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Case Report

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Combined Treatment of Giant Extra-Intracranial Arteriovenous Malformation

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ABSTRACT Arteriovenous malformations are formations consisting of dysplastic, dilated blood vessels in which blood, bypassing the capillary network, enters the venous bed and causes significant expansion of the veins. In addition, arteriovenous malformations can cause nutritional disorders (trophism) of the soft tissues located near them. Extracranial arteriovenous malformations are rare and dangerous vascular anomalies. At present, there are no universal algorithms for their treatment. This article presents the experience of surgical combined treatment of a patient with extracranial arteriovenous malformation. First, staged endovascular occlusion of the formation was performed, and then its complete removal in a hybrid operating room. This approach allowed performing a radical intervention with a positive result.

Keywords: arteriovenous malformation, extracranial malformation, hybrid surgery, combined treatment of AVM

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AVM — arteriovenous malformation
CSAG — cerebral subtraction angiography
CT — computed tomography
DRE — drug-resistant epilepsy
eAVM — extracranial arteriovenous malformation
ECA — external carotid artery
ICA — internal carotid artery
TCT — thyrocervical trunk
VA — vertebral artery

INTRODUCTION

Arteriovenous malformations (AVMs) are formations consisting of dysplastic, dilated blood vessels in which blood, bypassing the capillary network, enters the venous bed, causing significant venous dilation and the development of trophic disorders in the soft tissues surrounding the AVM.

AVMs are usually congenital and tend to increase in size over time, sometimes reaching large sizes. Typical AVM structures are dilated afferent arteries, a malformation tangle with randomly located and in places nodes resembling varicose veins, and efferent dilated veins that experience a load that is inadequate for their natural structure. The reasons for the increase in AVM size are: high arterial blood pressure on the vessel walls in all parts of the AVM, development of collateral vessels, and dilation of all vessels involved in the formation. The processes of angiogenesis (growth of new vessels from the previously existing vascular network) and vasculogenesis (*de novo formation* of the vascular network) also cause an increase in the size and volume of AVMs [1].

The classification of vascular malformations includes 4 main types:

1. Arteriovenous malformations (44–60%).
2. Cavernous malformations (19–31%).
3. Capillary telangiectasias (4–12%).
4. Venous developmental anomalies or venous angiomas (9–10%).

Intracranial AVMs are a "tangle" of vessels with a distinct center (core) that does not contain brain parenchyma, with draining "red veins" containing oxygenated blood at higher pressures than normal veins [2]. The incidence of intracranial AVMs is approximately 1.3 per 100,000 population per year, with a slight male predominance. The average age of

patients diagnosed with an AVM is approximately 33 years, 10 years younger than that of patients with aneurysms [3].

Extracranial AVMs are rare vascular anomalies. AVMs are known to develop in the fetus early in pregnancy and can sometimes be detected at birth [4].

The combination of extra- and intracranial AVMs is an even rarer phenomenon. No precise statistical data on the frequency of their detection have been found in the available literature. There are publications in the form of individual clinical observations.

Currently, there are no classifications and treatment algorithms for patients with extracranial and extra-intracranial AVMs based on multicenter studies, and therefore each clinical case should be carefully studied and used as the basis for further research of this rare but extremely dangerous pathology.

Clinical observation

Patient M., 47 years old, was admitted to the N.V. Sklifosovsky Research Institute for Emergency Medicine on March 6, 2023, as an emergency with repeated bleeding from trophic ulcers of the soft tissues of the head, located above a space-occupying lesion in the cervicoccipital region. It is known from the anamnesis that in November 2000 he received a traumatic brain injury (when falling from his own height, he hit the back of his head), he did not seek medical help, did not undergo examination. Over the next 5 years, the patient noted the appearance of a soft subcutaneous formation in the occipital region. Over the past 2 years, he has noted an increase in the size of the formation. An examination was conducted, an extracranial arteriovenous malformation of the cervicoccipital region (eAVM) was detected. In 2021, he was consulted by an X-ray endovascular surgeon, a neurosurgeon, it was recommended to continue observation. Current deterioration 2 months before hospitalization: EAVM increased significantly, pain in the cervical-occipital region began to bother, trophic ulcers of the scalp appeared. Repeatedly sought medical help due to erosive bleeding from ulcers. Upon admission to the N.V. Sklifosovsky Research Institute of Emergency Medicine, dressing was performed, bleeding was controlled (Fig. 1).

