www.jkns.or.kr

J Korean Neurosurg Soc 42 : 129-131, 2007

Case Report

Seok Kon Yeo, M.D.
Jeong Hoon Kim, M.D.
Chang Jin Kim, M.D.
Jung Kyo Lee, M.D.

Intracranial Epithelioid Hemangioendothelioma

Intracranial epithelioid hemangioendothelioma is extremely rare. We report a case of intracranial epithelioid hemangioendothelioma which developed in a 55-year-old man who presented with dysarthria for two weeks. The brain computed tomography scan and magnetic resonance image showed masses which had fat component at the left frontal convexity and at left posterior parietal area. Excisional biopsy at the left frontal convexity confirmed epithelioid hemangioendothelioma which is immunopositive for CD31, supporting endothelial differentiation, and negative for CD68, SMA and HMB-45.

KEY WORDS : Epithelioid hemangioendothelioma · CD31.

Department of Neurological Surgery, Asan Medical Center College of Medicine University of Ulsan Seoul, Korea

INTRODUCTION

The term epithelioid hemangioendothelioma (EHE) was proposed by Weiss and Enzinger to nominate an unique vascular tumor with an epithelioid appearance[20]. The EHE was initially believed to be a bronchiolo-alveolar tumor but was later confirmed to be of vascular origin[3]. The EHE is well-differentiated endothelial tumor with variable presentation and behavior and can occur at various sites, such as the lungs, liver, bones, head and neck, and vessels[1,6,10]. Central nervous system involvement is extremely rare (<0.02% of all brain tumors) [5,11,13,16,19].

CASE REPORT

We report a case of intracranial EHE in a 55-year-old man who presented with dysarthria for 2 weeks. The brain computed tomography (CT) scan and magnetic resonance image (MRI) showed fat containing masses with enhancing component at the left frontal convexity and at left posterior parietal (Fig. 1).

Craniotomy and grossly total resection of the left frontal mass was performed. Removed mass consisted of, a piece of pinkish soft tissue, measured 3.5×3×2 cm and the cut surface was pinkish yellow and hemorrhagic. Histologically, the atypical cells forming solid nests were scattered within the nodular mass containing fatty tissue and abnormal blood vessels. The atypical cells were immunopositive for CD31, supporting

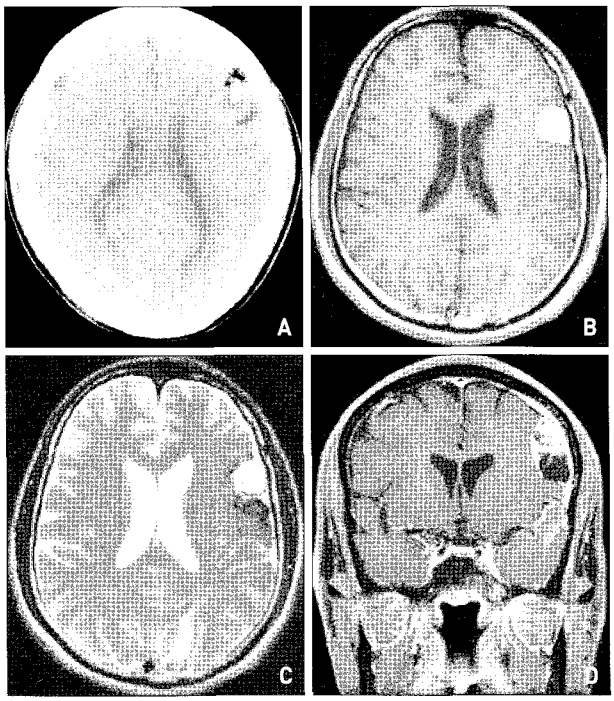

Fig. 1. Brain computed tomography scan revealing a 3×2 cm sized mass (A). Magnetic resonance image shows a fat containing extraaxial mass with enhancing component at the left frontal convexity and another small well-enhancing nodular lesion at the left posterior parietal (precuneus) (B, C, D).

