Hemangiopericytoma in the Trigone of the Lateral Ventricle

—Case Report—

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Abstract

A 40-year-old male presented with hemangiopericytoma in the lateral ventricle manifesting as headaches persisting for 6 months associated with vomiting and visual obscurations for one month. Computed tomography and magnetic resonance imaging of the brain showed a large tumor in the trigone of the right lateral ventricle. The highly vascular tumor was completely excised. The histological diagnosis was hemangiopericytoma. Hemangiopericytoma is rarely located in the lateral ventricle and is difficult to differentiate from meningioma by neuroimaging methods.

Key words: hemangiopericytoma, meningioma, trigone, lateral ventricle

Introduction

Hemangiopericytoma was first described as a subtype of angioblastic meningioma in 1928.³⁾ A similar lesion was described as a vascular variant of a meningioma derived from meningotheial cells with angioblastic features.⁸⁾ The term “hemangiopericytoma” was first used for malignant vascular tumor consisting of cells resembling capillary pericytes (Zimmerman’s pericytes) in the soft tissue of thigh, buttock, and retroperitoneum.²³⁾ The first case of primary intracranial hemangiopericytoma was reported in 1954.⁴⁾

Intracranial hemangiopericytomas are rare tumors that resemble meningiomas but behave more aggressively, with a tendency to metastasize and a higher propensity for recurrence.¹¹⁾ Intracranial hemangiopericytoma accounts for 2% of all meningiomas, are more common in males, and 56–75% of cases occur between 38–42 years of age.¹⁾ Most cases are located in the supratentorial compartment and less frequently in the posterior fossa or spine.^{1,10)}

Other very rare sites are the sella (3 cases),^{12,16,18)} pineal region (3 cases),^{14,21,22)} lateral ventricle (1 case),¹⁹⁾ and third ventricle (1 case).¹⁾

Here we describe a case of hemangiopericytoma located in the trigone of the lateral ventricle.

Case Report

A 40-year-old male presented with a 6-month history of headaches, which were characteristically dull, aching, generalized, and intermittently throbbing in nature, and associated with features of raised intracranial pressure like vomiting and visual obscurations for the last month. Neurological examination revealed bilateral papilledema and left homonymous hemianopsia. No other central nervous system abnormalities were found including the whole spinal cord.

Computed tomography (CT) revealed a large, hyperdense, homogeneously enhanced mass located in the trigone of the right lateral ventricle, with dilation of the contralateral lateral ventricle (Fig. 1). Calcification was noted within the tumor. Magnetic resonance (MR) imaging showed the mass in the trigone of the right lateral ventricle as 7 cm in

maximum diameter. The tumor was isointense on T₁-weighted images (Fig. 2A) and was hypointense on T₂-weighted images (Fig. 2B). After administration of gadolinium, the tumor showed intense homogeneous enhancement (Fig. 2C). Cerebral angiography was not performed.

The tumor was approached by a transcortical route through the superior parietal lobule. The tumor was characteristically reddish in color and bled profusely from the beginning of the excision.

Fig. 1 Axial computed tomography scan showing a massive right trigonal mass, with dilation of the contralateral ventricle.

Profuse bleeding occurred from the numerous blood vessels within the tumor during the piecemeal excision. The tumor was supplied by the branches of the anterior and posterior choroidal arteries. The arterial feeders to the tumor arose from the choroid plexus. The intraoperative impression was meningioma. The tumor was completely excised. The blood loss during the surgery was 5 liters which was adequately replaced with fresh units of whole blood, platelets, and fresh frozen plasma. No hypotension or disseminated intravascular coagulopathy occurred during the surgery secondary to the profuse blood loss and transfusion. In the immediate postoperative period the patient was conscious and alert and had left hemiparesis.

Histological examination of the tumor revealed hemangiopericytoma, with the characteristic densely cellular neoplasm of oval and spindle cells, called pericytes, with prominent slit-like vascular spaces lined by endothelial cells (Fig. 3). No whorl formation or psammoma bodies were present, and no necrosis was found in the tumor. Mitotic activity was also minimal. These findings were consistent with the diagnosis of hemangiopericytoma. Immunohistochemical staining revealed the tumor cells were positive for vimentin and CD34, but negative for epithelial membrane antigen (EMA) and smooth muscle actin. Electron microscopy of the tumor showed elongated vascular channels surrounded by stacks of tumor cells. Basal lamina of variable thickness surrounded each tumor cell (Fig. 4). The histology, immunohistochemistry, and

Fig. 2 (A) Preoperative axial T₁-weighted magnetic resonance (MR) image showing a large, isointense trigonal tumor. (B) T₂-weighted MR image showing the hypointense tumor. (C) Sagittal T₁-weighted MR image with contrast medium showing intense enhancement of the tumor.