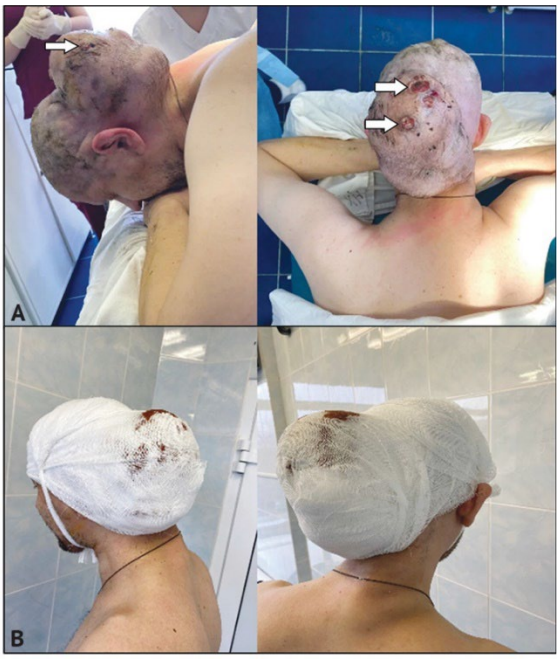

Fig. 1. Patient M. upon admission. A — trophic ulcers in the area of extracranial arteriovenous malformation (indicated by arrows); B — patient's appearance after treatment of trophic changes in the skin and their dressing

A comprehensive examination was performed. Computed tomographic (CT) angiography of the neck and head was performed. A giant extra-intracranial AVM of the cervico-occipital region, areas of ossification and thrombosis in it, pathologically dilated and tortuous afferent vessels were revealed (Fig. 2).

In order to verify the type of malformation, clarify the location of the afferent and efferent vessels and determine the tactics, digital subtraction cerebral angiography (CAG) was urgently performed. Selective angiography of the carotid arteries, vertebral arteries, and thyrocervical trunks (TCT) was performed. The size of the AVM was determined (18 × 12 × 14 cm). The afferent vessels were the branches of the right and left external carotid arteries (ECA), both vertebral arteries (VA), and branches of the TCT. No afferent branches were identified from the basin of the internal carotid arteries. Venous drainage was performed into the dilated superficial saphenous veins, which had nodular expansions, and into the "drain" of the sinuses (Fig. 3).

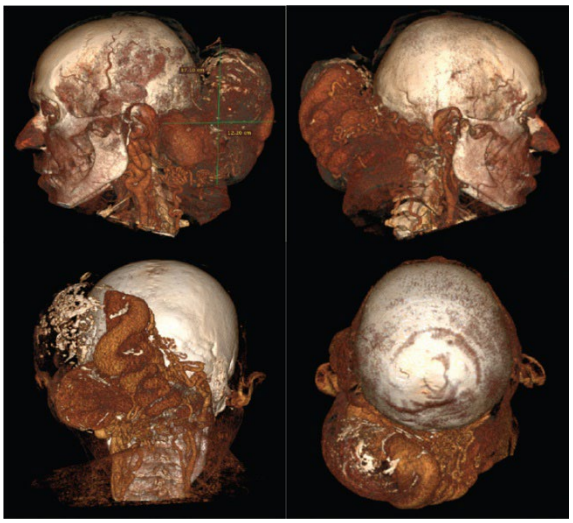

Fig. 2. Computed tomography angiograms of the neck and head of patient M. upon admission to hospital, 3D reconstruction. A giant extra-intracranial arteriovenous malformation of the cervicoccipital region

