Annals of Neurological Surgery

Case Report
Published: 13 Apr, 2018

A Giant Extradural Infantile Hemangioma of the Middle Cranial Fossa with Bone Erosion

*Qiang Li, Jianguo Xu and Yan Ju**

Department of Neurosurgery, West China Hospital of Sichuan University, China

Abstract

Background: Intracranial infantile hemangioma is a very rare disease, and its treatment remains a challenge.

Case Description: A 2-month old boy presented with a giant mass in the left middle cranial fossa. Postcontrast MRI showed homogenous enhancement, and craniotomy was performed. The tumor was totally resected, and diagnosis of intracranial infantile hemangioma was confirmed.

Conclusion: Surgery is a safe treatment option for intracranial infantile hemangioma. It relieves intracranial hypertension instantly, and ensures complete treatment of this rare disease.

Keywords: Infantile hemangioma; Craniotomy; Propranolol

Abbreviations

CT: Computed Tomography; HIF: Hypoxia Induced Factor; IH: Intracranial Hemangioma; MRI: Magnetic Resonance Imaging; MMP: Matrix Metalloproteinase; TIMP: Tissue Inhibitor of Metalloproteinase; VEGF: Vascular Endothelial Growth Factor

Introduction

Infantile Hemangioma (IH), also known as "capillary hemangioma" or "strawberry birthmarks", is the most prevalent benign neoplasm in infant, with an incidence of 4% to 10% [1]. However, intracranial infantile hemangioma is extremely rare, and only about 0.1% of pediatric IH patients had intracranial involvement [2]. Most of the lesions are within subarachnoid or ventricular spaces, and extradural IH is rarely reported [3]. In this article, we report a giant extradural IH located in the left middle cranial fossa causing bone erosion in a 2-month old boy.

Case Description

A 2-month and 15-day old baby was referred to our department because of incidental finding of intracranial mass. Ten days prior to his admission, the boy suffered from falling injury, and CT scan in the local hospital revealed a big mass in the left middle cranial fossa, without apparent brain injury. His physical examination in our hospital was unremarkable. A Computed Tomography (CT) angiography showed marked enhancement of the mass, damage to temporal bone, and no apparent vascular supply to the lesion. Gadolinium-enhanced Magnetic Resonance Imaging (MRI) scan showed a giant mass expanding from sphenoid to petrosal ridge, with intense homogenous enhancement. The lesion was about 5 cm in diameter. Left Sylvian fissure and middle cerebral artery was displaced. Bilateral subarachnoid fluid collection was present in frontal and temporal region (Figure 1). Because we could not exclude malignant tumor, such as sarcoma, and stereotaxic biopsy could cause catastrophic bleeding and aggravate its mass effect, we decided to resect the tumor via craniotomy. Under general anesthesia and readiness of blood transfusion, the patient underwent total resection of the tumor. The temporal bone was corroded, and tumor was seen just under temporal muscle. It was pink, yellow and greyish in color, pliable but a little bit tenacious in texture like sponge. The tumor was not as rich in blood perfusion as we previously thought it would be. The operation went smoothly, with little blood loss and no transfusion. Postoperative pathology confirmed diagnosis of infantile hemangioma. Glut1 was positive in immunohistochemistry. The patient's postoperative course was uneventful.

Discussion

Infantile hemangiomas are common benign tumors in pediatric population, and they usually affect head and neck region. Intracranial IH is very rare, and up to now, only about 40 cases of

OPEN ACCESS

Correspondence:
Yan Ju, Department of Neurosurgery, West China Hospital of Sichuan University, No. 37 Guoxue Road, Chengdu, Sichuan Province, 610041, P.R.China, Tel: +86-28-18980601975; E-mail: juyanped001@163.com

Received Date: *19 Mar 2018*
Accepted Date: *10 Apr 2018*
Published Date: *13 Apr 2018*

Citation:
Li Q, Xu J, Ju Y. A Giant Extradural Infantile Hemangioma of the Middle Cranial Fossa with Bone Erosion. Ann Neurol Surg. 2018; 2(1): 1011.

