

Case of a Pregnant Woman with Capillary Hemangioma of the Parasellar Region

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Abstract

Here we report a rare case of capillary hemangioma (CH) in a 28-year-old woman suffering from gradual worsening diplopia at 28 weeks of pregnancy. Magnetic resonance imaging (MRI) showed a mass lesion (about 3 cm in diameter) in the right parasellar region. We decided to observe as she was pregnant, and had no symptoms other than right abducent nerve palsy. Fortunately, her symptoms did not worsen until delivery. Computed tomography, enhanced MRI, and angiography after delivery revealed that the lesion was highly calcified and vascularized. A dorsum sellae meningioma or highly calcified pituitary adenoma was suspected and the endoscopic transsphenoidal approach was used for tumor removal.

The postoperative course was uneventful. The histological diagnosis was CH.

Intracranial CHs or CHs of skull are rare vascular tumors. These tumors are reportedly more common in female patients and may change in size in adults according to menstrual cycle and pregnancy. Only six cases, including that of the present study, were diagnosed during the perinatal period. Some of them experienced rapid symptom progression and tumor growth in their course; thus, we should pay further attention to pregnant or peripartum patients with brain tumor, suspected hemangiomas.

Keywords: capillary hemangioma, pregnancy, transsphenoidal approach

Introduction

Capillary hemangiomas (CHs) are benign vascular tumors, many of which present congenitally or in early infancy. Lesions typically occur in the skin and soft tissue and are usually found on the face, scalp, chest, or back. The incidence of CHs was reported to be 1.1%-2.6% in full-term neonates.¹⁾ However, intracranial CHs (ICHs) or CHs of the skull are rare. Herein, we report the case of a pregnant woman with CH thought to have arisen from the dorsum sellae.

Case Report

This case report was prepared after informed consent was obtained from the patient.

A 28-year-old woman presented with diplopia after 28

weeks of pregnancy. Upon neurological examination, incomplete right abducent nerve palsy was detected, and no other neurological deficits were observed. Non-enhanced magnetic resonance imaging (MRI) revealed a mass in the pituitary region. At her presentation to the neurosurgical department, she was 30 weeks pregnant; hence, we chose to observe the lesion until delivery. After delivery, enhanced MRI, computed tomography, and angiography were performed. The mass lesion was observed mainly around the right cavernous sinus, and the normal pituitary gland was compressed to the left side. The lesion was highly calcified and vascularized (Fig. 1A-F). The patient did not show symptoms of pituitary deficiency, and her pituitary hormonal data were normal, except for prolactin, which was 193.7 ng/mL. However, the level was thought to be within the normal range for peripartum women. A meningioma arising from the sellar floor or dorsum sellae or a

