

Supratentorial Leptomeningeal Hemangioblastoma

— Case Report —

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— Abstract —

Hemangioblastoma is a benign tumor that most commonly occurs in the cerebellum and associated with von Hippel-Lindau (VHL) disease. Supratentorial hemangioblastomas are exceptionally rare. We describe the magnetic resonance imaging (MRI) and histopathologic findings of a supratentorial leptomeningeal hemangioblastoma.

Key Words: Hemangioblastoma, Brain, neoplasms, MRI

Introduction

Hemangioblastomas (HBLs) are the histologically benign tumor that commonly occurs in the cerebellum. The presentation is usually sporadic, but 10~20% manifest as a component of autosomal dominant familial transmitted von Hippel-Lindau (VHL) disease characterized by retinal angiomas, pheochromocytoma, as well as renal, pancreatic, and bone cysts. HBLs rarely occur in the supratentorium. Only 114 supratentorial hemangioblastomas were reported from 1902

to 2000.¹⁾ Supratentorial hemangiomas arise in the intraparenchyma, intraventricular, meningeal and suprasella area.²⁻⁴⁾ It has been established that the clinical symptom and signs are due to mass effect and peritumoral edema.^{5, 6)} Magnetic resonance imaging (MRI) is the most useful method in diagnosis of posterior fossa HBLs. However, only a few cases of supratentorial HBLs with MRI findings have been reported. We describe the magnetic resonance imaging (MRI) and histopathologic findings of a supratentorial leptomeningeal HBLs.

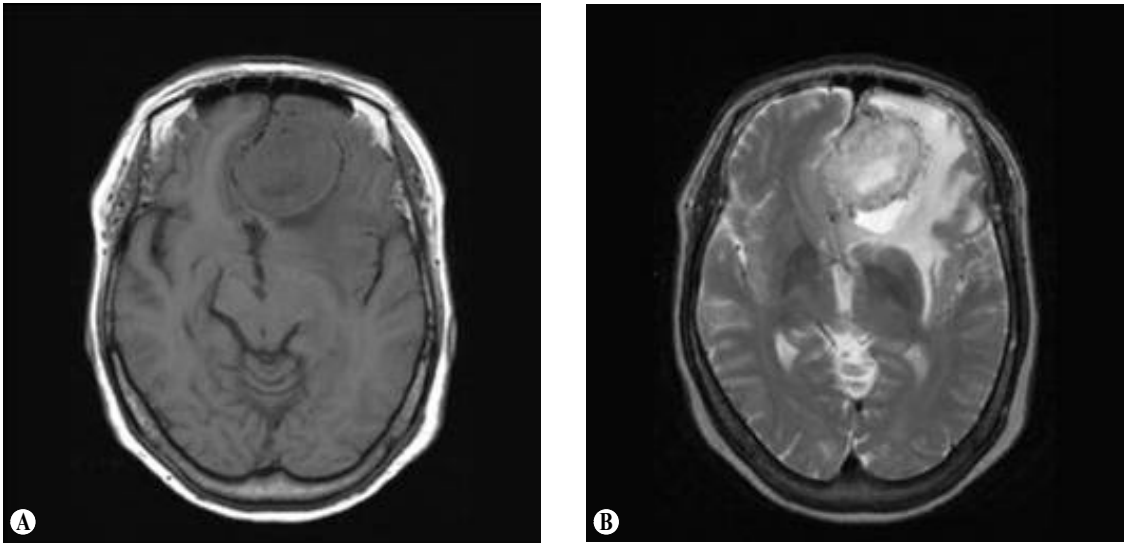


Fig. 1A, 1B. T1- and T2-weighted magnetic resonance images of the mass. A solid mass is noted in left frontal lobe with displacement of anterior falx and peripheral serpentine vascular structure. Note that the mass has a cystic portion and massive peritumoral edema.