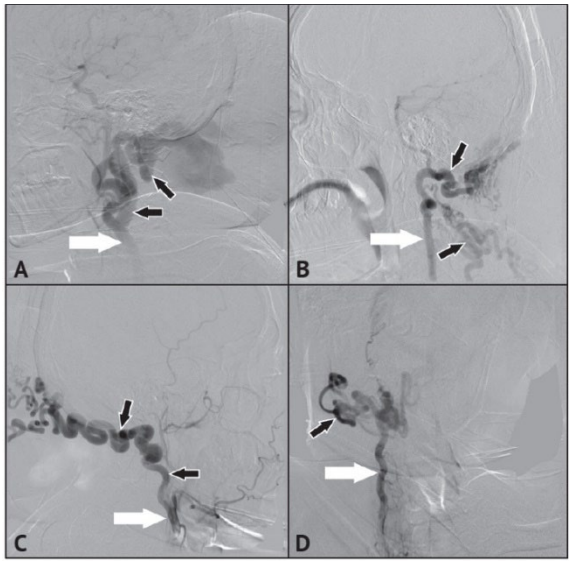

Fig. 3. Cerebral angiograms before surgery (subtraction mode): A — left lateral projection, the white arrow indicates the left external carotid artery and its afferent (black arrows); B - left oblique projection, the white arrow indicates the left vertebral artery and its afferents (black arrows); C - right lateral projection, the white arrow indicates the right external carotid artery, and the black arrows indicate the afferent from it; D - direct projection, the white arrow indicates the right vertebral artery, and the black arrow indicates the afferent from the right vertebral artery

A multidisciplinary consultation was held, consisting of a neurosurgeon, a X-ray endovascular surgeon, a microsurgeon, an anesthesiologist, a transfusiologist, and a therapist. A decision was made to conduct staged combined treatment in the conditions of the hybrid operating room of the N.V. Sklifosovsky Research Institute for Emergency Medicine.

Over the course of 4 weeks, the patient underwent 2 consecutive endovascular interventions with staged occlusion of AVM afferents and one combined intervention in a hybrid operating room.

Considering the high risks of developing hyperperfusion complications during endovascular afferent exclusion, a decision was made to conduct staged treatment.

Optima microcoils , *Amplatzer occluder Plug* II). Embolization of the afferent branch from the left ECA was performed (Fig. 4).

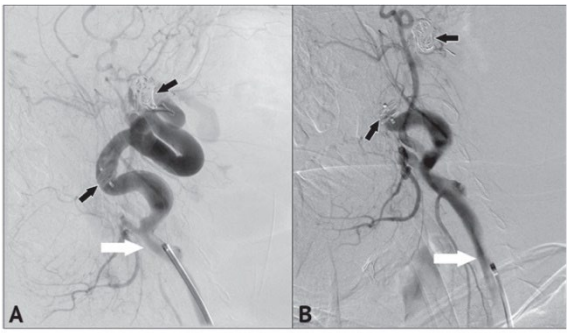

Fig. 4. Cerebral angiograms of the first stage of endovascular treatment (subtraction mode): A — left lateral projection, filling of a large afferent from the left external carotid artery; B — left oblique projection after exclusion of the afferent from the blood flow. The white arrow indicates the external carotid artery, black arrows indicate the occluder and microcoils in the lumen of the afferent

The second stage was performed 7 days later. After a control direct angiographic study, afferents from the right ECA, both VA, and thyrocervical trunks were embolized one by one with non-adhesive material (Fig. 5).

Next, the afferent arterial branches from the right VA and right TCT trunk were visualized and embolized with adhesive material. Fig. 6 shows the process of embolization with adhesive material of 3 pathological branches extending from the right VA.