Fig. 3 Photomicrograph of the tumor showing slit-like and elongated thin-walled staghorn vessels surrounded by compactly arranged solid cellular spindle cells of pericytic lineage. HE stain, $\times 400$.

Fig. 5 Postoperative axial T₁-weighted magnetic resonance image with contrast medium demonstrating complete excision of the intraventricular mass.

Fig. 4 Electron micrographs of the tumor showing basal lamina of variable thickness surrounding each tumor cell (A: bar = $2\ \mu\text{m}$) and two thick bands of basal lamina material (B: bar = $1\ \mu\text{m}$).

ultrastructural findings were consistent with hemangiopericytoma.

Adjuvant cranial radiotherapy was administered with a total dose of 60 Gy in 30 fractions in the immediate postoperative period. His left hemiparesis had recovered at 1-year follow up. There was no significant recovery of the left homonymous hemianopsia. Follow-up MR imaging at one year showed complete excision of the hemangiopericytoma (Fig. 5).

Discussion

The clinical features in our case were similar to any lateral ventricular tumor with headaches, features of raised intracranial pressure, and homonymous hemianopsia, which are common clinical findings in the majority of cases.¹⁹⁾ The findings of CT and MR imaging were very similar to the intraventricular meningioma, with brilliant enhancement after contrast administration. Angiography of hemangiopericytoma shows characteristic “corkscrew” vascular configurations with a long-lasting venous phase. The presence of calcification within the lesion by CT is strongly suggestive of meningioma rather than hemangiopericytoma. T₂-weighted MR imaging often shows multiple flow voids in hemangiopericytoma.^{1,17)}

Hemangiopericytoma is difficult to differentiate from meningioma by neuroimaging methods. Histology, immunohistochemistry, and electron microscopy findings are needed for the differential diagnosis.^{1,2,5,6,15)} Preoperative diagnosis of lateral ventricular hemangiopericytoma is extremely difficult and may pose a diagnostic dilemma based on the neuroimaging and gross appearances. Immunohistochemistry can differentiate hemangiopericytomas from meningiomas by showing positive staining for CD34 and vimentin.^{5,6,13)} Hemangiopericytomas show negative reaction to EMA and cytokeratin.¹³⁾ Connexin proteins form

gap junctions in human arachnoid villi and meningiomas. Therefore, connexin expression is a useful marker for the differentiation of meningioma and hemangiopericytoma.²⁾ The ultrastructure of hemangiopericytoma shows marked differences from those of syncytial and transitional meningiomas.⁵⁾ The tumor cells are plump and the cytoplasmic edges are blunt, but lack the complex cytoplasmic interdigitations and the desmosomes seen in meningiomas. Intensely electron-dense material fills the extracellular spaces, which appears as thickened basal lamina material. The plasma membranes of adjacent cells are closely opposed in some areas.⁵⁾

Hemangiopericytomas are described as arising from pericytes, which are mesenchymal cells with contractile properties similar to the smooth muscle cells located in the walls of the capillaries external to the endothelial cells. These pericytes are unrelated to either meningioma cells or the progenitor arachnoid cells. Intraventricular hemangiopericytomas probably originate from the pericytes found within the tela choroidea or the stroma of the choroid plexus.¹⁾

Intracranial hemangiopericytoma is an aggressive neoplasm with high malignant potential and incidence of recurrences.¹⁾ The principal treatment is radical or total excision, irrespective of the location in the brain, followed by radiotherapy. Survival is correlated with more aggressive primary resection and postoperative radiotherapy.^{1,9)} Hemangiopericytoma is amongst the very few primary intracranial tumors to metastasize outside of the central nervous system. The most commonly encountered sites of metastasis are bone, followed by the lung and liver, and rarely the pancreas and spleen.¹⁰⁾ The rates of distant metastasis at 5, 10, and 15 years are approximately 13%, 33%, and 64%, respectively. The median recurrence-free interval is approximately 50 months. The 5-, 10-, and 15-year recurrence rates are 65%, 76%, and 87%, respectively.¹⁰⁾ The time interval between recurrences shortens with each operation for recurrent hemangiopericytoma. Irrespective of the extent of the surgical excision, radiotherapy is strongly recommended. Radiation helps to reduce the tumor size, decrease the chance of distant metastasis, and increase the recurrence-free interval.¹⁾ Radiosurgery has been proposed as adjuvant therapy to treat selected patients with small and medium intracranial hemangiopericytomas that occur after surgery or radiation treatment.^{1,7,20)}

Our present case of hemangiopericytoma in the trigone of the lateral ventricle is extremely rare. Histological, immunohistochemical, and ultrastructural examination of lateral ventricular heman-

giopericytoma is mandatory as the neuroimaging differentiation from meningioma is difficult. Total excision is the ideal surgical option. Postoperative radiotherapy is recommended as the incidences of recurrence and distant metastases are high.

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