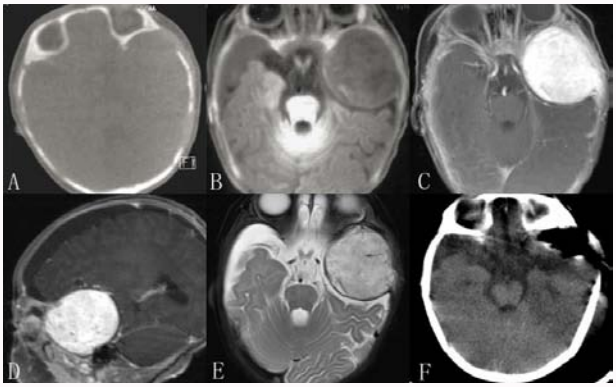

Figure 1: Radiological images for the present case.
A: CT in bone window showing erosion of left temporal bone.
B: T1- weighted MRI showing hypointensity of the lesion.
C: post-contrast T1-weighted MRI showing homogenous enhancement.
D: sagittal image of enhanced MRI showing bottom of the lesion, and subdural fluid collection.
E: T2-weighted MRI showing hyperintensity of the lesion.
F: Post-operative CT showing complete removal of the lesion.

intracranial infantile hemangioma has been reported in the literature [2-5]. The most prevalent location for IH is posterior fossa, mainly cerebellopontine angles or internal auditory canal [2]. It may be asymptomatic, or causes cranial nerve palsy, elevated intracranial pressure, hydrocephalus or intracerebral hematoma [5]. Intracranial IH occurs with or without PHACE syndrome, a rare neurocutaneous anomaly characterized by posterior fossa anomalies of the brain, arterial anomalies, cardiac anomalies, and eye anomalies [2,6]. Intracranial IH also occurs in the context of superficial IH, and search for intracranial lesions in dermal IH patients may be warranted [7].

The pathogenesis of IH is not fully elucidated. The abnormal capillaries inside IH arise from aberrant proliferation of immature endothelial cells stimulated by several angiogenic factors, such as Vascular Endothelial Growth Factor (VEGF), Matrix Metalloproteinase 2 (MMP2) and MMP9 [8]. It is reported that perinatal hypoxia is an important etiological factor for IH, and hypoxia induces tissue hypoxia induced factor-1α (HIF-1α), which promotes synthesis of VEGF protein, one of the most potent factors to promote vascular formation [9]. VEGF is the key to proliferation and migration of endothelial progenitor cells, which forms new vessels. In addition, VEGF can also induce synthesis of MMP, which degrades cellular basal membrane collagen to facilitate neo-vascular growth. MMP-9, in particular, can also mobilize endothelial progenitor cells from bone marrow, while VEGF enhance their proliferation.

The growth pattern of IH consists of two phases. A few weeks after birth, IH begins the rapid growing period until the age of 8-12 months old; after that, IH enters into a secondary recessive or slow growing phase [10]. The bi-phase pattern is closely related to serum levels of VEGF, MMP2, and MMP9. Przemyslaw et al. examined VEGF level in serum and tumor samples in 52 children; and they found that the serum VEGF was significantly higher in proliferative IH than recessive counterparts, and local VEGF was lower than that in peripheral blood [11]. Zhang et al. also found elevated serum level of VEGF in proliferative IH; while there were no difference between recessive IH, other vascular malformations and negative controls [12]. Zhong et al. reported increased MMP-2 and decreased Tissue Inhibitor of Metalloproteinses-2 (TIMP-2), a specific tissue inhibitor of MMPs, in proliferative IH samples than those in recessive IH [12]. Kleinman et al examined serum and tumor specimen expression of HIF-1α, VEGF and MMP9 in 10 children, and showed that all these mediators were significantly elevated in samples from children with proliferative IH [13]. These data suggest that angiogenic factors play an important role in the tumorigenesis and development of IH; targeting angiogenic signaling pathway may be a novel approach for IH treatment, even in the case of IH malignant transformation [14,15].

