A Rare Clinical Case of Giant Hemangiopericytoma

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The authors provide an example of successful treatment of a patient with a giant intracranial hemangiopericytoma. Hemangiopericytomas are aggressive tumors characterized by a high rate of recurrence and delayed metastasis. Despite the malignant nature of these tumors, they often reach a large size with minor clinical signs. Surgical removal of the tumor is still the primary treatment method.

Keywords: hemangiopericytoma, surgery of intracranial hemangiopericytoma.

Meningeal hemangiopericytomas (HPCs) are aggressive tumors characterized by a high rate of recurrence and delayed metastasis. Despite the malignant nature of these tumors, they often reach a large size with minor clinical presentations. To date, surgical treatment still remains the primary treatment of these tumors.

HPC is a malignant tumor arising from pericapillary cells or Zimmerman pericytes. These tumors usually occur in the lower extremities, pelvis, and retroperitoneum.

Intracranial HPCs are rare, as they amount to 2—4% of all meningeal tumors and less than 1% of all intracranial tumors.

According to the latest 2013 WHO classification of tumors of soft tissue and bone [25, 26], HPCs are included, as a histological variant, to a single nosological form — extrapleural solitary fibrous tumor called “solitary fibrous tumor with predominance of a hemangiopericytoma-like vascular component”. The term hemangiopericytoma is regarded as an obsolete synonym.

Despite this fact, taking into account the process localization and based on the current 2007 WHO classification of CNS tumors and a retrospective analysis of the literature on tumors previously diagnosed as HPCs, we reserve the right to use this term in the present study.

Clinical case

A 40-year-old male patient P. was admitted to the Central Clinical Military Hospital of the Russian Federal Security Service on March 13, 2009 with complaints of bilateral visual impairment, double vision, and pronounced persistent headache localized mainly in the fronto-parietal regions.

According to the medical records and the patient, the disease started in December 2008, when headache appeared. Later on, visual impairments (diplopia and tunnel vision) developed within 1 month. In January 2009, a medical examination by an ophthalmologist revealed congestion in the fundus. The patient was referred to a local health care facility for hospital treatment and examination. The examination revealed a giant mass lesion in the left cerebral hemisphere, presumably meningioma of the anterior clinoid process region of the left wing of the sphenoid bone. The patient was hospitalized to the Neurosurgical Department of the Central Clinical Military Hospital of the Russian Federal Security Service for further examination and treatment, where the diagnosis was confirmed during further examination. Decongestant and dehydration therapy with corticosteroids was started.

Examination revealed no splanchnopathy. The patient was conscious and cooperative. The patient’s criticism to his condition and the surrounding was reduced. The patient answered questions adequately, but occasionally, with elements of coprolalia. Neurological status: the patient did not bring his eyeballs laterally to the left, upgaze paresis, no accommodation, asymmetric face, descended right corner of the mouth. Muscle strength of the patient’s arms and legs was sufficient and amounted 5 points on both sides. The tendon reflexes were increased, there was anisoreflexia with an increase in the tendon reflexes right and extension of the reflexogenic zones. There were patellar and foot clonuses, right more than left. Positive Oppenheim’s and Pussep’s signs on the left side. The Marinesco–Radovici reflex on both sides. Distal hyperhidrosis. Ophthalmological examination: VIS OS = 0.9, VIS OD = 1.0. Visual fields: concentric narrowing of the visual field to 20—30° on the right of the fixation point and to 30—40° on the left of the fixation point, absolute paracentral scotomata of both eyes. Ocular fundus: the optic discs were hyperemic and edematous with blurred boundaries and protruded into the vitreous body; peripapillarily — edema, small hemorrhages, exudates, significantly narrowed and spastic arteries; dilated veins with ruptures and exudation. The crowded optic discs of both eyes.

Total selective carotid angiography (SCA) revealed a hypervascularized mass in the anterior and middle cranial fossae, left, with a size of 7×7 cm, fed by the branches of the left middle cerebral artery (MCA), left external carotid artery (ECA), and left intracranial segment of the internal carotid artery (ICA). The venous sinuses were without obstruction and thrombosis signs (Fig. 1).

