MULTIPLE CAVERNOUS MALFORMATIONS OF THE BRAIN

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Abstract. *Cavernous malformations (cavernomas, angiomas and hemangiomas) are the most common vascular neoplasms of the brain. They can be detected at any age as incidental findings or cause serious neurological disorders. Despite the non-oncological nature, with the manifestation and increase of symptoms, the patient with a high degree of probability requires treatment for cavernoma.*
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Cavernomas consist of pathological cavities separated by partitions and filled with blood. They can be located in any part of the brain, mainly in the hemispheres, but they can also be localized in the brain stem, basal ganglia or corpus callosum. The frequency of detection of cavernous malformations ranges from 0.6 per 100,000 population per year.

In most patients, clinical symptoms appear at the age of 40-60 years. In most cases, this is a single find. Multiple cavernous malformations or the presence of malformation in family members may indicate the hereditary nature of the pathology. With this disease, abnormal and swollen veins appear, similar to blackberries. Cavernomas can also bleed, as well as other vascular abnormalities. However, unlike bleeding from an aneurysm and AVM, bleeding from a cavernoma is usually small, and blood flows around the lesion. Profuse bleeding is rare. Cavernous malformations may exist asymptomatically or cause neurological symptoms: epileptic seizures, hemiparesis, visual impairment, damage to cranial nerves. The most severe complication of cavernoma is hemorrhage. In this case, the patient's condition worsens significantly, it is possible to develop pronounced neurological disorders and even death. According to various authors, the risk of hemorrhage ranges from 0.25% to 16.5% annually. The probability of hemorrhage increases in patients who have already suffered hemorrhages or when the neoplasm is localized in the brain stem or deep parts of the brain. Thus, in patients who have suffered a hemorrhage, the risk of repeated hemorrhage is already 33.9%.

Cavernous malformations (as well as other types of vascular malformations: AVM, telangiectasia and venous angiomas) do not belong to tumor diseases and represent atypical development of some blood vessels. However, the threat to the body from the cavernoma is the compression of brain tissue, which leads to neurological symptoms with which the patient first goes to the doctor. Cavernous venous malformations of the brain in ~ 80% of cases are lacalized supratentorially, but in general can be localized in any part of the brain, including the trunk. Most often they are isolated, but at the same time, one third of patients with sporadic education may have more than one malformation.

If the cavernous malformations have an insignificant size, the patient may be prescribed medication, as well as constant monitoring of the disease. Otherwise, when it comes to a neoplasm of significant size, which can lead to serious disorders in the brain, up to a fatal outcome, it is necessary to remove the cavernous malformation surgically. Cavernous malformations can be asymptomatic for a long time. However, until the neoplasm reaches a significant size and begins

to exert pressure on brain tissue, causing headaches and other neurological complications. But often the detection of a cavernoma can be accidental when a patient contacts a specialist with complaints about another neurological disease. The main method of diagnosis of cavernoma is magnetic resonance imaging (MRI), in which the vascular neoplasms of the brain are clearly visible. Large malformations are visualized by computed tomography as areas of increased density, surrounded by perifocal edema in the presence of subacute hemorrhage, and are not enhanced by contrast. Small malformations are difficult to distinguish. Magnetic resonance imaging is the method of choice. Classic are the signs of popcorn or berries, surrounded on the periphery by a ring-shaped zone of signal loss due to the presence of hemosiderin and characterized by increased magnetic susceptibility. Most of the cavernous venous malformations are an accidental finding and they mostly manifest themselves clinically during life. Clinical manifestations are caused by hemorrhages with the development of focal neurological symptoms or the development of seizures. The risk of hemorrhage is ~1% per year for hereditary multiple malformations and significantly less for sporadic single malformations. Macroscopically, the cavernous venous malformation looks like a mulberry berry. In histological examination, cavernous venous malformations are thin-walled irregularly shaped cavities, the walls of which are formed by the endothelium. Cavities can fit tightly to each other, or be separated by collagen fibers or fibrous tissue, unlike AVMs, between the loops of which brain matter may be present. In some cases, they are closely associated with venous malformations (DVA) and in this case it is a mixed vascular malformation. Mostly asymptomatic and, if necessary, behave conservatively. Clinically manifested malformations due to volumetric exposure, epileptogenic activity or repeated hemorrhages are resected, if possible.

Cavernomas can be asymptomatic all their lives. However, in a number of patients, clinical manifestations can be of two types, both individually and in combination:

- Hemorrhage. Sometimes, an increase in blood pressure inside the cavernoma leads to local destruction of the vascular wall and the formation of intracerebral hemorrhage. Unlike AVM, hemorrhages from the cavern are never massive and do not pose a threat to the patient's life, with the exception of extremely rare cases of cavern location in the lower parts of the medulla oblongata, where the centers of cardiovascular and respiratory regulation are located. However, if the hemorrhage focus is located in any functional area, hemorrhage of even a small volume can lead to the appearance of neurological symptoms (for example, the development of contralateral hemiparesis when the focus is located in the precentral gyrus of the frontal lobe).
- Epileptic syndrome. In some cases, due to the chronic presence of hemosiderin in the brain substance or with the development of acute hemorrhage, a focus of pathological cerebral bioactivity may form, clinically manifested by epiprimes of various structures (convulsive, non-convulsive, absences, vegetative, polymorphic and others).

Cavernous venous malformations should be differentiated from numerous other pathologies leading to multiple micro-hemorrhages, including:

- cerebral amyloid angiopathy
- chronic hypertensive encephalopathy
- diffuse axonal injury
- cerebral vasculitis
- hemorrhagic metastases
- Parry-Romberg syndrome

Large malformations can simulate:

- hemorrhage into metastases
- hemorrhage into the primary tumor (for example, glioblastoma)

Patients with asymptomatic cavernous malformations of any localization should be under dynamic supervision (especially in the brain stem) and annually perform an MRI of the brain. The relatively benign nature of the formations makes it possible to refuse surgical treatment in the absence of rapid growth or pronounced neurological manifestations (hemorrhage and /or epileptic seizures). Some authors believe that only dynamic observation of patients for 3 years makes it possible to determine the leading clinical syndrome and indications for surgical intervention in the complex of treatment. Symptomatic cavernous malformations should be treated aggressively due to the high risk of recurrent hemorrhage (cortical and subcortical cavernous malformations should be removed using the smallest corticotomy or, when possible, through furrows). Even if the operation entails high risks of damage to important areas such as the brain stem, surgical removal can prevent further deterioration due to repeated hemorrhage. Currently, neuronavigation is used to minimize access. Neuronavigation is important for the treatment of small deep-seated cavernous malformations, it can also help in planning an access point to minimize surgical access. Intraoperative ultrasound examination of the brain through the formed trepanation window is actively used to determine localization. Radiosurgery (stereotactic) as an alternative method of treating deep or epileptogenic has no real advantages compared to conservative treatment and is mostly not recommended for children. However, with multiple can be considered as, perhaps, the only method of treatment.

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