Case Report

A 68-year-old woman who had a recent memory loss for two months was admitted to Yeungnam University hospital. Her hemoglobin level and hematocrit level were 10.7 g/dl and 33.3%, respectively. Her platelet count was 1,000 K/uL. On magnetic resonance imaging (MRI), T1- and T2-weighted images showed a solid mass with multifocal cystic portion and peripheral signal voids in the left frontal convexity. Edema and displacement of anterior falx by the mass were noted (Figs. 1A and 1B). This mass and adjacent dura revealed strong enhancement after contrast study (Fig. 1C). A gross findings of this mass revealed a clear margin and had a plenary vascularity. Microscopic findings demonstrated proliferation of stromal cells,

which have pleomorphic nuclei with abundant vacuolated cytoplasm between florid vascular networks (Fig. 2A). Immunohistochemical stainings for the specimen were done.

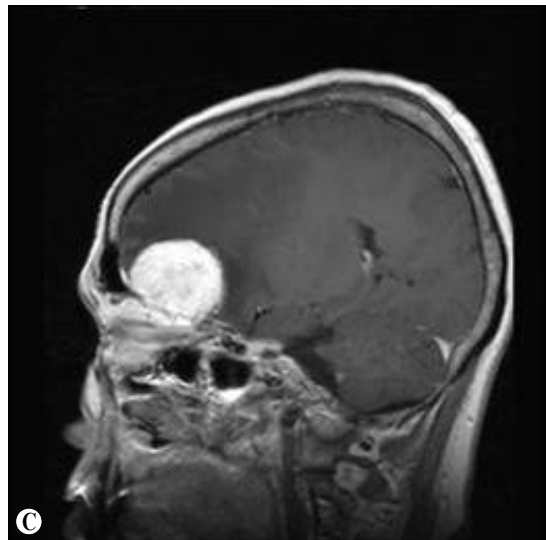


Fig. 1C. Contrast study shows marked enhancement of the mass. Noted that adjacent dural enhancement (“dural tail” sign).