• Received : March 28, 2007
• Accepted : July 6, 2007
• Address for reprints :
Jeong Hoon Kim, M.D.
Department of Neurological Surgery
Asan Medical Center, College of Medicine, University of Ulsan
Pungnap 2-dong, Songpa-gu
Seoul 138-736, Korea
Tel : +82-2-3010-3550
Fax : +82-2-476-6738
E-mail : jhkim1@amc.seoul.kr

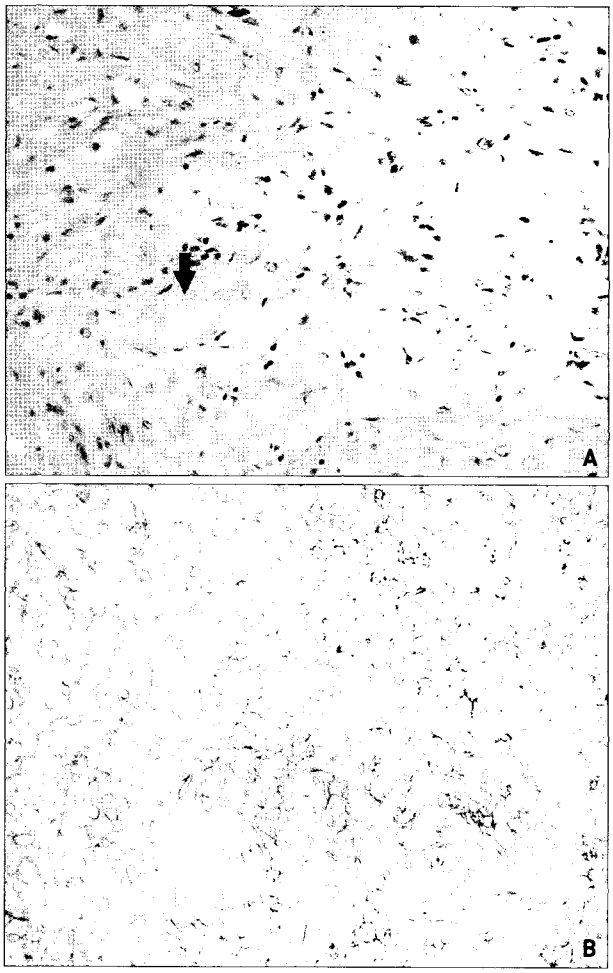

Fig. 2. The atypical cells forming solid nests are scattered within the nodular mass, which contain abnormal blood vessels (A H&E×400). The atypical cells are immnopositive for CD31, supporting endothelial differentiation, and negative for CD68, SMA and HMB-45 (B CD31 staining). Based on the histologic findings (epithelioid feature and intracytoplasmic vacuoles containing intraluminal RBCs (arrow)) and immunohistochemical staining (CD31 positivity), epithelioid hemangioendotheliom is most likely although this tumor could not be accurately classified under the current classification of the vascular neoplasm.

endothelial differentiation, negative for CD68, representing histiocytic marker, and negative for SMA and HMB-45, supporting angiomyolipoma. Based on the histologic findings and immunohistochemical staining, EHE was most likely although this tumor could not be accurately classified under the current classification of the vascular neoplasm (Fig. 2).

DISCUSSION

Central nervous system involvement of EHE is extremely rare (<0.02% of all brain tumors) but may occur in several different ways, as EHE can arise from the brain, dura matter, crainum, and spine[5,11,13,16,19].

The differential diagnosis of EHE includes a chordoid meningioma, chordoma, myxoid chondrosarcoma, metastatic carcinoma, hemanigoma, angiosarcoma, chordoid glioma, yolk sac tumor, hemangiopericytoma and hemanigoblastoma. Immunohistochemical profiling is useful because chordoma is immunopositive for S100 and cytokeratin, chordoid glioma stains for glial fibrillary acidic protein, and meningioma is at least focally immunoreactive with epithelial membrane antigen. Diffuse staining of epithelioid cells for endothelial cell markers, such as CD31, factor VIII-related antigen, or CD34, shows the endothelial nature of the neoplasm[2,7,8,13,16].

EHE can occur at various sites, such as the lungs, liver, bones, head and neck, and vessels[1,6,10]. Therefore, it is important to survey not only brain but also the skeleton, the visceral organs to determine the full extent of the disease.

Treatment options and prognosis are still controversial. Also, unlike other aggressive vascular tumors (such as hemangioendothelial sarcoma or angiosarcoma), the histological grading system is not useful for predicting prognosis[1,10]. If EHE is completely removed, it is associated with a favorable prognosis and adjuvant therapy is not needed[9]. Radiation therapy, which might be used in an attempt to sclerose the blood vessels, was attempted to restrain growth of incompletely removed tumors[8,9,15,17]. The benefits of pre- and postoperative irradiation of EHE are uncertain when total resection is possible[4,9,13,16]. Treatment with α-interferon became a commonly used modality capable of controlling the growth in most of the cases[4,9,14,18]. Therefore, radiation therapy and/or α-interferon have been recommended for partially excised or surgically inaccessible lesion.

We report a case of epithelioid hemangioendothelioma in the left frontal lobe. We performed grossly total resection of the left frontal mass and will surgically remove mass at left posterior parietal.

