Hemangioblastomas are benign tumors that develop from the vessels of the central nervous system and can be a manifestation of autosomal dominant von Hippel-Lindau disease. Statistically, they account for 1.5–2.5% of all intracranial tumors and 2–15% of spinal cord tumor lesions. There are very few publications on the intramedullary localization of these neoplasms.

The patient, 45 years old, a serviceman, presented with complaints of headache, slight unsteadiness of gait, as well as slight weakness in the right extremities, more pronounced in the right upper extremity, periodic numbness of the upper extremities, which progressed and made further service impossible. On neurological examination: pupils D=S, light reflexes were brisk, eye movement was fully preserved and horizontal nystagmus was present. BNI - PS - I, BNI - NS - I. HB - I. GR - I. Swallowing and phonation were fully preserved. There was a slight hemiparesis on the right. Hemihypesthesia on the right was more prominent in the upper extremity. Ataxia of mixed genesis. Pelvic organs function was preserved. Periodic constipation for up to 7 days. Magnetic resonance imaging revealed a multifocal brain lesion. Supratentorially, a cystic mass measuring 2.60×2.12×2.14 cm with a solid component up to 1.5 cm in the diameter was detected in the of the thickened corpus callosum. Intramedullary cystic-solid lesion of the medulla oblongata extending to the cervical spinal cord with conventional dimensions of the solid component 1.76×1.23×1.57 cm and the cystic component was 1.52×1.62×1.22 cm. Magnetic resonance imaging of the cervicothoracic region of the spinal cord revealed significant hydromyelitic expansion of the central canal from the C2 level to the Th3 level (up to 10mm in the diameter). Endovascular embolization of the neoplasm with a liquid embolic agent (Phil) and following microsurgical en bloc tumor resection were performed.

Hemangioblastomas with an intramedullary location are extremely difficult and risky for the surgical removal. The presence of a cystic component in the hemangioblastoma structure or perifocally gives a chance to remove such a neoplasm avoiding risk of functional deterioration. Preoperative endovascular obliteration of hemangioblastoma vascularity is considered as an effective measure, although it is associated with a risk of cerebral ischemia in corresponding brain structures. Another crucial issues of intramedullary hemangioblastoma surgery includes multimodal intraoperative neuromonitoring and maximum possible intraoperative magnification to minimize injury of adjacent brain structures. Present practice shows that multiple intramedullary cysts in the spinal cord commonly regressing after neoplasm removal and leads to improvement of neurological deficit within a relatively short period of time.

Conclusions. Hemangioblastomas with intramedullary growth and perifocal cyst can be surgically removed with a good functional result and comprehensive approach includes preoperative selective angiography with endovascular embolization, multimodal intraoperative neuromonitoring and appropriate microsurgical technique.

Keywords: hemangioblastoma; von Hippel-Lindau disease; intramedullary lesion; brain stem; endovascular embolization; microsurgical removal

Introduction
Hemangioblastomas are benign tumors that develop from the vessels of the central nervous system and can be a manifestation of autosomal dominant von Hippel-Lindau disease [1–5]. Statistically, they account for 1.5–2.5% of all intracranial tumors and 2–15% of spinal cord neoplasms [6–8]. Hemangioblastomas are more commonly located in the cerebellum, supratentorial brain structures or in the spinal cord. The literature review of the PubMed database
for the period from 1975 to 2022, found only 26 articles describing intramedullary tumor growth, suggesting the sporadic localization of hemangioblastoma in this brain area.

Patients with hemangioblastomas usually have minor focal neurological symptoms that progress over time. When lesion located in the spinal cord, sensory and motor abnormalities are mostly often observed, when in the cerebellum - static and coordination disturbances usually revealed [5, 9, 10]. In rare cases hemangioblastoma is observed as an incidental finding [11]. Sensorineural hearing loss has been reported as the first clinical manifestation of cerebellar hemangioblastoma [12]. Only one case of hemangioblastoma with brain stem hemorrhage has been described [13].

In rare cases, sudden development of neurological deficit due to intramedullary hemorrhage have been reported in patients [14, 15]. A combination of hemangioblastoma and syringomyelia is rarely observed [5].