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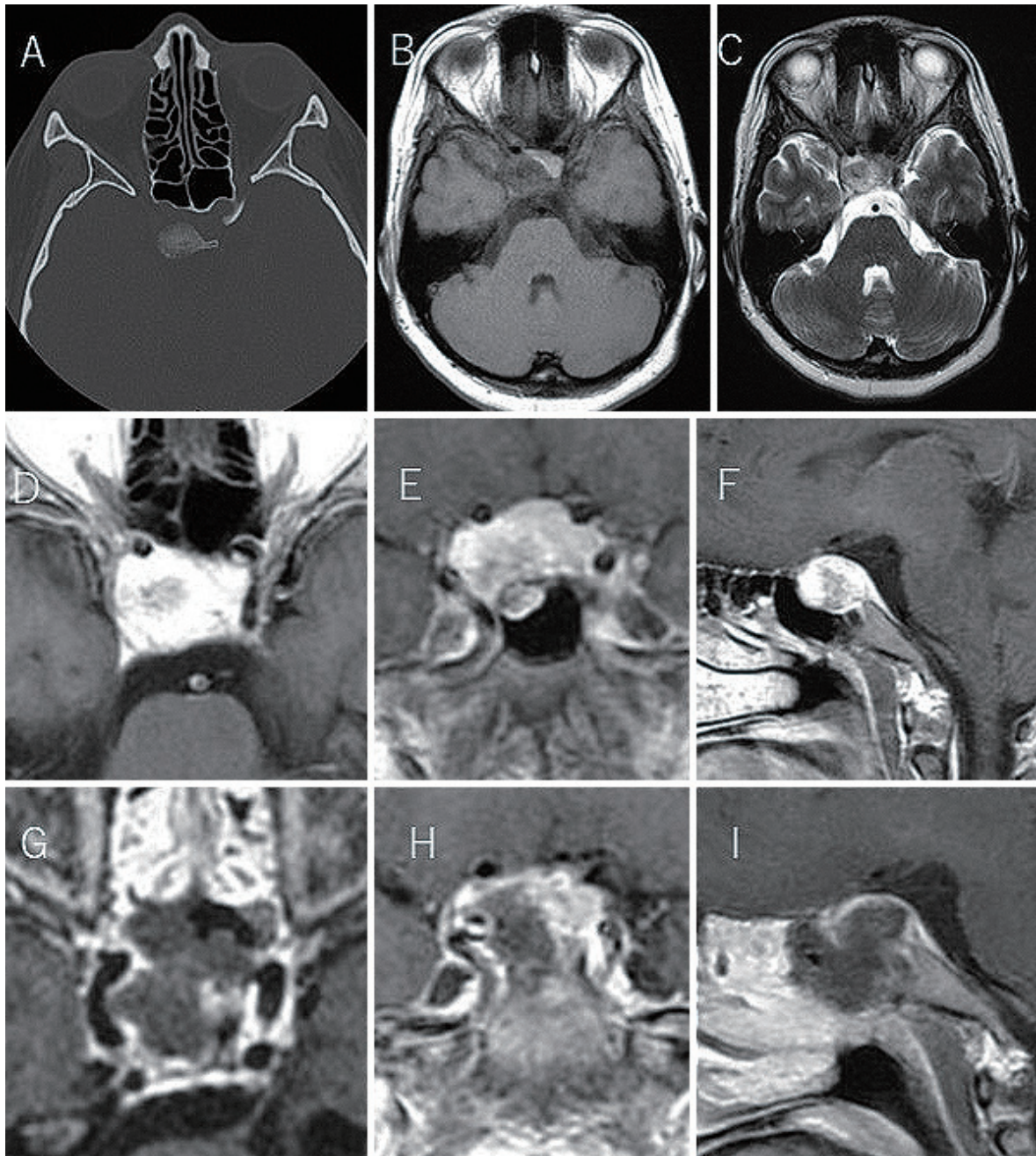


Fig. 1 Perioperative image studies.

Preoperative computed tomography of the bone window shows the dorsum sellae enlarged into the sellae turcica. (A) The lesion shows hypointensity on T1-weighted magnetic resonance imaging (MRI) (B) and mixed intensity on T2-weighted MRI. (C) The normal pituitary gland seems to be compressed to the left frontal side by the tumor. The lesion is well enhanced on gadolinium-enhanced MRI (D, E, F).

Postoperative gadolinium-enhanced MRI showed no residual tumor, and the pituitary gland remained normal (G, H, I).

highly calcified pituitary adenoma was suspected. Follow-up was also presented as an option because it was soon after giving birth. However, the patient and her family requested early resection and confirmation of the diagnosis. Therefore, we planned to perform tumor resection using the endoscopic transsphenoidal approach (eTSA) two

months after the delivery. The patient's diplopia improved when she was admitted to our department to undergo surgery, which was subsequently performed following her and her family's approval.

