

터키안 및 주변 해면정맥동, 사골동, 접형동에 발생한 용종성 혈관내피 과증식증

- 증례 보고 -

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Papillary Endothelial Hyperplasia of the Sella, Cavernous Sinus, Ethmoid and Sphenoid Sinus

- Case Report -

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Papillary endothelial hyperplasia (PEH), usually originates in extracranial soft tissue, is considered a reactive proliferation of endothelium and an unusual form of thrombus organization. However the occurrence of PEH in the cranial cavity is very rare. The authors report a case of a PEH involving intracranial sella, parasella, cavernous sinus, ethmoid and sphenoid sinus in an 11-year-old boy.

The patient was presented with sudden right visual disturbance. Magnetic resonance image showed a well-enhanced mass in the right cavernous sinus extending into the ethmoid and sphenoid sinus. The cerebral angiography demonstrated a vigorous tumor staining supplied by both internal and external carotid arteries.

The right fronto-temporal craniotomy and subtotal resection of the tumor were performed. The mass was comprised in a highly vascular red-black nodular tissue associated with a mixture of fresh and organizing thrombi. The histopathological examination revealed papillary structures lined by endothelium within vascular space with organized thrombus. While the patient was waiting for second operation in order to remove remaining small tumor in the ethmoid and sphenoid sinuses, the lesion exhibited rapid regrowth and the size became to the preoperative size at three months after operation.

At reoperation, gross total removal was accomplished by a combination of trans-cranial and trans-sphenoidal approaches. Postoperative radiotherapy was given.

KEY WORDS : Papillary endothelial hyperplasia · Intracranial · Gross total removal · Radiotherapy.

서론 (organizing thrombi) (angiosarcoma) 가
(papillary endothelial hyperplasia, PEH) PEH ,
1,6) , PEH 1
13)

중례

(5×3×3cm)가

11

, T₂

, T₁

(Fig. 1).

(finger count) 15cm 가

(Fig. 2).

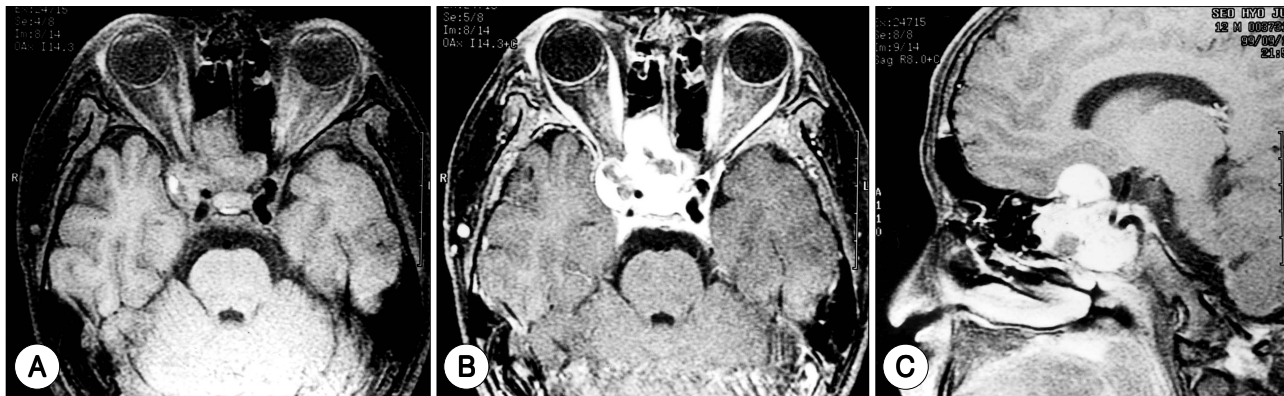


Fig. 1. Preoperative magnetic resonance image. A : Axial T1 weighted magnetic resonance image showing isointense to slightly hypointense signal in sellar, parasellar, sphenoid sinus. B, C : Contrast axial and sagittal magnetic resonance images showing strongly enhancing large mass.