CONCLUSION

Intracranial epithelioid hemangioendothelioma extremely rarely develops in central nervous system. Biopsy confirmed epithelioid hemangioendothelioma which is immunopositive for CD31, supporting endothelial differentiation, and negative for CD68, SMA and HMB-45.

References

1. Adler B, Naheedy J, Yeager N, Nicol K, Klamar J : Multifocal epithelioid hemangioendothelioma in a 16-year-old boy. **Pediatr Radiol 35** : 1014-1018, 2005
2. Baehring JM, Dickey PS, Bannykh SI : Epitheloid Hemanigoendothelioma of the Suprasellar Area : A Case Report and Review of the Literature. **Arch Pathol Lab Med 128** : 1289-1293, 2004
3. Bhagavan BS, Dorfman HD, Murthy MS, Eggleston JC : Intravascular bonchiolo-alveolar tumor (IVBAT) : a low-grade sclerosing epithelioid angiosarcoma of lung. **Am J Surg Pathol 6** : 41-52, 1982
4. Chen TC, Gonzalez-Gomez I, Gilles FH, McComb JG : Pediatric intracranial hemangioendotheliomas : case report. **Neurosurgery 30** :

410-414, 1997
5. Chow L, Chow W, Fong DT : Epithelioid hemangioendothelioma of the brain. **Am J Surg Pathol 16** : 619-625, 1992
6. Dial DH, Liebow AA, Gmelich JT, Friedman PJ, Miyai K, Myer W, et al : Intravascular, bronchiolar, and alveolar tumor of the lung (IVBAT) : an analysis of twenty cases of a peculiar sclerosing endothelial tumor. **Cancer 51** : 452-454, 1983
7. Elias KR, Ryan CK : Epithelioid hemangioendothelioma and the elusive vacuole. **Liver Transpl 9** : 310-312, 2003
8. Fryer JA, Biggs MT, Katz IA, Brazier DH, Shakespeare TP : Intracranial epithelioid hemangioendothelioma arising at site of previously atypical meningioma. **Pathology 30** : 95-99, 1998
9. Hamlat A, Casallo-Quilliano C, Saikali S, Lesimple T, Brassier G : Epithelioid hemangiendothelioma of the infundibular-hypothalamic region : case report and literature review. **J Neurooncol 67** : 361-366, 2004
10. Kleer CG, Unni KK, Mcleod RA : Epithelioid hemangioendothelioma of bone. **Am J Surg Pathol 20** : 1301-1311, 1996
11. Kubota T, Sato K, Takeuchi H, Handa Y : Successful removal after radiotheraphy and vascular embolization in a huge tentorial epithelioid hemangiendothelioma : a case report. **J Neurooncol 68** : 177-183, 2004
12. Miettinen M, Lindenmayer AE, Chaubal A : Endothelial cell markers CD31, CD34, and BNH9 antibody to H- and Y-antigens- evaluation of their specificity and sensitivity in the diagnosis of vascular tumors and comparison with von Willebrand factor. **Mod Pathol** 7 : 82-90, 1994
13. Nora FE, Schelthauer BW : primary epithelioid hemangioendothelioma of the brain. **Am J Surg Pathol 20** : 707-714, 1996
14. Palmieri G, Montella L, Martignetti A, Bianco AR : Interferon alpha-2b at low dose as long-term antiangiogenic treatment of a metastatic intracranial hemangioendothelioma : a case report. **Oncol Rep** 7 : 145-149, 2000
15. Puca A, Meglio M, Rolle M, Zannoni GF : Intracranial epithelioid hemangioendothelioma : case report. **Neurosurgery 38** : 399-401, 1996
16. Rushing EJ, White JA, D'Alise, Chason DP, White CL III, Bigio EH : Primary epithelioid hemangioendothelioma of the clivus. **Clin Neuropathol 17** : 110-114, 1998
17. Tammam AG, Lewis PD, Crockard HA : Cerebello-pontine angle epithelioid hemangioendothelioma in a 4-year-old boy. **Childs Nerv Syst 13** : 648-650, 1997
18. Tancredi A, Puca A, Carbone A : Multifocal cerebral hemanigo-endothelioma : case report and review of the literature. **Acta Neurochir (Wien) 142** : 1157-1164, 2000
19. Taratuto AL, Sevlever ZG, Saccoliti SM : Epithelioid hemangioendothelioma of the central nervous system. **Pediatr Neurosci 14** : 11-14, 1988
20. Weiss SW, Enzinger FM : Epithelioid hemangioendothelioma : a vascular tumor often mistaken for a carcinoma. **Cancer 50** : 970-981, 1982