Intraoperatively, the bone around the right cavernous and clival portions of the internal carotid artery were

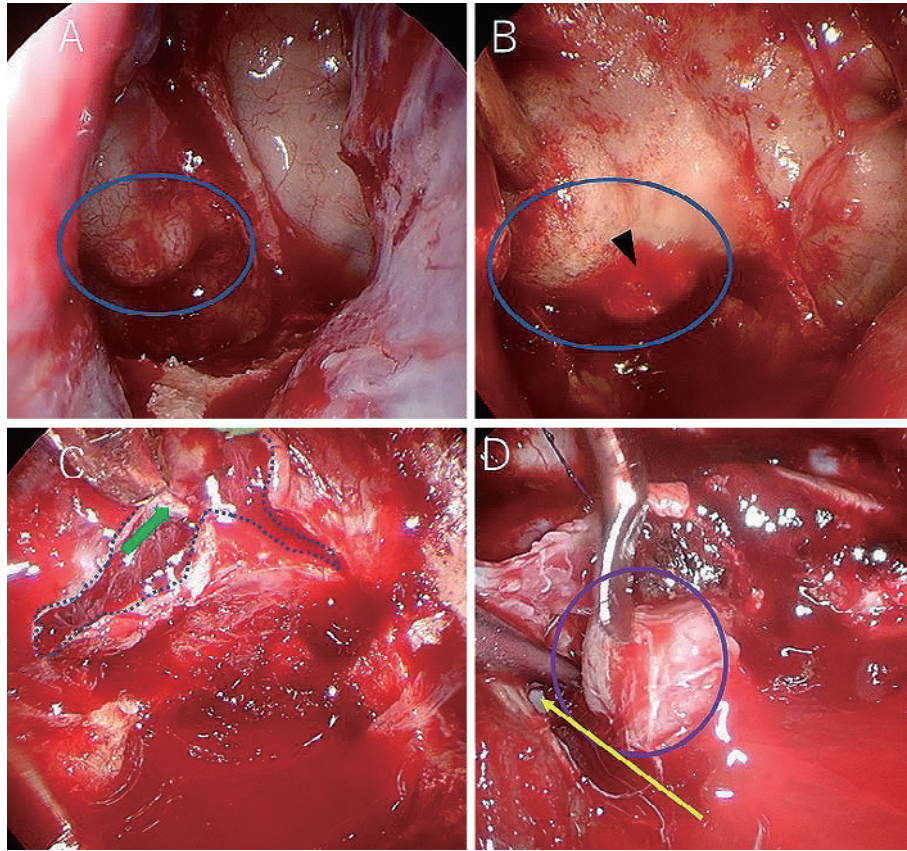


Fig. 2 Intraoperative findings.

A. View just after opening of the frontal wall of the sphenoid sinus. Vascular collection and dilatation of the sphenoid sinus mucosa (blue circle).

B. View after removal of the sphenoid sinus mucosa. Highly thickened sphenoidal bone inferior to the sellar floor (blue circle). The most reddish part inferior to the sellae was the abnormal bone structure (black arrowhead).

C. View after cutting the dura mater of the sellar floor. The blue dotted line shows the cutting edge of the dura mater. The rigid membrane appeared to be the dura mater (green arrow) separating the tumor (inferior and lateral to the membrane) and the normal pituitary gland (superior and medial to the membrane).

D. View after opening the cavernous sinus medial wall. The tumor adhered to the medial wall (purple circle). The internal carotid artery (yellow arrow) was identified.

highly thickened (Fig. 2A, B). We used a Sonopet ultrasonic aspirator (Stryker, Kalamazoo, MI, USA) for bone removal around this site to reduce the risk of internal carotid artery injury. The thickened part of the bone was sparser than the normal bone, and there was active bleeding. After removal of the sellar floor and cutting of the dura mater of the sellar floor, a normal pituitary gland was identified in the left upper side of the sellae, and an elastic soft tumor was detected on the right inferior side. The tumor and normal pituitary gland were divided by a membrane that seemed to be the dura mater (Fig. 2C). The soft part of the tumor adhered to the medial wall of the cavernous sinus and was removed along with the tumor. After removal of the medial wall, no residual tumor was observed in the cavernous sinus (Fig. 2D). We then stopped the cavernous sinus bleeding by repairing its medial wall using Surgical Nu-Knit[®] (Johnson and Johnson, New Jersey, NY, USA).