Brain CT revealed a large mass lesion characterized by sufficiently distinct nodular contours, sized 78×76×64 mm,
located predominantly in the left fronto-temporo-parietal region, with basal areas adherent to the right frontal lobe by 10—12 mm. The mass was intimately adjacent to the left carotid siphon, floor of the anterior cranial fossa, and anterior tubercle of the sella turcica. The bones forming the floor of the anterior cranial fossa were corroded. The mass compressed the anterior parts of the left lateral ventricle and likely extended to its lumen. When intravenous contrast enhancement was used, the mass accumulated a contrast agent somewhat unevenly (up to +60 — +85 Hounsfield Units). The mass was fed by numerous arterial vessels with a diameter of up to 2—3 mm, which were located primarily on the middle and anterior sides of the mass. Minor perifocal edema was observed (up to 7 mm, at the lateral margin of the mass in the left temporal region) (Fig. 2).

The surgery was carried out on 25.03.09. The arcuate incision was performed in the left fronto-parieto-temporal region. The extended pterional approach was performed. Resec-

Fig. 1. Selective carotid angiography (S).
Hypervascularized mass lesion of the anterior and middle cranial fossae, left, sized 7×7 cm, and fed by the branches of the left MCA, ECA, and intracranial segment of the ICA. The venous sinuses are with no signs of obstruction and thrombosis.

Fig. 2. Pre-operative contrast-enhanced CT.
a — axial view; b — frontal view. A large mass lesion with distinct nodular contours, sized 78×76×64 mm, with basal areas adherent to the right frontal lobe by 10—12 mm is observed in the left fronto-temporo-parietal region. The mass compresses the anterior parts of the left lateral ventricle and likely extends to its lumen. The mass unevenly accumulates a contrast agent. Minor perifocal edema is observed.
tion of the external part of the left wing of the sphenoid bone was performed extradurally. The dura mater (DM) was severely tense, bulging, no brain pulsation was observed. DM was opened along the lateral margin of the basal parts of the left frontal lobe. An operating microscope and microsurgical instruments were used for further surgery. The frontal lobe was drawn aside the skull base. A bright pink undulating tumor separated from the brain substance was found. It was soft, readily bleeding, and looked more like a paraganglioma rather than a meningioma. There was heavy bleeding (bright red blood), and hemostasis was performed by pressing the tumor using cotton rolls with hydrogen peroxide. Acute swelling and bulging of the brain developed with its fungus bulging to the DM defect. Decongestant measures were undertaken. Microsurgical manipulations were renewed after a persistent decrease in the tension and bulging of the brain. The tumor was easily fragmented and removed using conventional vacuum suction. A tumor portion was sent to urgent histologic examination, the preliminary result of which was a mixed type meningioma. The tumor was mainly homogeneous and bleeding. Only medial-basal regions of the tumor structurally resembled a nodular cellular meningioma with a dense stroma, with more distinctly developed vessels in the stroma and a higher density. Blood supply of the tumor was carried out mainly through the transitional pial vessels that resulted in very intense bleeding when the tumor was resected along the boundary with the brain substance. The tumor matrix was found in the medial segments of the sphenoid bone wing at the boundary with the anterior clinoid process, where the tumor stromal structure was the most pronounced. DM in this area was eroded, bleeding, and thoroughly coagulated. The tumor was completely removed. The left supraclinoid ICA segment, the ICA bifurcation, and the initial segments of the MCA and anterior cerebral artery were visualized. Final hemostasis was achieved using Surgicel hemostatic cotton. Distinct brain pulsation appeared. The brain smoothened and partially retracted. No anatomical damage to the brain matter due to brain traction occurred. DM was continuously sutured without tension using an atraumatic suture. The bone flap was laid on the place and fixed with a bone suture. Layered wound closure. Intraoperative blood loss was about 3,500 mL.

