



# HAEMANGIOPERICYTOMA; CLINICOPATHOLOGICAL ANALYSIS OF CENTRAL NERVOUS SYSTEM

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**ABSTRACT...** Hemangiopericytoma (HPC) in central nervous system is a rare tumor, his tumor has a high recurrence rate and the characteristics of extracranial metastases. **Objectives:** To investigate the clinicopathological features, imaging features, immunohistochemical phenotype of haemangiopericytoma (HPC) of central nervous system. **Design:** Hospital based crosssectional prospective study. **Period:** From 24th October to 26th October 2012. **Setting:** First People's Hospital of Jining City, China. **Methods:** The clinical manifestations, imaging features, histopathological and immunohistochemical features were analyzed combining the review of the literature in one case of central HPC. **Results:** The Gross examination revealed the size of the tumor was 5cm × 4cm × 1.5cm; the section is gray, medium soft texture, and part of the area had capsule. The microscopic examination showed that the tumor cells were abundant and the same size, showing round, oval or short spindle shape. The cytoplasm was eosinophilic, and part of it was slightly translucent. The nuclei were ovoid, and the nucleoli were inconspicuous. A lot of capillaries lined by endothelial cells were seen in the tumor tissue, and the blood vessels were dilated like "staghorn" in some areas. Immunohistochemistry showed that tumor cells expressed Vimentin, CD34, CD99, Bcl-2, PR protein. They didn't express EMA, SMA, and S-100 protein. The proliferation index of ki-67 is about 4%. **Conclusions:** The central haemangiopericytoma is a rare tumor, having no specific clinical manifestations and imaging features. The final diagnosis requires a combination of histopathological and immunohistochemical examination, and it should be differentiated from meningioma, solitary fibrous tumor, hemangioblastoma and mesenchymal chondrosarcoma, etc.

**Key words:** Central nervous system; Haemangiopericytoma; Clinicopathological features; Immunohistochemistry

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Hemangiopericytoma (HPC) in central nervous system is a rare tumor, Stout and Murray<sup>1</sup> first reported it in 1942. This tumor has a high recurrence rate and the characteristics of extracranial metastases. Most scholars believe that the HPC previously originated in the meninges in the past few decades, but Joseph and others<sup>2</sup> confirmed HPC originated from Zimmerman peripheral cells of capillaries at the molecular level in 1995, rather than meningeal cells. These cells are a variation of the smooth muscle cells and have multiple differentiations potential. However, HPC in central nervous system has been called angioblastic meningioma. It was still divided into perivascular cell subtypes of meningiomas by the World Health Organization classification until 1997.

In recent years, the research by

immunohistochemistry and electron microscopy has found that HPC in central nervous system and meningioma are different in histology, biological behavior and treatment, prognosis, etc. WHO classification finally defines it as hemangiopericytoma in central nervous system in 2007<sup>3</sup>. This article will report one case which occurred in the left frontal lobe and diagnosed in our hospital. We will summarize its clinical manifestations, imaging features and histopathological features as follows.

## MATERIALS AND METHODS

### Clinical data

A 50-year-old female presented with headache before menstruation for more than 30 years. This pain was mainly located in the forehead but not very serious. The patient suddenly felt

glossolalia 15 hours prior to pain and she had no nausea, vomiting, coma, convulsions and or limb movement disorder. She was admitted to the hospital for treatment.

Physical examination revealed a temperature of T 36.3°C, Pulse rate 68/min, Respiration 18 times/min, BP 120/80 mmHg. The patient was conscious and she breathed smoothly. The pupils had the same size and shape, and their diameter was 3mm. The light reflex is sensitive. The neck was soft, and the heart and lung were normal. The limbs activity was good. The muscle strength, muscle tension and tendon reflexes were normal. Neurophysiological reflex was present, and the pathological reflex was also not elicited. CT scan showed that the margins of the tumor was clear. The tumor was nodular with a high density shadow, and it was significantly enhanced. No obvious abnormalities were seen in the surrounding brain tissue. MRI scan and enhanced scan (1.5) showed an irregular shaped mass presenting uneven equivalent T1 and equivalent T2 signal in the left frontal cerebral convexity. Multiple spots were seen in the long T1 and long T2 signal area, and their size was about 0.41cm×0.36cm×0.32cm.