The best diagnostic imaging for IH is MRI because it displays precise anatomy for surrounding tissues. The tumor is characterized by iso- or hypo-intensity in T1-weighted imaging, and progressive, but homogenous enhancement after Gadolinium injection [16]. High signal intensity in T2 weighted image is typical for IH, and clear delineation of the tumor is useful to discern peripheral structures. Under microscope, the tumor consists of solid or lobular small capillaries, sinusoidal vessels, varied size of intraluminal spaces lined by plump endothelial cells, and thin basement membrane. These endothelial cells stain strongly for Glut1, a highly selective and definitive marker for IH diagnosis [17]. Glut1 is a glucose transporter protein located in normal capillary endothelial cells, and it is positive in 97% of IHs, but not in any non-IH vascular malformations [17]. The Glut1 positive endothelial cells are clonogenic, can convert to mesenchymal phenotype, and re-differentiate into stable endothelial cells, pericytes/smooth muscle cells or adipocyte, suggesting they have properties of stem cells [18]. It is interesting that proliferation of Glut1 positive cells can be inhibited dramatically by limited exposure to Rapamycin, indicating that this anti-angiogenic agent may be used to boost the effect of propranolol or corticosteroids, or may reduce their duration [18].

Treatment for intracranial IH includes observation, surgery and conservative treatment using steroids, interferon, or beta-blockers [2]. Since IH tend to stabilize or recess spontaneously after proliferative stage, observation seem to be a reasonable choice, when there is no life-threatening or disfiguring situations [10,19]. However, if the mass causes compression to critical structures, hydrocephalus, elevated intracranial pressure or intracerebral hematoma, surgery is indicated [3,5,20,21]. Our patient presented a giant mass with bone erosion and considerable compression to temporal and frontal lobe, surgical resection is appropriate. We could not determine nature of the mass beforehand, and had it been a malignant tumor, surgery would be the treatment of choice for her. Otherwise, if the lesion had been smaller, given its extradural location, conservative treatment with steroids or propranolol would be considered. Propranolol is a classical beta-adrenergic receptor antagonist that is used to treat heart arrhythmia. Recently, it has been applied “off-label” for intracranial IH with satisfactory results [2,8,16]. Propranolol constricts capillary vessels and reduces blood flow inside IH, and also reduces VEGF and MMP level via HIF-1α-dependent signaling pathway [22]. In the long run, it suppresses IH growth through induction of endothelial cell apoptosis [23]. However, since 10% to 30% dermal IH relapse after propranolol treatment, the efficacy and safety of propranolol for intracranial IH needs further study [24].

Conclusion

Our patient underwent total resection of the mass. When fully prepared, surgery is safe. It relieves intracranial hypertension successfully, and ensures complete treatment for intracranial IH.

Funding Source

This study is supported by National Natural Science Foundation Grant No.81602190.