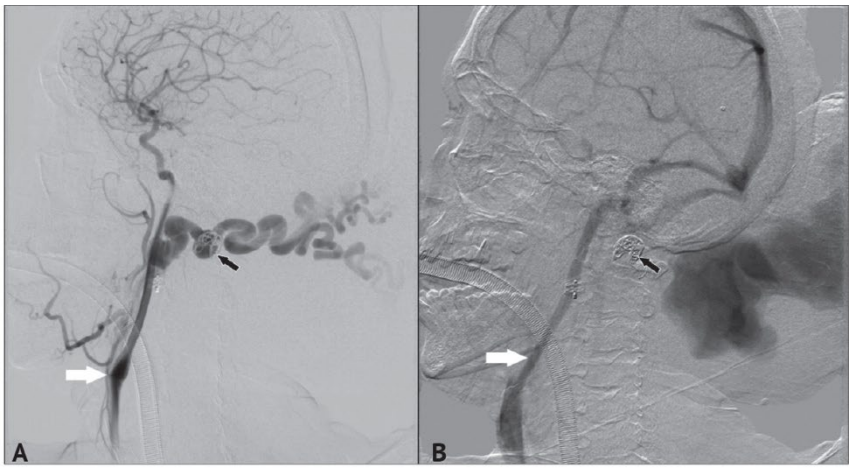

Fig. 5. Cerebral angiograms of the second stage of endovascular treatment (subtraction mode): A — left lateral projection, filling of the extracranial arteriovenous malformation from the right external carotid artery, the white arrow indicates the bifurcation of the right common carotid artery, the black arrow — a microcoil in the afferent from the external carotid artery; B — left lateral projection (late arterial phase), after embolization of a large afferent from the right external carotid artery (the white arrow indicates the right external carotid artery, the black arrow — a microcoil in the lumen of the afferent)

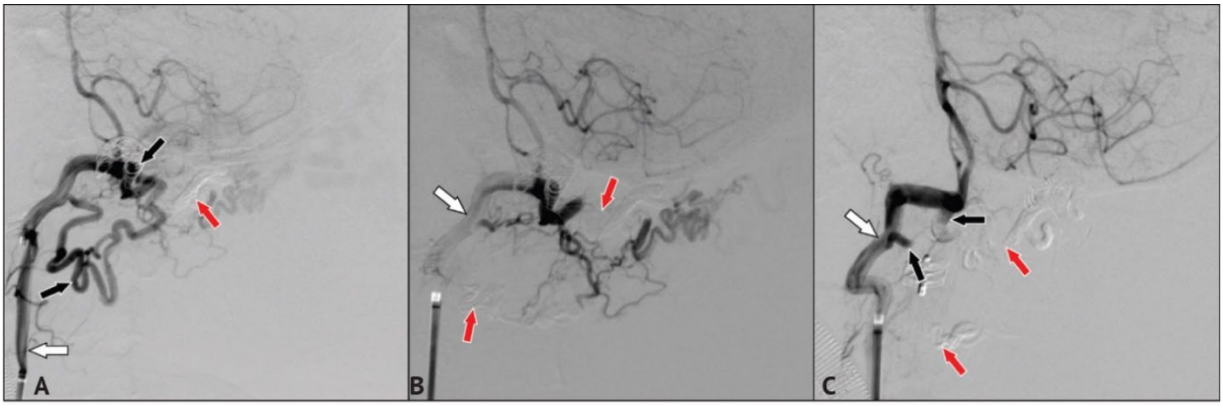

Fig. 6. Cerebral angiograms endovascular embolization afferents from the right vertebral artery (subtraction mode). Three pathological branches supplying blood to the extracranial arteriovenous malformation. The white arrow indicates the right vertebral artery, the black arrow indicates its pathological branches, and the red arrow indicates adhesive material in the lumen of the afferents

Next, the afferents of the left VA and TCT were excluded from the blood flow. Two large high-flow afferents originating from the left VA were visualized, as well as one large-caliber afferent from the left TCT. The afferent of the larger diameter from the VA was excluded with microcoils and adhesive material. After achieving complete reduction of blood flow in it, the remaining afferent from the left VA was embolized with microcoils in the next step (Fig. 7).

Next, visualization of the left TCT was performed using selective angiography, which revealed the distal sections of pathological vessels filling from the left VA and TCT.

A step-by-step closure of afferents from the left TCT was performed. Afferent branches departed from the left TCT by two trunks: smaller and larger caliber. After complete exclusion of the trunk with a larger diameter by adhesive material, a redistribution of blood flow towards the trunk of a smaller caliber was noted, which was occluded by complete embolization of the left TCT.

Despite the fact that the largest afferents of the main arteries of the head (from both ECA, PA and both TCT) were excluded from the blood flow, the blood supply to the EAVM continued to be carried out from the muscular branches of the CA, branches of the TCT, ascending cervical arteries on both sides and the costocervical trunk on the left. Endovascular occlusion of the above-mentioned arteries was not possible due to their small diameter and topographic and anatomical features of their origin from the main arteries. At the same time, antegrade blood flow in all main arteries was preserved (Fig. 8).

