Cystic Hemangiopericytoma in the Third Ventricle

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Primary intracranial hemangiopericytoma is rare and resemble meningioma on imaging study. It shows meningeal attachment, and is usually isointense with gray matter on T1-weighted MR image with heterogeneous enhancement and prominent vascular flow voids on T2-weighted image. Cystic type of hemangiopericytoma is very rare and only 3 cases have been reported in the literature which arised in the middle fossa, cerebellum, and occipital area. Ventriclental hemangiopericytomases were reported in 9 cases, and all of them were solid type. Authors experienced a peculiar case of cystic hemangiopericytoma in the 3rd ventricle and report it with review of the literature.

KEY WORDS: Ventricle · Cystic · Hemangiopericytoma.

Introduction

The hemangiopericytoma (HPC) is a malignant tumor originating from Zimmermann’s pericytes around capillaries and postcapillary venules which is most commonly located in the musculoskeletal system and the skin.

Meningeal HPC has angioblastic tendency. It was thought to be an angioblastic subtype of a meningioma by Bailey, et al. in 1928, and termed as angioblastic meningioma by Cushling and Eisenhardt in 1938. Peripheral HPC was described in 1942 by Stout and Murray, until that time these tumors were accepted as a variant of meningiomas¹⁰. Begg and Garret reported a primary intracranial HPC in 1954. The current (2000) classification of the World Health Organization (WHO) has eliminated the term angioblastic meningioma in favor of HPC, and, HPC is accepted as distinctive mesenchymal neoplasms unrelated to meningioma⁹.

On MRI of the brain, it shows certain features suggesting HPC compared to meningioma. It appears as iso- or hypointense lesions on T1WI and hyper- or iso-intense on T2WI, with strong enhancement. Importantly, there are angioblastic characteristics which are “corkscrew” feeders, dural feeding of ICA and ECA origin⁷. Cystic change occurs in rare occasion, and may be confused with other tumor with cystic component. Authors experienced a peculiar case of cystic HPC in 3rd ventricle and reported with review of the literature.

Fig. 1. Pre-operative T1-weighted magnetic resonance images (A) showing lobulated cystic mass in the posterior portion of lateral ventricle. Post-contrast axial (B), coronal (C) and sagittal (D) images show ring enhanced cystic mass with mural nodule.
**Case Report**

A 58-year-old female patient presented with headache for 6 months. The headache progressed gradually, and recently it became aggravated on lying down and was accompanied with vomiting. She also complained decreased memory, urinary incontinence, and paraparesis (motor grade IV). On admission, neurological status was relatively well-preserved except mild disturbance in memory and gait. No history of medical or systemic disease were reported. Plain chest X-ray, electrocardiogram were normal. Complete blood count and other hematologic studies were within normal limits.

MRI of the brain revealed a cystic lesion in the 3rd ventricle (Fig. 1). It was $3 \times 3$ cm in size, and majority of the mass was cyst showing thin-walled ring-enhancement with intensely enhancing solid portion, which was $0.5 \times 0.5$ cm in size and noted on the left part of the tumor. Four-vessel cerebral angiography revealed no gross abnormalities in the cerebral vessels.

The operation was performed by anterior transcaldesal approach. When the corpus callosum was incised, cyst wall came into view, which was thick and gray in color. Cystic fluid was aspirated to shrink the mass. Bleeding was not so troublesome and cyst wall was coagulated for further shrinkage. With special attention not to injure the nearby brain, the mass was excised carefully. There was bleeding from the solid portion of the tumor, but it was successfully controlled with electrical coagulation.

Postoperatively, there was transient tremor in her right hand.

Histological examination of the tumor revealed a dense cellular neoplasm of oval to spindle cells. Cells were surrounded with prominent “stag-horn” sinusooidal vascular channels. These findings were consistent with HPC (Fig. 4). To confirm the diagnosis, immunohistochemical staining was performed. The tumor cells were negative for factor VIII, CD34, GFAP and epithelial membrane antigen (EMA).

Adjuvant cranial radiotherapy was administered with a total dose of 5940cGy in 33 fractions in the immediate postoperative period. On 2 months after operation, follow-up MRI was taken, which shows small enhancing mass in the splenic portion of the corpus callosum (Fig. 2). After 30 months, the brain MRI shows no interval change (Fig. 3).

**Discussion**

Intracranial HPC is rare vascular tumors of CNS and it was accepted as a hemangiopericytic or angioblastic variant of meningioma. The first intracranial HPC was reported by Begg and Barrett in 1954. Currently, HPC is accepted as distinctive mesenchymal neoplasms unrelated to meningioma. Intracranial HPC accounts for 2 to 4% of all meningioma, comprising lesser than 1% of all intracranial tumors\(^1\). Unlike meningiomas, HPC is more common in males than females\(^2\).

Location of HPC is similar to that of meningioma, and most of them have dural attachments. But, rarely it was reported to occur in the pineal, sellar, suprasellar regions, ventricle and in the posterior fossa\(^2\).
On the CT scan of the brain, it is hyperdense with homogeneous enhancement. It usually shows a broad-based dural attachment, although some authors have suggested that it shows relative narrow-based attachment, compare to classic meningioma. On the brain MRI, HPC is iso-intense with cortical gray matter on T1WI, with strong enhancement, however pattern of enhancement is rather heterogeneous compare to classic meningioma. The "dural tail" sign can be present and the presence of associated brain edema is also mild to moderate. It has been reported, that the dural attachment of a HPC is narrower than the one occurring in a meningioma. HPC shows prominent internal vascular flow voids in T2WI MRI than classic meningioma. Despite the advances in neuroimaging techniques, it may cause diagnostic confusion with meningioma. It was reported that magnetic resonance spectroscopy may be useful to distinguish HPC from meningioma because of the higher levels of myo-inositol.