Computed tomography (CT) and magnetic resonance imaging (MRI), 3D-CT angiography, and selective angiography are used to diagnose hemangioblastoma. Positron emission tomography/CT may be used in controversial cases [1].

Due to high vascularization, partial removal of hemangioblastoma is practically impossible. Given the high intraoperative risk of ischemic complications in vital brain areas, it is recommended to apply selective angiography to determine the vascular anatomy of hemangioblastoma and to assess the feasibility of interventional devascularization. Preoperative embolization can be indicated to reduce intraoperative bleeding and create optimal conditions for the safe tumor removal [16–19]. Endovascular embolization is usually performed one day before the planned microsurgery. Possible complications associated with embolization include ischemic stroke in the vertebrobasilar circulation and extravasation of embolic material [9, 20‒22].

The occurrence of a hematoma from tentorial veins previously coagulated and transected in the early postoperative period after resection of cerebellar hemangioblastoma with an approach to the tentorium has been described [23]. A. F. Joaquim and E. Ghizoni observed better neurological outcome in patients with preserved neurological status before surgery [9].

Embolization may be contraindicated in some cases, for example, in the case of cerebellar hemangioblastoma, which is supplied by a persistent (primitive) hypoglossal artery. Due to existent embryonic carotid-vertebrobasilar anastomosis, embolization of neoplasm is impossible because of high risk of severe brain stem brainstem ischemia [24, 25]. Unfortunately, there is extremely limited information in the literature regarding preoperative embolization of brain stem hemangioblastomas [21, 26].

Microsurgical technique [4, 10] and maximum magnification of the microscope are crucial in the removal of hemangioblastoma. J. Joseph et al. (2018) advise to leave the "tumor capsule" attached to the brain stem to preserve vital functions, preventing complications associated with the risk of damage of adjacent anatomical structures [27]. Although these neoplasms do not have a capsule as such, it is likely that they are the walls of a perifocal cystic neoplasm. It is preferable to remove the tumor by atraumatic circular devascularization in a single block to reduce the risk of intraoperative bleeding [21]. An extremely important element of functional preservation in intramedullary neoplasms is multimodal intraoperative neuromonitoring [28‒30].

Von Hippel-Lindau disease is a serious problem for both patients and their families. Timely diagnosis and treatment of this disease can improve prognosis and survival rates [8].

In the long term after surgery (after 6, 24, 36 months), gradual recovery in neurological status is commonly observed [31].

Case report

The patient, 45 years old, a serviceman of UAF, presented with complaints of headache, slight unsteadiness of gait, as well as slight weakness in the right extremities, more pronounced in the right upper extremity, which made further service impossible. The patient had periodic numbness of the upper extremities, mainly the right, which progressed.

Reportedly, the patient considers himself ill for about two years when he first noticed slight numbness in the right upper extremity. Over the last few months, the patient’s condition has worsened due to an increase in sensory disturbances in the upper extremities and the appearance of movement disorders in the right extremities.

Neurological examination: consciousness is clear (15 points on the Glasgow Coma Scale (GSC). Pupils D=S, photoreactions are spared, ophthalmodynamics is fully preserved. Horizontal nystagmus jerks are observed. BNI - PS - I, BNI - NS - I. HB - I. GR - I. Swallowing and phonation are unchanged. There is slight hemiparesis on the right. Hemihypesthesia on the right is more in the right upper extremity. Ataxia of mixed genesis. Pelvic organs function is preserved. Periodic constipation for up to 7 days.

Ophthalmologist: visual fields are not changed. The discs of the optic nerves are pale pink, the borders are clear, the pupils are symmetrically narrowed, white spots in the macular area on the right, retinal dystrophy. Degeneration of the macular area on the right.

Otolaryngologist: chronic sensorineural hearing loss (AD=AS). Static and coordination dysfunction.

MRI of the brain (February 18, 2022): multifocal brain lesions (Fig. 1-3). Supratentorially, in the projection of the corpus callosum thickening with an accent to the left, a cystic mass measuring 2.60×2.12×2.14 cm, surrounded by a zone of edema up to 3.48 cm wide was determined. A solid component with a conventional diameter of up to 1.5 cm. Posterior portions of the lateral ventricles are compressed (Fig. 2). Intramedullary cystic-solid lesion of the medulla oblongata with extension to the cervical spinal cord with a conventional size of the solid component of 1.76×1.23×1.57 cm
Fig. 1. A cystic mass measuring 2.60×2.12×2.14 cm is determined in the projection of the corpus callosum thickening with an emphasis to the left.