Figure 9 shows 3D *reconstructions* of CT angiograms after completion of the endovascular stage of treatment. Dilated terminal branches of the left ECA, which were absent before endovascular occlusion of the afferents (Figure 9A *)* , and *a de novo* formed afferent from the right ECA (Figure 9B *) are visualized* .

Given the high risks of complications after each stage of endovascular treatment of AVM, the patient was in the intensive care unit to monitor vital signs. The risk of hyperperfusion complications was the basis for performing staged treatment, with the time between surgeries being at least 7 days. Cerebral perfusion was monitored using CT perfusion (Fig. 10).

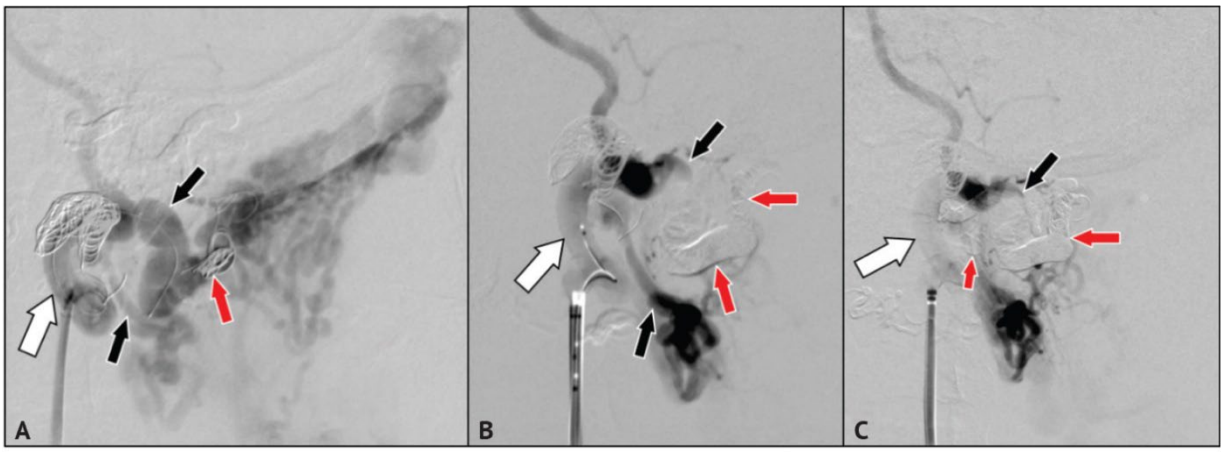

Fig. 7. Cerebral angiograms (subtraction mode): A–C — lateral projections, stages of exclusion from the blood flow of afferents originating from the left branches of the vertebral artery thyrocervical trunk. White arrows point to the vertebral artery, black arrows to the afferents, red arrows to the microspirals and adhesive materia

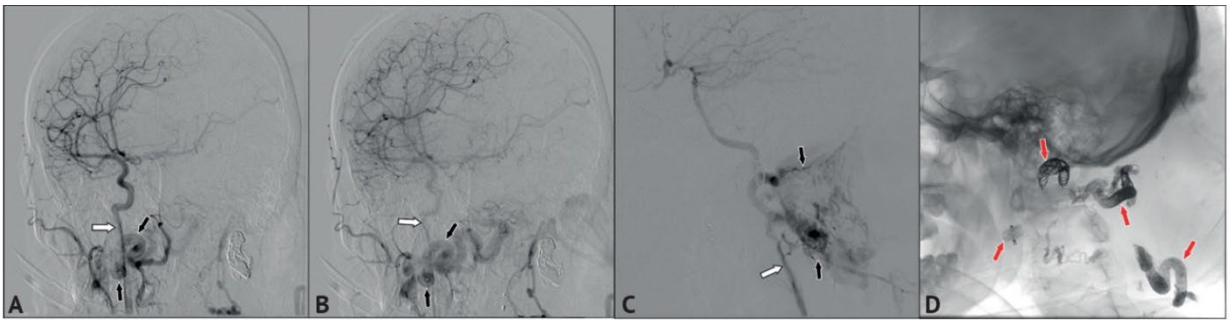

Fig. 8. Cerebral angiograms (subtraction mode): A, B — lateral projection, the right internal carotid artery is visualized (white arrow), de novo formed afferent from the right external carotid artery (black arrows); C — lateral projection, patent left vertebral artery (white arrow) and its muscular branches supplying blood extracranial arteriovenous malformation (black arrows); D — left-sided lateral projection, red arrows indicate occluding materials after all stages of endovascular treatment