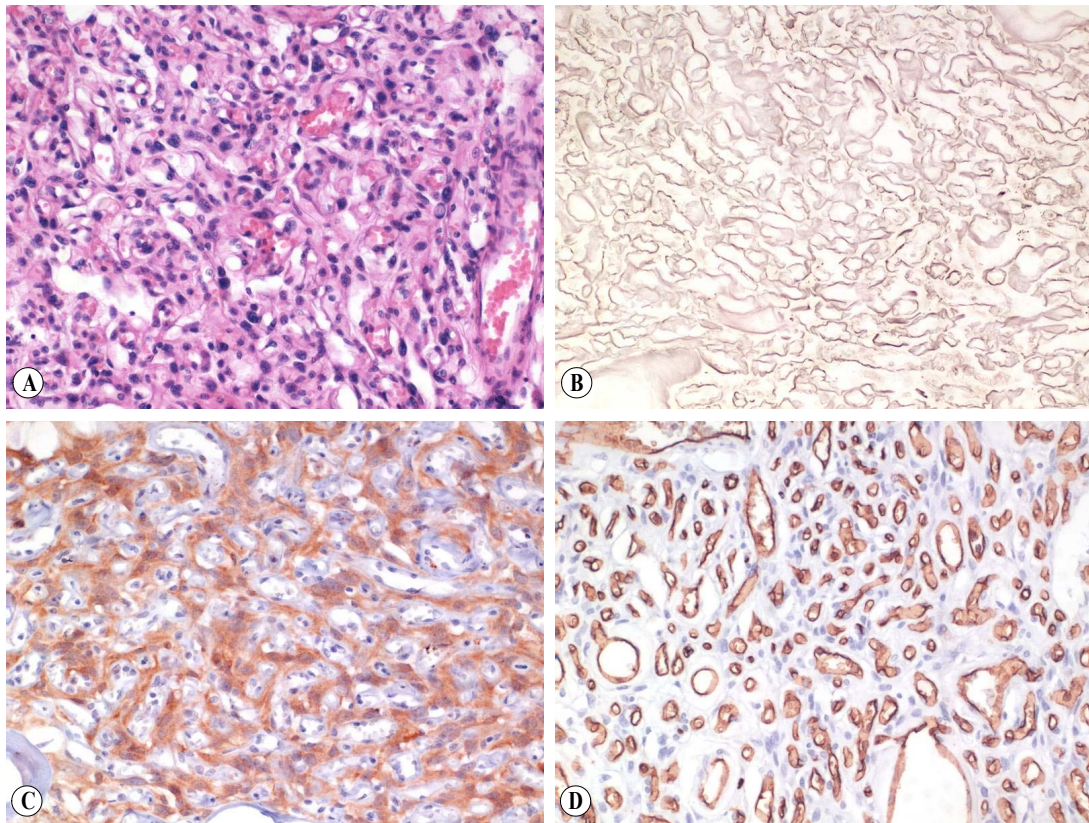


Fig. 2. Microscopic finding and immunohistochemical staining of the tumor.

- A. Tumor consists of stromal cells with multiple vascular channels (x200, hematoxylin stain).
- B. Reticulin staining reveals complex vascular channels (x200, reticulin stain).
- C. Tumor cells show diffuse immunoreactivity for neuron specific enolase (x200, immunohistochemical stain).
- D. Anti-CD34 highlights endothelial cells lining capillary-sized vessels (x200, immunohistochemical stain).

Reticulin staining revealed complex vascular channels (Fig. 2B). Tumor cells showed diffuse immunoreactivity for neuron specific enolase (Fig. 2C). Anti-CD34 highlighted endothelial cells lining capillary-sized vessels (Fig. 2D). These microscopic findings and immunohistochemical stainings were characteristic of HBLs. Follow-up MRI of the patient 1year after the operation showed no residual mass.

Discussion

HBLs of the central nervous system are the most common tumor in VHL disease, affecting 60~80% of all patients and can occur as a sporadic entity.⁷⁾ Multiple HBLs occur in 20% of the patients with VHL disease and in 5% of sporadic patients.⁸⁾ HBLs are most commonly located in the cerebellum

and other locations can be spinal cord and medulla oblongata. They also originate in supratentorium such as frontoparietal, parasagittal leptomeninges, posterior temporal lobe, lateral ventricle and suprasellar region.^{2-4, 9, 10)} HBLs are subdivided into four groups: simple cystic, macrocystic, solid, and microcystic solid types.⁵⁾

The radiological findings on computed tomography (CT) and MRI of posterior fossa HBLs were well described. The key features of posterior fossa HBLs are solid or cystic mass with mural nodule. Pathological vessels usually appear as serpentine signal voids on MRI. Few cases of supratentorial HBLs have been described. Case report that describes MRI findings of the supratentorial HBLs show no major difference from infratentorial HBLs.^{2-4, 6, 8)} The differential diagnosis of a cyst with mural nodule in cerebral hemispheres are a benign astrocytoma, ependymomas, choroids plexus papillomas and meningiomas.⁶⁾

The histologic findings that suggest the HBLs are the presence of large numbers of thin-walled, fairly closely packed blood vessels and intervening polygonal stromal cells. The stromal cells are characterized by a pale and finely vacuolated cytoplasm.⁶⁾ These neoplastic stromal cells are unknown origin and accompanied by intensive angiogenesis.⁷⁾ The main histopathologic differential diagnoses are angioblastic meningioma, metastatic renal cell carcinoma and hemangiopericytoma. The appropriate

immunohistochemical stains helps to confirm the correct diagnosis. The stromal cells of HBLs show immunoreactivity for neuron-specific enolase, vimentin, and S100 protein and do not react to epithelial membrane antigen and glial fibrillary acidic protein.

In HBLs, CD34 immunohistochemical reactivity is typically strong and diffuse in contrast to the more focal heterogeneously weaker staining of tumor cells in fibrous meningioma and hemangiopericytoma. Reticulin staining demonstrates the complex capillary networks of HBLs.^{4, 6)}

In summary, HBLs must be considered if we find supratentorial solid or cystic mass with mural nodule and immunohistochemical stains should be done to make the diagnosis clear.

요 약

혈관아세포종은 소뇌에 가장 흔하게 발생하며 von Hippel-Lindau disease와 연관될 수 있다. 저자들은 천막상부 연수막에서 기원한 혈관아세포종의 자기공명영상과 병리학적 소견에 대해서 기술하고자 한다.

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