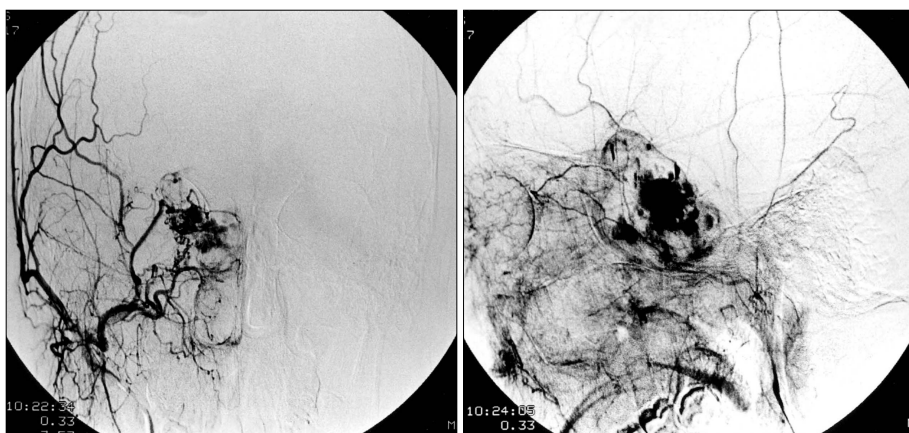


Fig. 2. Right external carotid angiography demonstrates a tumor blush.



Fig. 3. Magnetic resonance images at three months after the first operation. The mass was extended into the sellar, parasellar regions, sphenoid sinus and ethmoid sinus. Right cavernous sinus was encircled by the mass with invasion of leptomeninges.

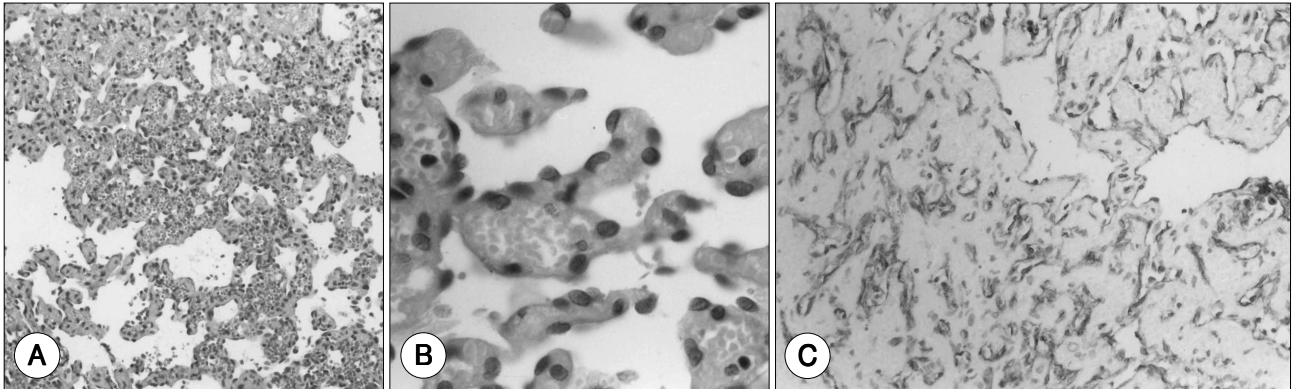


Fig. 4. A : Numerous papillary projections with fibrin core, lined by thin endothelial cells(H-E, original magnification $\times 100$). B : Plump endothelial cells lacking pleomorphism or mitosis(H-E, original magnification $\times 400$). C : An immunohistochemical preparation using the antibody CD34 highlights the complex vascular network(original magnification $\times 200$).



Fig. 5. Magnetic resonance images at 12 months after the second operation. There was no evidence of remnant tumor. Cavernous sinus and sphenoid sinus was filled with muscle flap graft.

(trans - nasal biopsy)

(right fronto - temporal craniotomy and extradural approach)

(5500cGy)

(endothelial cell)

(Fig. 4). 12

가 가

(Fig. 5).

가 (trans - sphenoidal approach)

고 찰

(papillary endothelial hyperplasia, PEH) 1923 Pierre Masson¹¹⁾

(hemorrhoidal plexus) (heman-gioendotheliome vegetant intravasculaire)

Intravascular angiomatosis, Masson 's pseudoangiosarcoma, Masson 's hemangioma^{1,6,7)}

(musculi rectus abdominis)

(superficial temporal artery and vein)

(Fig. 3)

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3

(organized thrombi) 가 3 (extravascular form) , 1993 56%, 40%, 4% PEH 320 가 13) 2% DNA flow cytometric analysis endothelial basic fibroblast growth factor autocrine loop 가 가 (M : F = 1 : 1.2). 1983 Hashimoto 91 PEH 3 가 2cm 6) 12 가 3,5,9) PEH가 가 PEH (fibrous pseudocapsule) (extravascular) (angioma), (arteriovenous malformation), (pyogenic granuloma), (lymphangioma), (vascular ha-