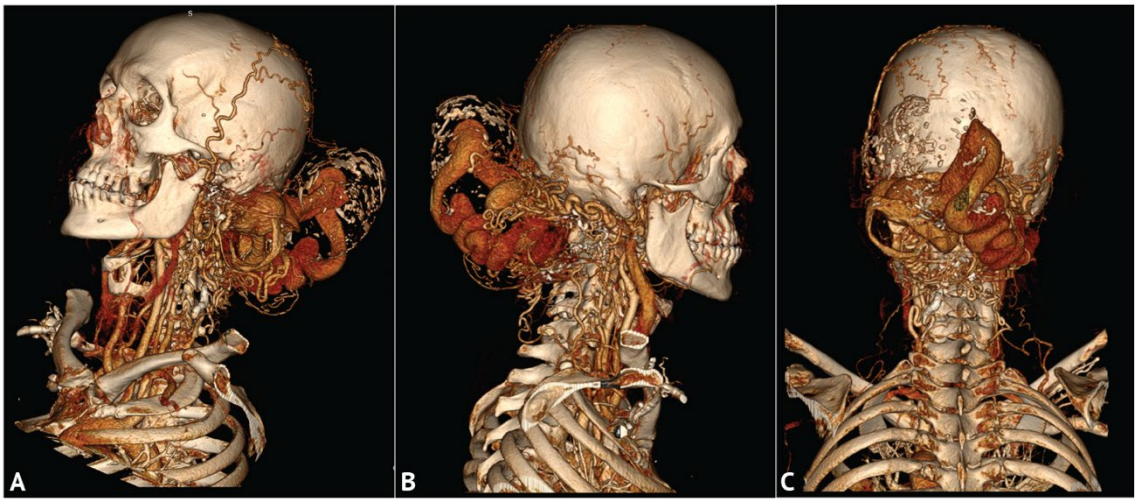

Fig. 9. Different projections of 3D CT angiogram after endovascular stages (A–C)

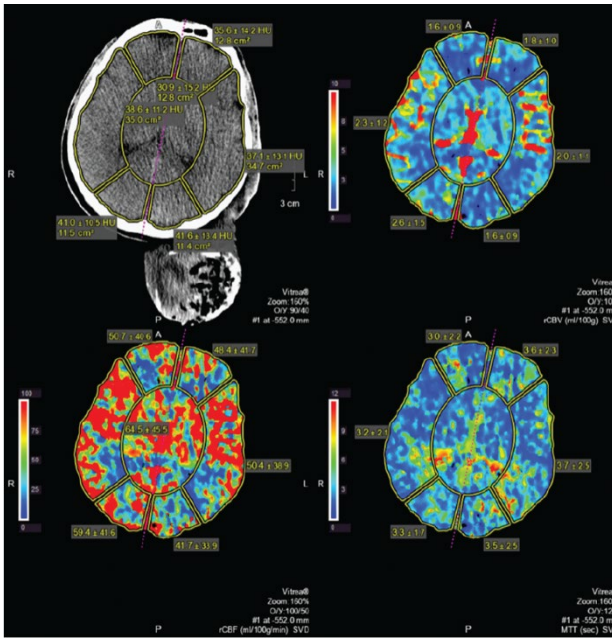

Fig. 10. CT perfusionograms after endovascular stages in CBV (upper right), CBF (lower left) and MTT (lower right) modes. Perfusion parameters are within normal values, interhemispheric asymmetry is not noted

After completion of the endovascular stages of treatment, AVM removal is planned in a hybrid operating room. In order to reduce possible intraoperative blood loss, it is planned to exclude the afferent of the right ECA from the blood flow.