Postoperatively, the patient had no neurological or hormonal deficits, and she was discharged one week after surgery. The tumor was thought to have been completely resected (Fig. 1G-I). Histological examination revealed a dense growth with a capillary-like vascular structure and endothelial cells. Immunohistochemical staining revealed that test results for cluster of differentiation (CD) 31 and CD34 were mostly positive, but those for epithelial membrane antigen and glial fibrillary acidic protein were negative (Fig. 3). The tumor was diagnosed as CH. Additionally, this vascular structure was observed in the thickened sphenoidal bone. Without additional treatment, tumor regrowth was not detected by follow-up enhanced MRI over 1.5 years.

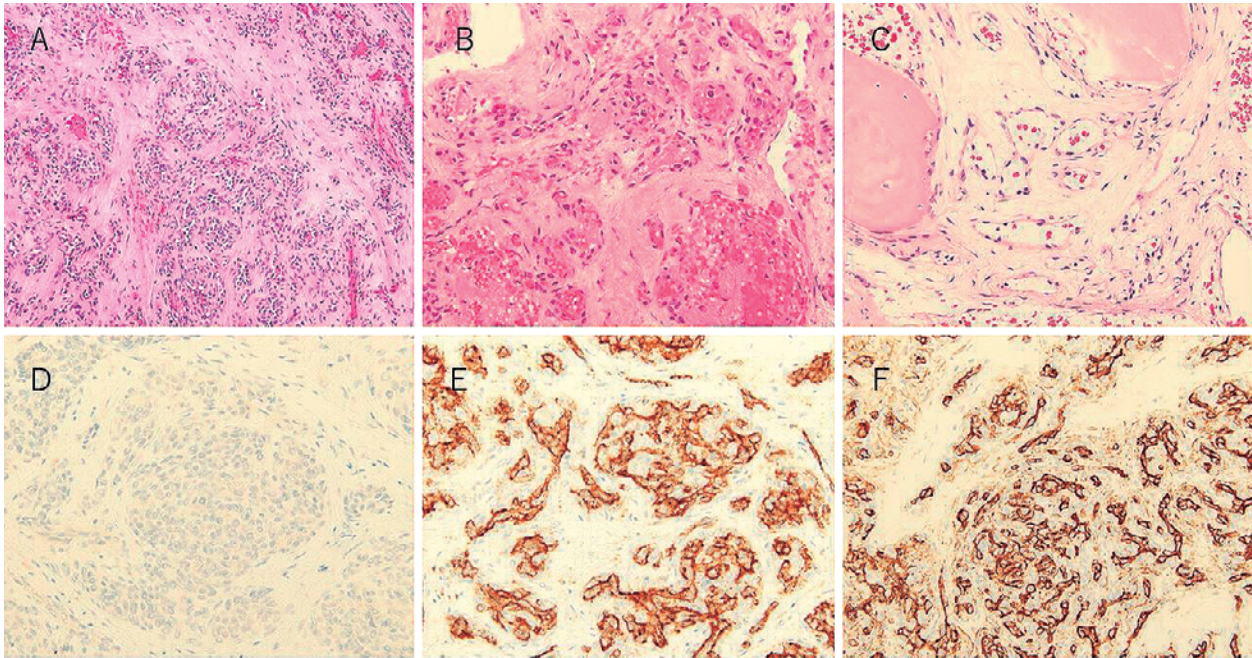


Fig. 3 Microscopic and immunohistochemical findings of the tumor.