References

1. Haggstrom AN, Drolet BA, Baselga E, Chamlin SL, Garzon MC, Horii KA, et al. Prospective study of infantile hemangiomas: clinical characteristics predicting complications and treatment. Pediatrics. 2006;118(3):882-7.
2. Kang E, Friedman N, Mamoun I, Tamburro J, Golden A. Beta Blockade as Treatment for Intracranial Infantile Hemangioma: Case Report and Literature Review. Pediatr Neurol. 2016;59:13-7.
3. Shakir HJ, McBride P, Reynolds RM. Dural-based infantile hemangioma of the posterior fossa: Case report. Surg Neurol Int. 2016;7:52.
4. Haine E, Sevely A, Boetto S, Delisle MB, Cances C. Infantile Hemangioma of the Posterior Fossa in a Newborn: Early Management and Long-Term Follow-up. Neuropediatrics. 2017;48(5):378-81.
5. Grabb PA. Surgical management of intracranial capillary hemangiomas in children: report of 2 cases. J Neurosurg Pediatr. 2016;17(3):310-7.
6. Heyer GL, Dowling MM, Licht DJ, Tay SK, Morel K, Garzon MC, et al. The cerebral vasculopathy of PHACES syndrome. Stroke. 2008;39:308-16.
7. Bihannic AL, Michot C, Heckly A, Loget P, Beucher A, Brassier G, et al. Capillary haemangioma arising from the anterior choroidal artery. Childs Nerv Syst. 2005;21(4):265-71.
8. Rotter A, de Oliveira ZNP. Infantile hemangioma: pathogenesis and mechanisms of action of propranolol. J Dtsch Dermatol Ges. 2017;15(12):1185-90.
9. Orozco-Covarrubias L, García-Valencia C, Ocariz SD, Ruiz-Maldonado R. Demographic and clinical characteristics in a cohort of Mexican mestizo children with infantile hemangioma. Dermatologia Revista Mexicana. 2014;58(3):215-24.
10. Chang LC, Haggstrom AN, Drolet BA, Baselga E, Chamlin SL, Garzon MC, et al. Growth characteristics of infantile hemangiomas: implications for management. Pediatrics. 2008;122(2):360-7.
11. Przewratil P, Sitkiewicz A, Andrzejewska E. Local serum levels of vascular endothelial growth factor in infantile hemangioma: Intriguing mechanism of endothelial growth. Cytokine. 2010;49(2):141-7.
12. Zhang L, Lin X, Wang W, Zhuang X, Dong J, Qi Z, et al. Circulating level of vascular endothelial growth factor in differentiating hemangioma from vascular malformation patients. Plast Reconstr Surg. 2005;116(1):200-4.
13. Kleinman ME, Greives MR, Churgin SS, Blechman KM, Chang EI, Ceradini DJ, et al. Hypoxia-induced mediators of stem/progenitor cell trafficking are increased in children with hemangioma. Arterioscler Thromb Vasc Biol. 2007;27(12):2664-70.
14. Pourazizi M, Kabiri S, Abtahi-Naeini B. Intralesional Bevacizumab (Avastin®) as a Novel Addition to Infantile Hemangioma Management: A Medical Hypothesis. J Res Pharm Pract. 2017;6(3):190-1.
15. Jeng MR, Fuh B, Blatt J, Gupta A, Merrow AC, Hammill A, et al. Malignant transformation of infantile hemangioma to angiosarcoma: response to chemotherapy with bevacizumab. Pediatr Blood Cancer. 2014;61(11):2115-7.
16. Cavalheiro S, Campos HG, Costa SD. A case of giant fetal intracranial capillary hemangioma cured with propranolol. J Neurosurg Pediatr. 2016;17(6):711-6.
17. North PE, Waner M, Mizeracki A, Mihm MC Jr. GLUT1: a newly discovered immunohistochemical marker for juvenile hemangiomas. Hum Pathol. 2000;31(1):11-22.
18. Huang L, Nakayama H, Klagsbrun M, Mulliken JB, Bischoff J. Glucose transporter 1-positive endothelial cells in infantile hemangioma exhibit features of facultative stem cells. Stem cells. 2015;33(1):133-45.
19. Tortori-Donati P, Fondelli MP, Rossi A, Bava GL. Intracranial contrast-enhancing masses in infants with capillary haemangioma of the head and neck: intracranial capillary haemangioma? Neuroradiology. 1999;41(5):369-75.
20. Philpott C, Wray A, MacGregor D, Coleman L. Dural infantile hemangioma masquerading as a skull vault lesion. AJNR Am J Neuroradiol. 2012;33(6):E85-7.
21. Daenekindt T, Weyns F, Goethem AV, Peuskens D, Engelborghs K, Wuyts J. Giant intracranial capillary hemangioma causing enlarged head circumference in a newborn: case report. Surg Neurol. 2007;68(2):195.
22. Li P, Guo Z, Gao Y, Pan W. Propranolol represses infantile hemangioma cell growth through the beta2-adrenergic receptor in a HIF-1alpha-dependent manner. Oncol Reports. 2015;33(6):3099-107.
23. Storch CH, Hoeger PH. Propranolol for infantile haemangiomas: insights into the molecular mechanisms of action. Br J Dermatol. 2010;163(2):269-74.
24. Chang L, Lv D, Yu Z, Ma G, Ying H, Qiu Y, et al. Infantile hemangioma: factors causing recurrence after propranolol treatment. Pediatric Res. 2018:175-82.