Intraventricular Hemangiopericytoma

Intraventricular hemangiopericytoma is a rare intraventricular tumor, with only 10 cases reported in the literature till 2016.

Diagnosis

They may pose a difficult diagnostic dilemma based on their radiographic and gross appearances. Because of this difficulty, histological confirmation is required to make a definitive diagnosis [1)].

Differential diagnosis

Differential diagnosis of hemangiopericytoma from intraventricular meningioma and solitary fibrous tumors is very important because the clinical behavior of hemangiopericytoma is very aggressive, including local recurrence or distant metastases in the central nervous system and periphery. Histological confirmation is required to make a definitive diagnosis [2)].

MR spectroscopy (MRS) can be useful in characterizing intraventricular tumor and distinguishing CNCs from meningiomas and other intraventricular tumors [3)].

Treatment

These lesions have a propensity to recur and metastasize in the central nervous system and periphery, thus making the goal of treatment a complete surgical resection followed by postoperative radiation therapy in most cases [4)].

Case reports

2016

A 40-year-old man who presented with raised intracranial pressure. His MRI showed a 3.3×3.2×3.2cm heterogeneously lesion with contrast enhancement in the left frontal horn obstructing the foramen of Monro and causing hydrocephalus. The tumor was excised through an anterior interhemispheric, transcallosal approach, and histopathology revealed an anaplastic hemangiopericytoma (World Health Organization grade III). To our knowledge this is the first report of this rare pathology being located within the frontal horn of the lateral ventricle [5)].

A 23-year-old man with a left intraventricular hemangiopericytoma presenting with headache, word-finding difficulties, blurred vision, nausea, vomiting, photophobia, and right-sided weakness and numbness. Using a left superior parietal lobule approach, a piecemeal resection was completed, achieving radiographic gross total resection. Pathology was consistent with a hemangiopericytoma. He was treated adjunctively with 60 Gy of local radiation. At 6-month follow-up, the patient had

resolution of his aphasia and improvement in his headaches, with no signs of recurrence or metastasis on imaging.

Standard treatment for central nervous system hemangiopericytoma includes aggressive surgical resection. The role of adjuvant radiation is less well defined but is commonly pursued postoperatively. Regardless of extent of resection or adjuvant treatment, close follow-up to evaluate for evidence of local recurrence and distant metastasis is essential. [6].

2011

A 67-year-old right-handed woman presented with a case of hemangiopericytoma in the lateral ventricle manifesting as digit number memory disturbance, sensory aphasia, and right quadrantanopsia. Magnetic resonance imaging demonstrated a 6-cm homogeneously enhanced mass in the trigone of the left lateral ventricle. The tumor was totally removed via the left inferior temporal gyrus. The histological findings were consistent with hemangiopericytoma. Differential diagnosis of hemangiopericytoma from meningioma and solitary fibrous tumors is very important because the clinical behavior of hemangiopericytoma is very aggressive, including local recurrence or distant metastases in the central nervous system and periphery. Histological confirmation is required to make a definitive diagnosis. The present patient did not receive radiation therapy, but developed no local recurrence or metastases. Complete surgical resection and awareness of hemangiopericytoma in the lateral ventricle are very important for good clinical outcome [7].

2010

A 65-year-old woman presented with a very rare hemangiopericytoma in the body of the lateral ventricle. Magnetic resonance imaging demonstrated a homogeneously enhancing mass lesion occupying the bilateral medial portions of the body of the lateral ventricle. Cerebral angiography disclosed a vascular-rich tumor, fed mainly by the left lateral posterior choroidal artery. After devascularization of the feeding vessel by endovascular coiling, the patient underwent complete surgical excision of the tumor via an anterior transcallosal approach, followed by radiation therapy, and has thus far been disease-free for 5 years. The present patient represents the first reported case of hemangiopericytoma arising in the body of the lateral ventricle. In this location of hemangiopericytoma, preoperative embolization could provide a promising option in terms of reducing the intraoperative blood loss and achieving total tumor extirpation with minimum damage to the surrounding structures. Since preoperative identification of hemangiopericytomas confers therapeutic advantages,it is important to be aware that they can occur at unusual ventricular sites [8].

2009

A 31-year-old female presented with a particularly rare hemangiopericytoma (HPC) in the right lateral ventricle manifesting as a 6-month history of visual disturbance and headache. Left hemianopsia and choked disc were identified by an ophthalmologist who referred her. Magnetic resonance imaging demonstrated a 5-cm homogeneously enhanced mass in the trigone of the right lateral ventricle. The tumor was totally removed by two stage surgery. The histological findings were consistent with HPC. HPC is very important to differentiate from meningioma and solitary fibrous tumors because HPC is more aggressive. The histological and immunochemical findings are important for the differential

diagnosis. The present case showed no local recurrence or metastasis without radiation therapy for 4 years, indicating that radiation therapy is not absolutely imperative for patients with intraventricular HPC showing low MIB-1 staining index after total removal [9].

2004

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 [10].

A 55-year-old woman presented with gradual onset of left side weakness, gait ataxia, and tendency to miss objects in the left visual field of uncertain duration. Magnetic resonance imaging with contrast showed a tumor with homogenous enhancement in the right lateral ventricle. The patient underwent right temporoparietal stealth-assisted craniotomy and surgical removal of the tumor. Histopathologic examination with ancillary tests confirmed hemangiopericytoma. Awareness that hemangiopericytoma can occur as an intraventricular tumor is important for clinicians and pathologists. Because of radiologic similarity, this tumor is not to be confused with intraventricular meningioma because the prognosis is different [11].

1999

Abrahams et al. present the first reported case of a hemangiopericytoma (HPC) occurring in the third ventricle. Most of these lesions are based in the meninges. There is only one other reported case of an intraventricular HPC; in that case the lesion was found in the lateral ventricle. A 40-year-old right-handed man presented with a 3-month history of headaches. Clinical evaluation, including computerized tomography and magnetic resonance imaging studies, revealed a 1-cm enhancing lesion in the third ventricle. Given the findings on the preoperative imaging studies, the lesion was not consistent with some of the more commonly occurring tumors of the third ventricle, namely colloid cysts. A transcortical approach and resection of the lesion was performed without complication. The final pathological findings were consistent with those of an HPC. Hemangiopericytomas rarely occur in the ventricles and may pose a difficult diagnostic dilemma based on their radiographic and gross appearances, as shown in this case. Because of this difficulty, histological confirmation is required to make a definitive diagnosis. These lesions have a propensity to recur and metastasize in the central nervous system and periphery, thus making the goal of treatment a complete surgical resection followed by postoperative radiation therapy in most cases [12].

1991

A 41-year-old male with a 2-month history of headache and paresthesia of the right shoulder and arm. The tumor was partially removed, followed by 50 Gy local Linac irradiation given over 6 weeks. Four months later the residual tumor demonstrated a marked decrease in size and vascularity. The residual tumor was totally removed with less operative bleeding than at the initial operation. This is

the first reported case of hemangiopericytoma located in the trigone of the lateral ventricle [13].

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