Micrographs of the extraosseous fraction of the lesion showing numerous small vascular structures with swollen endothelial cells (hematoxylin eosin [HE] staining; original magnification, $\times 100$ (A), $\times 400$ (B)). Intraosseous fraction showing numerous thin-walled vessels occupying the trabecular bone (HE original magnification, $\times 400$ (C)). Immunohistochemical analysis showing the tumor was negative for (D) epithelial membrane antigen (EMA original magnification, $\times 400$) but positive for (E) cluster of differentiation (CD) 31 (original magnification, $\times 400$) and (F) CD34 (original magnification, $\times 400$).

Discussion

CHs are commonly found in the skin and soft tissues of infants.¹⁾ In the present case, the tumor had the following two parts: intraosseous and extraosseous-epidural. Based on the radiological images and the operative findings, we thought the tumor originated from the sphenoid bone (in the dorsum sellae) and grew mainly in the bone, and part of the tumor destroying the cortical bone grew in an epidural fashion to the parasellar and sellar regions. Twelve CHs around the sphenoid bone (sellae,²⁾ cavernous sinus,²⁻⁹⁾ sphenoid bone^{10,11)} have been reported in the literature. Apart from our case, there were no cases in which the tumor showed hyperostosis, and both intraosseous and extraosseous tumors were detected by histopathological studies in the literature.

Radiographically, ICHs need to be differentiated from meningioma, hemangiopericytoma, metastatic lesions, and other vascular malformations, including intravascular papillary endothelial hyperplasia and chondrosarcoma.¹²⁾ Daenekindt et al. reported that T2-high and homogeneous enhancement by Gd-T1 with a clear rim, multiple flow voids on MRI, intralesional hemorrhage, and absence of a dural tail or bone changes are typical findings associated with ICHs. However, some tumors show the dural tail sign¹³⁾ or bony change (erosion¹⁴⁾ or hyperostosis²⁾ like meningioma. Consequently, ICH may not be diagnosed on

radiological examination.

The most common cutaneous CHs spontaneously regress around school age. On the other hand, ICHs and CHs of skull bones rarely occur, and Abe et al. stated that ICHs showed a reduced apoptotic index compared with cutaneous CHs, and that the spontaneous involution of central nervous system CHs is unlikely.¹⁾ Therefore, many ICHs are treated with surgical resection.¹⁾ The prognosis after surgery is good, and recurrence after gross total resection (GTR) has not been reported. However, patients with residual cavernous sinus or transverse sinus tumors experience recurrence relatively early after surgery (3-6 months).¹⁵⁾ Stereotactic fractionated radiotherapy has been selected in several cases of residual tumor. Tumor control using this approach has been reported to be effective. The literature shows that few patients were followed up for <3 years, and the long-term prognosis is still unknown. Most cases that received radiation therapy were cavernous sinus cases^{6,9)} with operative difficulties in cavernous sinus surgery. Although eTSA has been performed on ICHs in and close to the cavernous sinus, it often results in a biopsy and partial removal. However, Massman et al. recently reported the first GTR of an ICH limited to the right cavernous sinus by eTSA.⁷⁾ Pas et al. also reported the resection of a CH of the sphenoid sinus intrasellar and parasellar extensions using an endoscopic transpterygoid approach.¹¹⁾ We also achieved GTR of a CH in the parasellar region using eTSA. Similar

Table 1 Summary of intracranial capillary hemangioma cases identified during the peripartum period