Its boundary was clear, and its wide base lesions were connected to the skull plate. There was no obvious edema in the peripheral brain parenchyma area, and the rest of the brain parenchyma showed no abnormal signal.

There wasn't significant expansion in the ventricular system, and the midline structures were still in the middle position. After the intravenous injection of contrast agent Gd-DTPA, the lesions were significantly enhanced uniformly. The dural tail sign was visible (Figure 1), and the peripheral brain parenchyma showed no abnormal enhanced shadow. The radiologists and the clinicians diagnosed it as meningioma, and they recommended surgery. The base area of tumor located in the left frontal meninges, and the tumor adhered to the dura tightly. Separating the tumor from the surrounding brain tissue, the surgeons had resected the tumor completely and

sent it for pathological examination. The patient had recovered quickly after surgery, and she was not subjected to radiotherapy.

## Methods

The tumor specimen was fixed with 4% formaldehydesolution,goingthroughconventional paraffin embedding and slicing by 4um. After hematoxylin-eosin staining and reticular fiber staining, the glass slides were observed under the microscope. The immunohistochemistry used was Envision two-step method. The antibodies included Vimentin, CD99, BCL-2, CD34, PR, EMA, SMA, S-100 and Ki-67. All of the antibodies came from Fuzhou Maixin company. The experimental procedure was carried out according to the reagent instructions.

## RESULTS

### Macroscopy

A nodular mass, the size was about 5cm × 4cm × 1.5cm, attached to little meningeal tissue, the area of meningeal tissue was about 3.5cm×2cm. The section was gray, medium soft texture, and part of the area had capsule.