Table 1. Clinical feature of reported intracranial PEH

Authors	age/sex	Clinical presentation	Underlying vascular malformation	Surgery	Course
Najib et al.(1982)	16 yrs/F	Neurocutaneous disseminated form, seizure	-	Subtotal	Local recurrence 19 months later
Chen et al.(1984)	3.5 mos/F	Signs of raised intracranial pressure, seizure	-	Biopsy	Died 6 months later
Izukawa et al.(1987)	55 yrs/F	Hemianopsia, sensory dysphasia, hemiparesis, seizure	Cavernous hamangioma	Total	No follow up
Sickler and Lanford.(1990)	12 days/F	Signs of raised intracranial pressure	-	Subtotal	Recurrence 2 months later, treated by chemotherapy
Wen et al.(1991)	15 yrs/F	Signs of raised intracranial pressure	-	Subtotal	No progression for 6 months
Patt et al.(1992)	27 yrs/F	Unilateral deficit of cranial nerves , , , headache	Venous angioma	Total	No recurrence for 6 months
Tsuji et al.(1994)	18 yrs/F	Seizure, hemiparesis	Arteriovenous malformation	Total	No recurrence for 2 yrs
Kristof et al.(1997)	70 yrs/F	Transient diplopia	-	Subtotal	Recurrence 3 months later, treated by irradiation
	51 yrs/M	Diplopia	-	Subtotal	No progression
	24 yrs/F	Diplopia	Cavernoma	Total	No progression for 4 months
Duong et al.(1997)	51 yrs/F	Signs of raised intracranial pressure, visual field defect	-	Total	No recurrence for 6 months
Hagiwara et al.(1999)	29 yrs/F	Headache, gait disturbance	-	Total	No recurrence for 21 months
Present case(2000)	11 yrs/M	Visual impairment	-	Subtotal, complete removal 3months later	Recurrence 3 months later, treated by reoperation

Table 2. Radiological findings of reported intracranial PEH

Authors	Age/sex	CT		MRI			Angiography
		Non-enhanced	Enhanced	T1WI	T2WI	Enhanced	
Nagib et al.(1982)	16 yrs/F	-	Marked, homogeneous	-	-	-	Avascular
Chen et al.(1984)	3.5 mos/F	Mildly high	Marked, homogeneous	-	-	-	-
Izukawa et al. (1987)	55 yrs/F	Mixed density	Not enhanced	-	-	-	Avascular
Sickler and Lanford.(1990)	12 days/F	-	-	Isointensity	-	-	Hypervascular
Wen et al.(1991)	15 yrs/F	Not detected	-	-	High intensity	-	Hypervascular
Patt et al.(1992)	27 yrs/F	-	Marked, homogeneous	-	-	-	Hypervascular
Tsuji et al.(1994)	18 yrs/F	High (hematoma)	Not enhanced	-	High intensity	-	Avascular
Kristof et al.(1997)	70 yrs/F	-	-	Isointensity	-	Marked, homogeneous	Hypervascular
	51 yrs/M	-	-	Isointensity	High intensity	Marked homogeneous	Avascular
	24 yrs/F	Mildly high	Mildly enhanced	-	High intensity	Slightly enhanced	-
Duong et al.(1997)	51 yrs/F	Mildly high	Marked, homogeneous	Isointensity	-	Marked homogeneous	-
Hagiwara et al.(1999)	29 yrs/F	Mildly high	Marked, homogeneous	Low intensity	High intensity	Marked homogeneous	-
Present case(2000)	11 yrs/M	High	Marked, homogeneous	Isointensity	High intensity	Marked homogeneous	Hypervascular

T1WI, T1-weighted image ; T2WI, T2-weighted image ; (-), not done or not described

, 3 , 4 , 가 PEH 2 , 12 10 . PEH 4 , 3 . (Table 2), 7 4 (neurocu- , 8 5 taneous) PEH 1 가 12). , 2 . PEH 가 T₁ , PEH 가 T₂ 5 4 T₁ , PEH 가 PEH 가 8 4 , 4 6 가 5 3,7,9,13) 가 6 PEH 4 2 19 1 12) 가 PEH (angiomatous meningioma), 4 2 9,16) (angiosarcoma) (vascular malformation), (vascular) , (avascular) (cavernous hemangioma), 1 12) (arteriovenous malformation), (capillary telangiectasia) ,

3

3

PEH

가

결론

PEH

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PEH

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