Histological diagnosis: the examination carried out at the Central Clinical Military Hospital of the Russian Federal Security Service revealed the mixed-type meningioma with a dense cell arrangement. No necrosis foci or mitotic figures were found. An immunohistochemical examination in the laboratory of neuromorphology at the Burdenko Neurosurgical Institute of the Russian Academy of Medical Sciences revealed: hemangiopericytoma, pronounced focal expression of Vim, CD 34, Factor XIII, and the Ki-67 proliferation marker >5%, Grade III (Fig. 3).

The postoperative period was complicated by right-sided pleuropneumonia, formation of epidural hydroma at the surgical area, and persisting cephalgic syndrome. The patient received combined therapy, including replacement of postoperative blood loss (transfusion of packed red cells, in the total amount of 1,745 mL, and fresh frozen plasma – 4,900 ml), antibacterial therapy, sanitation lumbar punctures, and decongestant therapy. Cerebrospinal fluid cultures were sterile, with no growth. The wound healed by primary intention. Regression of general cerebral and focal neurological symptoms and regression of mental disorders were observed in the immediate postoperative period.
Fig. 4. Contrast-enhanced CT. First day after surgery.

a — axial view; b — frontal view. The tumor is radically removed. There are no hemorrhagic complications in the removed tumor bed.

Fig. 5. Contrast-enhanced CT in the axial plane (a). Contrast-enhanced MRI in the axial (b) and frontal (c) planes. Five months after surgery (no signs of tumor recurrence).
Contrast-enhanced CT on the 1st postoperative day revealed no areas of pathological uptake of a contrast agent. The tumor was completely removed (Fig. 4).

Control contrast-enhanced CT and MRI 5 months after surgery revealed no signs of recurrence (Fig. 5).

However, given the histologic pattern, radiologists of the Neurosurgical Institute recommended radiotherapy, at the place of residence, of the resected tumor area at the total focal dose (TFD) of 56 Gy.

Five-year follow-up of the patient showed no tumor recurrence.

Discussion

The term hemangiopericytoma was first used by A. Stout and M. Murray [23] in 1942 to describe a tumor located in the retroperitoneal space and consisted predominantly of proliferating pericytes.

Meningeal HPCs were first described in 1928 by R. Bailey et al. [2] and were considered an angioblastic type of meningioma. Later, immunohistochemical, ultrastructural and genetic studies demonstrated a fundamental difference between HPC and all other types of meningiomas [12, 18]. The latest WHO classification (2007) of CNS tumors classifies HPCs as mesenchymal non-meningioma tumors and assigns them two grades: II and III, according to their proliferative potential [14]. HPC consists of small oval cells with a large number of thin-walled vessels of various calibers and has the characteristic antler-like vascular pattern [18]. Many authors indicate the characteristic neuroradiological and angiographic features of HPCs. Unlike meningiomas, the majority of HPCs have the multilobar structure with indistinct boundaries and infiltration of the surrounding brain tissue without hyperostosis, bone erosions, and calcifications. The presence of perifocal edema is also observed. HPCs are predominantly supplied by the branches of the ICA and posterior cerebral artery, unlike meningiomas, that are mainly supplied by the meningeal branches of the ECA. Some authors [10, 16] indicate the presence of a large number of small corkscrew-like vessels in the stroma and pronounced vascular ture, which is the distinctive feature of HPCs.

According to the literature [11], preoperative embolization of the tumor-feeding vessels is an effective method to reduce blood supply. However, given the features of HPC blood supply, it is not always possible.

Surgery is currently regarded as the primary method of treatment for meningeal HPCs [10, 13, 15]. The frequency of local recurrences is high even after radical resection of HPC. According to some authors [15], this indicator amounted up to 50%. Postoperative radiation therapy reduces the frequency of HPC recurrence [19—21, 24]. In the case of a hard-to-reach location of tumors, sparing surgery combined with postoperative radiation therapy is advisable [22]. According to most authors [8, 9, 19, 20], the most efficient TFD was 50—60 Gy.