Author, (references)	Age	Timing of CH identified	Delivery	Location	Dura-attach	Symptoms	Symptoms' progression or relief after delivery	Treatment	Outcome
Present case	28	30 weeks of gestation	Spontaneous	Dorsum sellae	+	CN6 palsy	Relief	eTSA (GTR)	No recurrence (18 months)
Massman et al. ⁷⁾	23	5 weeks after delivery	N.A.	Cavernous sinus	+	None	Progression (complete CN3,6 palsies, hemifacial pain)	eTSA (GTR)	No recurrence (1 year)
Simon et al. ¹⁵⁾	31	38 weeks of gestation	Cesarean	Cerebellar tentorium	+	Headache, vomit	N.A. (Craniotomy was performed 1 week later after cesarean section)	Craniotomy × 3 (near total × 2, GTR × 1)	No recurrence after third surgery (41 months)
Mirza et al. ¹⁶⁾	28	A few months after delivery	N.A.	Cerebellar tentorium	+	Migraine-like headache	Progression (Secondary generalized tonic-clonic seizure, worsening headache)	Craniotomy (GTR), perioperative Dex infusion	No recurrence (1 year)
	41	3.5 months after delivery	N.A.	Convexity	+	During pregnant headache and vague visual disturbance	Progression (Transient left hemianopia)	Craniotomy (GTR), perioperative Dex infusion	No recurrence (13 weeks)
Smith et al. ¹⁷⁾	26	36 weeks of gestation	Cesarean	Middle cranial fossa	+	Headache, vomit, photophobia, confusion CN6 palsy	Relief (Dexamethasone infusion for 3 weeks)	Preoperative Dex infusion Craniotomy (GTR)	Symptom resolution (4 months)

Abbreviations N.A., data was not available; CN3: oculomotor nerve; CN6, abducent nerve; Dex, dexamethasone; eTSA, endoscopic transphenoidal approach; GTR, gross total resection

cases are expected to continue to increase in the future as the number of eTSA cases increases. However, surgery, including cavernous sinus opening or manipulation near the ICA, should be performed by surgeons accustomed to eTSA with a high-resolution endoscope, ultrasonic aspirators, precise navigation system, reliable hemostatic materials, and instruments, particularly for eTSA.

ICHs and pregnancy

There were 46 cases of ICH and 8 of CH of the skull bone. Among them, 6 cases, including the present case, were diagnosed during the perinatal period.^{7,15-17)} Table 1 shows all symptomatic cases of women during the peripartum period.

This tumor is reportedly more common in female patients and may change in size in adults according to the menstrual cycle and pregnancy. Unlike meningiomas, CHs are reported to occur due to a lack of estrogen and progesterone receptors.¹⁾ Simon et al. suggested that an increase in cardiac output and fluid retention during preg-

nancy leads to rapid tumor growth during pregnancy.¹⁵⁾ The hypervolemia associated with normal pregnancy averages 40%-45% above the non-pregnant blood volume. The maternal blood volume expands most rapidly during the second trimester and then rises at a much slower rate during the third trimester to plateau during the last several weeks of pregnancy. After delivery, the blood volume rapidly decreases to non-pregnant levels by 6 weeks postpartum. Additionally, the cardiac output of pregnant women peaks at 28-32 weeks of gestation (excluding delivery) and then declines slightly. During labor and immediately postpartum, the cardiac output temporally increases and decreases to non-pregnant levels by 6-8 weeks postpartum.¹⁸⁾ The patient experienced worsening diplopia during pregnancy and gradual remission after delivery. This clinical course correlated with the above hypothesis. A case reported by Smith et al.¹⁷⁾ demonstrated symptom relief after delivery with dexamethasone infusion. The authors reported that brain edema and mass effects were reduced after delivery. However, among the six cases with

CHs during perinatal period, three were detected after delivery, and all experienced worsening of symptoms after delivery when the blood volume and cardiac output decreased. In one case reported by Massman et al., the rapid growth of ICH after delivery was detected using MRI.⁷⁾ Furthermore, the case reported by Simon et al. was diagnosed during pregnancy, but the patient experienced recurrence twice after delivery.¹⁶⁾ Therefore, the possibility of changes in hemodynamics increasing the incidence of ICH is unlikely. In another intracranial vascular lesion, hemangioblastoma, placental growth factor and its receptor, vascular endothelial growth factor receptor 1, can lead to the development of peritumoral edema and cysts during pregnancy.¹⁹⁾ Similarly, unknown growth factors may lead to ICH growth.