Fig. 2. Intramedullary cystic-solid lesion of the medulla oblongata with extension to the cervical spinal cord.

Fig. 3. A hydromyelitic expansion of the central canal of the spinal cord from the C2 to the Th3 level with a maximum diameter of up to 1.53 cm.
and the cystic component with a conventional size of 1.52×1.62×1.22 cm.

MRI of the cervicothoracic spinal cord (February 18, 2022): a hydromyelic expansion of the central canal of the spinal cord from the C2 to the Th3 level with a maximum diameter of up to 1.53 cm was detected. At the level of the C6-C7 intervertebral disc, a focus of paramagnetic accumulation was detected intramedullary on the left with a diameter of up to 0.33 cm (Fig. 3).

Digital selective cerebral angiography verified a hypervascularized round shaped focal lesion at the level of craniovertebral junction (Fig. 4).

Endovascular intervention (July 18, 2022): embolization of hemangioblastoma was performed using liquid embolic agent (Phil) after superselective catheterization of feeding artery which was originated from V-4 segment of right vertebral artery. Control angiograms after intervention demonstrated total obliteration of tumor vascularity and absence of embolic complications (Fig 5).

After surgery the consciousness is clear (15 points on GSC). Pupils D=S, photoreactions are brisk. Swallowing and phonation preserved to the full extent. Hemiparesis on the right side (up to 3 points) and hemihypesthesia on the right side increased.

Surgery (July 19, 2022): removal of the intramedullary neoplasm. Surgical intervention was performed using intraoperative neurophysiological monitoring with such modalities as motor evoked potentials (MEP), somatosensory evoked potentials (SSEP), free run EMG, tEMG. The recording electrodes were placed in the abductor digiti minimi muscle after intubation. M-responses and somatosensory evoked potentials of the right extremity were absent. Given the clinical manifestations and localization of the mass, the most important task of monitoring was the preservation of vital bulbar functions.

Median suboccipital approach with osteoplastic trepanation and partial removal of the posterior semi-arch of the atlas was performed. The superior cervical part of the spinal cord is sharply expanded filling the entire space of the canal. Free fragments of white brittle matter (extravasation of adhesive compositions) were visualized subarachnoidally. A bluish neoplasm is
visualized along the dorsolateral surface of the spinal cord with multiple medium- and small-diameter vessels on the surface, mostly filled with embolized mass, and a large number of dilated venous collectors on the dorsocaudal surface of the neoplasm (drainage neoplasms). Dissection of the arachnoid membrane from the solid part of the neoplasm with the transition to the thinned wall of the cystic mass, located medially from the solid component was performed step by step under the maximum magnification of the microscope without coagulation. During dissection, the contents of the yellow cyst in a volume of up to 4-5 ml were drained. Under maximum microscope magnification (x24), a thin layer of the brain matter was separated from the neoplasm on its dorsal-medial, medial, caudal, and oral surface with gradual transition to the oral-ventral surface of the neoplasm, a dissection of the arteries supplying blood to the neoplasm from the ventral-lateral side was performed. During the circular dissection, the arteries supplying solid part of the neoplasm were found to be completely obliterated by the adhesive composition. After the dissection, the arteries supplying almost avascular tumor were transected. Two drainage veins (one in the oral direction, the other in the caudal-dorsal direction) were found, draining into the spinal veins. The latter were coagulated and transected along the contour of the solid part. The tumor was removed in one block without blood loss. During tumor removal, signs of irritation of the CNX were observed. M-responses at the end of surgery were recorded, but reduced from the CNX on the right. MEP from the upper extremities were without changes. SSEP did not change during the surgery.

Pathomorphological report (29.07.2022): Hemangioblastoma 9161/1 Gr II.