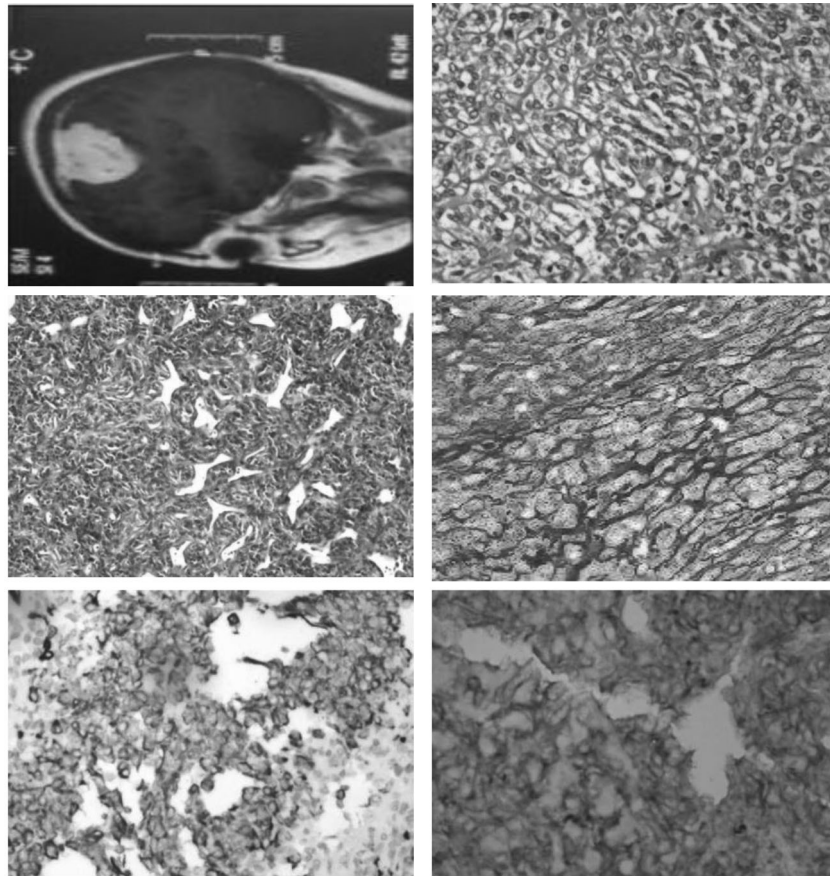
### Microscopy

The tumor cells were abundant and the same size, showing round, oval or short spindle shape. The boundary of the cell membrane was unclear, and the cytoplasm was eosinophilic, and part of it was slightly translucent (**Figure 2**). The nuclei were ovoid or short spindle shape, and the nucleoli were inconspicuous. A lot of capillaries lined by endothelial cells were seen in the tumor tissue. These blood vessel walls were very thin presenting crack-like and they were dilated like "staghorn" in some areas (**Figure 3**). Reticular fiber staining showed abundant reticular fibers between the tumor cells (**Figure 4**).

## DISCUSSION

### Clinical features

HPC in central nervous system occurs mostly in the elderly patient, and with slight preponderance in male patients. The average age of patients is



- Figure-1.** Enhancement CT scan: intravenous injection of contrast agent Gd-DTPA with visible dural tail Sign of the mass.
- Figure 2:** The boundary of the cell membrane was unclear, and the cytoplasm was eosinophilic, and part of it was slightly translucent. The nuclei were ovoid or short spindle shape, and the nucleoli were inconspicuous.
- Figure 3:** Capillaries lined by endothelial cells are seen in the tumor tissue with very thin walled blood vessels presenting a crack-like and also dilated "staghorn" – like patterns in some areas
- Figure 4:** Reticular fiber staining showing abundant reticular fibers between the tumor cells

40 to 45 years old<sup>4</sup>. The tumors are often located in the intracranial area, especially near the torcular herophili, and they are connected with the dura mostly. Few of the cases occur in the meninges, and very few occur in the brain parenchyma. The symptoms of patients are associated with tumor's location, if the tumors occur in the intracranial area, the symptoms will be headache, epilepsy and symptoms of intracranial hypertension. However, those tumors which occur in the spinal meninges, the symptoms will be muscle weakness and parasthesia. The development of the disease is different, and the process is often slow-growing, from several months to several years.

### Imaging Features

CT scan shows the tumor presenting nodular or

lobulated high density shadow which is enhanced significantly. There is obvious edema around the tumor usually. The calcification is rare, but it also can be seen in the intracranial<sup>5</sup> site. The tumor may invade or destroy the surrounding bone tissue, and it also can invade the brain tissue. The tumor may show hemorrhagic and cystic change. MRI scan showed the tumor with mixed high signal in T2W1 weighted phase, and the significantly strengthened but often uneven signal when implementing the enhanced scanning tumor in T1W2 weighted phase. The dural tail sign is rare. There are signs of vascular flow voids and signals of necrosis or cystic degeneration.

### Pathological features

The macroscopy shows the margins of the tumor

were clear; the shape was round or slightly lobulated. The section looked like the shape of fish; the colour is gray to reddish brown. The texture is tough or uneven soft and hard. The tumor is easy bleeding, and it can have necrosis and cystic changes. The microscopy showed many tumor cells and were of the same size, showing fusiform or polygonal shape, with no specific arrangement. The boundary of the cell membrane was unclear, and the cytoplasm was eosinophilic. The nuclei were ovoid or short spindle shape, and the nucleoli were inconspicuous. The nuclear atypia and mitoses can be seen. There are rich reticular fibers between cells, and the reticular fiber staining showed abundant reticular fibers between the tumor cells. These blood vessel walls are very thin presenting crack-like and they are connected like "staghorn" in some areas. The tumors can destroy the surrounding brain and bone tissue, but there is no bone hyperplasia<sup>6</sup>.