Conclusions

We report the case of a pregnant woman with CH thought to have arisen from the dorsum sellae. ICH is rarely diagnosed by radiological examination. Therefore, in most cases, surgical intervention is necessary for histological diagnosis and/or control of the mass effect in large lesions. The eTSA is effective for ICHs that are limited to the sellar region. ICHs may sometimes grow peripartum.

Conflicts of Interest Disclosure

The authors do not have any disclosures or conflicts of interest to declare.

References

- 1) 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* 109: 151-158, 2005
- 2) Morace R, Marongiu A, Vangelista T, et al.: Intracranial capillary hemangioma: a description of four cases. *World Neurosurg* 78: 191.E15-191.E21, 2012
- 3) Nawashiro H, Osada H, Wada K, Minamimura K: Intermittent diplopia and periorbital pain due to capillary hemangioma. *Intern Med* 50: 2065, 2011
- 4) Saliba I, El Fata F, Berthelet F, Moumdjian R: Trigeminal nerve haemangioma eroding the petrous carotid canal. *J Laryngol Otol* 123: 1258-1261, 2009
- 5) Phi JH, Kim SK, Cho A, et al.: Intracranial capillary hemangioma: extra-axial tumorous lesions closely mimicking meningioma. *J Neurooncol* 109: 177-185, 2012
- 6) Tsao MN, Schwartz ML, Bernstein M, et al.: Capillary hemangioma of the cavernous sinus report of two cases. *J Neurosurg* 98: 169-174, 2003
- 7) Massman LJ, Conteh F, Cochran EJ, Zwagerman NT: Intracranial capillary hemangiomas: a peripartum presentation and review of the literature. *World Neurosurg* 145: 220-228, 2021
- 8) Low JCM, Maratos E, Kumar A, King A, Al-Sarraj S, Barazi S: Adult parasellar capillary hemangioma with intrasellar extension. *World Neurosurg* 124: 184-191, 2019
- 9) Grosu AL, Nieder C: Stereotactic fractionated radiotherapy for recurrent capillary hemangioma of the cavernous sinus. *Strahlenther Onkol* 182: 179-182, 2006
- 10) Suss RA, Kumar AJ, Dorfman HD, Miller NR, Rosenbaum AE: Capillary hemangioma of the sphenoid bone. *Skelet Radiol* 11: 102-107, 1984
- 11) Pas M, Shimono T, Morisako H, et al.: Capillary hemangioma of the sphenoid sinus with intrasellar and parasellar extensions. *Radiol Case Rep* 17: 156-160, 2022
- 12) Daenekindt T, Weyns F, Kho KH, Peuskens D, Engelborghs K, Wuyts J: Giant intracranial capillary hemangioma associated with enlarged head circumference in a newborn. *J Neurosurg Pediatr* 1: 488-492, 2008
- 13) Yang G, Li C, Chen X, et al.: Large capillary hemangioma of the temporal bone with a dural tail sign: a case report. *Oncol Lett* 8: 183-186, 2014
- 14) Zheng SP, Ju Y, You C: Giant intracranial capillary hemangioma in a 3-year-old child: case report and literature review. *Clin Neurol Neurosurg* 114: 1270-1273, 2012
- 15) Simon SL, Moonis G, Judkins AR, et al.: Intracranial capillary hemangioma: case report and review of the literature. *Surg Neurol* 64: 154-159, 2005
- 16) Mirza B, Shi WY, Phadke R, et al.: Strawberries on the brain - intracranial capillary hemangioma: two case reports and systematic literature review in children and adults. *World Neurosurg* 80: 900.e13-900.e21, 2013
- 17) Smith IF, Skelton V: An unusual intracranial tumour presenting in pregnancy. *Int J Obstet Anesth* 16: 82-85, 2007
- 18) Cunningham FG, Leveno JK, Bloom LS, Spong YC, Dashe SJ: Williams Obstetrics, ed 24. New York, McGraw-Hill Medical, 2014
- 19) Nayak N, Kumar A: Cerebellar hemangioblastoma during pregnancy: management options and review of literature. *Surg Neurol Int* 11, 2020

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