MRI of the brain after surgery (August 10, 2022) (Fig. 7-8): Neoplasm in the projection of the corpus callosum - no changes (Fig. 7). There were postoperative changes in the medulla oblongata on the right in the form of a postoperative cavity of cerebrospinal fluid density measuring up to 0.70×0.70×1.62 cm. There was an area of edema and gliosis around it. CSF pseudomeningocele developed after the surgery.

MRI of the cervicothoracic spinal cord (10/08/2022): hydromyelic expansion of the central canal of the spinal cord from the C2 to the Th3 level has significantly decreased and has a maximum diameter of up to 0.6 cm. At the level of the C6-C7 intervertebral disc, a focus of paramagnetic accumulation intramedullary on the left, with a diameter of up to 0.33 cm was preliminary identified without changes (Fig. 8).

In the early postoperative period, consciousness was clear (15 points on the GSC). Pupils D=S, photoreactions were brisk. Swallowing and phonation were fully preserved. Hemiparesis and hemihypesthesia on the right were without negative dynamics compared to the condition after embolization on 07/18/2022. Impaired sense of urge to urinate and defecate.

After the rehabilitation course within a month, an improvement was recorded in the form of a decrease in hemiparesis. Gait was independent. Strength in the right extremities – 5 points (Fig. 9). He noted the improvement of sensitivity in the right extremities, restoration of sensitivity and the normalization of pelvic organs function.

He had meningoencephalitis with maximal cytosis up to 1749 cells/μL, protein – 1.98 g/L. During treatment, he received colistin, meropenem, gentamicin, ciprofloxacin, rifampicin, flucconazole.

Hemangioblastomas are extremely complex neoplasms due to the risk of bleeding during removal, limitation of blood supply to vital areas of the brain, the need to remove the solid part in one block with a preliminary comprehensive examination and blocking the blood supply of the neoplasm alone, as there are cases of transit involvement of important arterial vessels in

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Fig. 6. Intraoperative neuromonitoring data: M-responses and somatosensory evoked potentials of the right upper extremity are absent. During tumor removal, signs of irritation of the CNX. M-responses at the end of surgery are reduced from CNX on the right. MEP from the upper extremities were without changes. SSEP unchanged during surgery
Fig. 7. Postoperative changes in the medulla oblongata with a postoperative cavity of cerebrospinal fluid density up to 0.70×0.70×1.62 cm

Fig. 8. After surgery, a decrease in the expansion of the central canal of the spinal cord from the C2 to the Th3 level with a maximum diameter of up to 0.6 cm is detected

the hemangioblastoma wall, especially in case of their localization in the parastem or brainstem areas.

The presence of a cystic component in the structure of hemangioblastoma or perifocally, even with partial separation of a solid nodule from vital structures, gives a chance to remove such a neoplasm with functional preservation. Preoperative endovascular removal of a solid fragment of hemangioblastoma is an effective measure that prevents blood loss and greatly facilitates surgical removal, although it is associated with the risk of impaired cerebral circulation in important brain areas, so it should be performed by highly trained personnel with careful evaluation of preoperative angiograms. The second important component of intramedullary hemangioblastoma surgery is maximum possible intraoperative magnification (in our case, x24), to minimize traumatization of adjacent brain areas. Anesthetic management, if necessary, with controlled intraoperative hypotension, and professional multimodal intraoperative neuromonitoring with the maximum possible dynamic function monitoring are also important. Coordinated teamwork of relevant specialists allows the removal of such neoplasms with a satisfactory functional outcome.

As practice demonstrates, multiple intramedullary cysts of the spinal cord distal to the neoplasm are of hydromyelic origin and regress as a result of removal of the neoplasm with a decrease in neurological deficit within a relatively short time after surgery.
Conclusions
Hemangioblastomas with intramedullary growth and the presence of a perifocal cyst can be operated on with a good functional outcome using a comprehensive approach, which involves selective angiography with embolization of vessels supplying the neoplasm alone, intraoperative multimodal neuromonitoring, the use of microsurgical techniques with the maximum magnification of the microscope, and tumor dissection from adjacent brain matter without coagulation.

Information disclosure
Conflict of interest
The authors declare no conflict of interest.
Informed consent
The written informed consent was obtained from the patient.

Fig. 9. Two months after surgery. Complete recovery of limb movement.
References


