelioid form. Kaposiform hemangioendothelioma are biphasic tumors with both vascular and lymphatic component, demonstrating irregular, infiltrating nodules of compressed blood vessels, with a dense hyaline stroma. Retiform hemangioendothelioma expresses a net-like pattern of infiltrative growth. Pseudomyogenic and composite subtypes are rare form of bones and soft tissue hemangioendotheliomas. All of these patterns of growth were not observed in our case [14, 15]. Hemangiopericytoma characteristically grows in a sheet like, storiform pattern, around an intratumoral staghorn blood vessel. All of the above-mentioned differential diagnoses were against our case. Overall, ICH are usually benign, slow growing tumors with good prognosis and long free survival rate.

4. Conclusion

We present a case of 59-year-old female with ICH. Perioperative radiological diagnosis is usually difficult to make. Al-
though ICH is rare to occur, it should be in the differential diagnosis of defined mass with peripheral enhancement. ICH has a good prognosis with low rate of recurrence after surgical resection.

Ethics approval Informed consent

Verbal and written informed consent was obtained from the patient for their anonymized information to be published in this article.

Patient consent

Consent to publish the case report was obtained. This report does not contain any personal information that could lead to the identification of patient.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Conflict of interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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