Chemotherapy was ineffective in treatment of patients with meningeal HPCs [8]. According to E. Galanis et al. [8], only 1 of 7 patients had the positive dynamics during chemotherapy with doxorubicin. However, some authors [1] report on the efficacy of a combination of ifosfamide and epirubicin.

Intracranial HPCs are aggressive tumors with a high rate of recurrence and delayed metastasis [7, 17]. N. Mena et al. [16] who observed 94 cases of HPC reported on the recurrence rate of 70% and the metastasis rate of 27%. The bones, lungs, and liver are the most frequent metastasis localizations, but there are reports on other metastasis localizations [5, 20]. According to the literature [6], metastases were most frequently observed 63—99 months after the diagnosis. A case of HPC metastasis after 20 years was also described. Bone metastases appear as osteolytic lesions on plain radiographs. In most cases, tumor recurrence tends to precede the emergence of delayed metastasis [3].

Conclusions

The analysis of the present clinical case and world literature indicates the efficacy of surgical removal of meningeal HPCs as the first and primary treatment. The specific feature of this clinical case is that the tumor reached a giant size with minimal clinical manifestations. Despite a careful additional examination, it was extremely difficult to establish the correct clinical diagnosis prior to surgery.

Given high vascularization of HPC, radical tumor resection is the main requirement for successful surgical treatment and elimination of postoperative hemorrhagic complications. Radiation therapy is auxiliary treatment after surgery. In cases of doubts in a histological diagnosis based on light microscopy, an immunohistochemical study is the essential prerequisite for the correct diagnosis.

REFERENCES

Hardly detectable and quite rare brain hemangiopericytomas (HPCs) are malignant tumors with main characteristics often resembling those of cerebral meningiomas. However, they have a high rate of recurrence and delayed metastasis.

The paper provides an interesting clinical case of giant intracranial HPC at the Neurosurgical Department of the Hospital of Federal Security Service. The clinical case is discussed in details, both patient’s complaints and the dynamics of clinical manifestations are analyzed, the results of objective neurological examination are presented. In this case, the clinical syndrome resembled that occurring in patients with basal meningiomas with initial growth from the clinoid region. The results of objective neuroimaging examinations are provided, including contrast-enhanced CT and total selective carotid angiography. The paper is well illustrated with photographs.

Next, stepwise surgery for tumor resection is described in detail (and it is very instructive), including selection of an adequate surgical approach, its details and the implementation technique, peculiarities of dura mater (DM) opening, imaging characteristics of the identified tumor and its structure, which is non-uniform in different tumor portions. The tumor blood supply sources identified during the surgery and successful hemostasis methods are described in details. Nevertheless, hemostasis was assiduous, and blood loss was 3 L. The relation between the tumor and the great vessels, including the supraclinoid internal carotid artery (ICA), ICA bifurcation, and system of the middle and anterior cerebral arteries, is fundamentally clarified. Everything is convincingly and thoroughly described, but it would be desirable to see illustrative images of the surgery stages that are lacked in the article. The postoperative period proceeded with moderate complications that were timely and successfully stopped.

Control examinations (contrast-enhanced CT), conducted in the postoperative period, demonstrate success of the surgery. The immunohistochemical study conducted at the Neurosurgical Institute clarified the diagnosis and tumor grade (Grade III). Later, radiation therapy was conducted, and the patient was followed up for 5 years, no tumor recurrence was observed.

A brief discussion of the problem and the analysis of the literature in a historic context are provided at the end of the article. It is emphasized that, unlike brain meningiomas, main blood supply is provided through the branches of the ICA and posterior cerebral artery, rather than through the meningeal arteries. Due to pronounced vascularization of HPC, the feeding vessels should be preoperatively embolized when possible.

In conclusion, the authors emphasize the undoubted significance of surgical treatment of these tumors as the basis of the treatment process, complemented by radiation therapy, if necessary.

The article is very interesting and informative, especially for modern practicing neurosurgeons. It reflects the important tendencies in the search for appropriate ways to improve the technique of microsurgical operations for resection of such complex tumors as hemangiopericytomas.

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