Generally speaking, the most HPC in central system is low-grade, equivalent to WHO II grade. When the tumor cells have a higher density or obvious anaplastic features and active pathological mitoses (>5/10HPF), and the proliferation index of Ki-67(MIB-1) is 5%- 10%, as well as large areas of hemorrhage and necrosis. All of these indicators have hinted a higher degree of malignancy, and it is equivalent to anaplastic HPC (WHO grades III)<sup>7</sup>. Immunohistochemistry expresses Vimentin, CD34, CD99, PR and Bcl-2. Desmin is focally positive, and SMA or CK can be positive occasionally. EMA is negative or focally positive, but GFAP and S-100 are negative.

### Ultrastructure and genetic characteristics

The electron microscopy shows the tumor cells are pleomorphic, and the tumor cells form vascular architecture closely. The cytoplasm may contain a small amount of intermediate filaments, and there is also the formation of dense body occasionally. The cell synapse is elongated. The extracellular basement membrane is protruding, which encircle around the cell presenting basement membrane-like amorphous structure. There are no complete desmosomes and gap junctions between cells<sup>8</sup>. The genetic characteristics

include the recombination of chromosome 12q13 and the mutation of 6p21, 7p15 and 19q13. There is homozygous deletion of CDKN2A gene in about 25% of the HPC of central nervous system<sup>7</sup>.

### Differential Diagnosis

- (1) Angiomatous meningioma Radiographic examination showed that the tumor was homogeneous enhancement, and calcification or bone hyperplasia was visible. As the tumor grows slowly, the dural tail sign is common. Meningiomas express EMA, but don't express CD99 and Bcl-2 in immunohistochemistry.
- (2) Solitary fibrous tumor of the central nervous system (SFT): The scope of the tumor is relatively limited, and it often have pseudocapsule.

Microscopy shows that the tumor cells are sparse and dense alternately, and it often has the deposition of bundles of collagen and thick-walled blood vessels. Both of these two tumors express CD34 and Bcl-2, but HPC express them relatively weak or only partially positive, however SFT express them diffusely<sup>9</sup>.

- (3) Hemangioblastoma: This tumor often occurs in young individuals, and to occur in certain families. It occurs mostly in the cerebellum, and it has relatively clear boundaries, rarely invading the surrounding tissues. The microscopically it shows vesicular lipid-rich interstitial cells and capillary network. Immunohistochemistry shows interstitial cells express NSE and EGFR.
- (4) Mesenchymal chondrosarcoma: When this tumor occurs outside the bone, it particularly occurs in the dura. When it lacks cartilage, the histological features of undifferentiated small round cells looks like HPC, but these tumor cells have poorly differentiation and obvious atypia. Immunohistochemistry shows these small round cells express S-100 and Leu7, so it is not difficult to identify them<sup>10</sup>.

### Treatment and Prognosis

Hemangiopericytoma (HPC) in central nervous system is an intermediate tumor and its prognosis

are not good. It has the tendency for potentially malignant change and recurrence. The interval of recurrence is associated with proliferation index. The lower index of Ki-67 will indicate that the patients with longer recurrence time, lower transfer rate and longer survival time. Through surgery and postoperative radiotherapy, we can reduce the risk of tumor recurrence and delay the recurrence time<sup>11</sup>.

The central HPC is a rare disease, and it lacks specific clinical manifestations, however, its imaging features are very similar with meningioma. Therefore, the diagnosis requires pathological examination. Because its biological behavior, histology, treatment and prognosis are different from meningioma, long-term follow-up after surgery is necessary.

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Sr. #	Author-s Full Name	Contribution to the paper	Author=s Signature
1	Dr. Henry Mwakyoma	Introduction and literature review, histological analysis, Analysis of results, Discussion and conclusion of results, Preparation of manuscript for submission	<i>Hmwky</i>
2	Dr. Xiang Longquan	Introduction and literature review, initial steps in recruitment of a case and taking of microphotographs, histological analysis, Analysis of results, Discussion and conclusion of results	<i>xianglongquan</i>