Intracranial HPC has more aggressive clinical behavior than meningioma, with higher local recurrence rate, reaching 64% at 15 years. Also, intracranial HPC may cause extracranial metastasis to bones, followed by the muscle, liver, lungs, abdominal cavity, lymph nodes, skeletal muscle, kidney, pancreas, skin, breast, adrenal glands, gallbladder, diaphragm, retroperitoneum, and heart. Metastasis appear at a mean period of 8 years after initial therapy. Guthrie et al. calculated that the probability of metastasis at 5, 10, and 15 years was 13, 33, and 64%, respectively.

Surgery is the treatment of choice for HPC, total removal is advisable, because of its propensity to recur. The prominent feature of these tumors are extreme rich of vascularity, which may results in substantial intraoperative blood loss. So, preoperative embolization is emphasized in order to reduce intraoperative bleeding. Fortunately, our case was cystic type with little vascularity and surgery can be performed without significant intraoperative bleeding. Perioperative mortality for HPC ranges from 0 to 27%6. The role of postoperative adjuvant radiotherapy has been established. Guthrie et al. demonstrated that patients with HPC who received postoperative radiotherapy showed significantly increased disease free survival time (mean of 74 vs 29 months, \(P < 0.05\)) as well as a long overall survival (92 vs 62 months). The dose of the radiation should be at least 50-55 Gy.

The radiosurgery has been recognized as another alternative to control recurrence of HPC. Sheehan et al. reported that Gamma knife radiosurgery provides local tumor control for 80% of recurrent HPC. Galanis and co-workers reported 10 cases of recurrent meningial HPC managed with stereotactic radiosurgery. Of these, three previously non-irradiated patients (all with lesions <25 mm) achieved complete response, which persisted for a median of 3 years. In seven lesions (70%), a partial response occurred with a median duration of 12 months, whereas three lesions (30%) remained stable. These authors recommended radiosurgery in the treatment of smaller recurrent meningial HPC. But, the role of stereotactic radiosurgery in the management of local intracranial recurrence has some controversies. The role of chemotherapy in the treatment of extracranial metastasis needs to be defined.

Microscopically, HPC was very cellular and nuclei were generally round to fusiform in shape, showing neither coarse chromatin nor nucleolar prominence. Cytoplasm was scarce and poorly defined. Vascularity was abundant, with thin-walled vascular networks showing a "stag-horn" like arrangement and lining with flat endothelial cells. Immunohistochemistry can differentiate HPC from meningiomas by showing positive staining for CD34, factor VIII and vimentin but negative for S-100 protein and EMA. The antigen CD34 (endothelial/vascular marker), a transmembrane glycoprotein present on human progenitor cells and endothelial cells, is a very sensitive marker for endothelial differentiation, stained neoplastic endothelium more strongly than normal endothelium. Factor VIII antigen (FVIII, endothelial/vascular marker) is restricted to endothelial cells and megakaryocytes, which is more specific for endothelial neoplasms than CD31 and CD34 and useful as a confirmatory marker. Although with limited value in diagnosis, vimentin (mesenchymal marker), a mesenchymal intermediate filament, can be demonstrated in most properly fixed tissues, but it is used to identify antigen loss during the process. S-100 protein (neuronal, nerve sheath and melanocytic marker) is widely distributed in peripheral and central nervous systems, the S-100 protein may play a role in ionic regulations and it is expressed in astrocytes, oligodendrocytes and Schwann cells. EMA (epithelial marker) is expressed in the vast majority of epithelial like tumor such as epithelioid
and synovial sarcoma, etc). Because of its tendency to systemic metastasis, periodic follow-up is mandatory, with frequent clinical examinations, chest X-rays, liver enzyme profiles, and even whole body bone scan. Guthrie et al. reported 5, 10, and 15-year survival rates were 67, 40, and 23%, respectively. Our patient showed no evidence of local recurrence of systemic disease at 30 months after the diagnosis.

Intraventricular HPC may originate from pericytes found within the tela choroidea or the stroma of the choroid plexus. The mural nodule must be included in the differential diagnosis with cystic HPC, subependymoma, ependymoma, anaplastic ependymoma, cystic astrocytoma, hemangioblastoma and paraventricular malformative cyst. The mechanisms of cyst formation of HPC are not fully explained but there are some possible mechanisms of cyst formation, central degeneration and necrosis of the tumor due to insufficient blood flow, edudation of plasma complements, intratumoral bleeding, reforming process of the subarachnoid spaces and aggregation and enlargement of microcysts. To our knowledge, all the ventricular hemangiopericytomas reported in the literature were solid type (total 9 cases: 6 cases in lateral ventricle, 3 cases in 3rd ventricle). The cystic HPC reported in literature were only 3 cases, one of them was in middle fossa, another was in cerebellum, the last one was within occipital area. Authors believe this report may be the first report of intraventricular cystic HPC.

Conclusion

The diagnosis of the intracranial HPC became possible thanks to the development of imaging study for the brain, including brain MRI and cerebral angiography. But, unusual location or cystic variant of HPC is difficult to diagnose by radiological studies only. And, more aggressive nature of HPC than meningioma require close follow-up to detect local recurrence and systemic evaluation for systemic progression of the disease.

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References