The patient was delivered to the endovascular operating room of the hybrid surgical unit. At the first stage, the instrument was selectively installed in the right ECA, which is the afferent of the AVM with a pronounced fistulous part. Considering the large diameter of the afferent and the high-velocity parameters of the blood flow, a decision was made to implant a vascular occluder. After the occluder was installed, there is no blood flow in the afferent (blocked) (Fig. 11).

After angiographic control and confirmation of the radicality of the occlusion and all large afferents of the EAVM, small muscular branches from the vertebral arteries, from the branches of the sciatic nerve, from the ascending cervical arteries on both sides, from the distal branches of the superficial temporal arteries and the costocervical trunks on the left were visualized. It was impossible to exclude the above-mentioned afferents from the blood flow by embolization due to the anatomical features of their origin, tortuous course and small diameter.

The patient was transferred to the adjacent neurosurgical operating room in a hybrid block (Fig. 12).

The patient is in the prone position, with the head fixed in a Mayfield clamp in the midline position (Fig. 13).

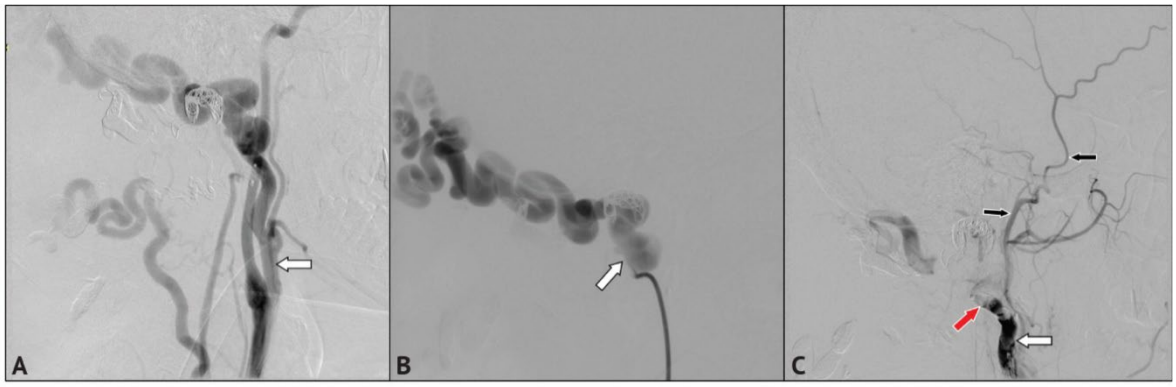

Fig. 11. Cerebral angiograms (subtraction mode): A — lateral projection of the right external carotid artery (white arrow); B — lateral projection, de new afferent from the right external carotid artery (white arrow); C — lateral projection, occluded de novo afferent from the right external carotid artery, the red arrow indicates the adhesive embolisate in its lumen

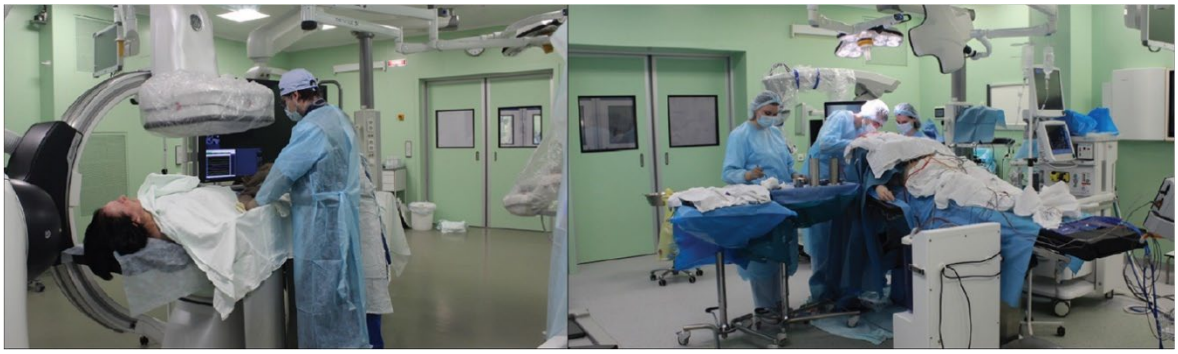

Fig. 12. Photograph of a hybrid operating room, left – angiographic, right – neurosurgical

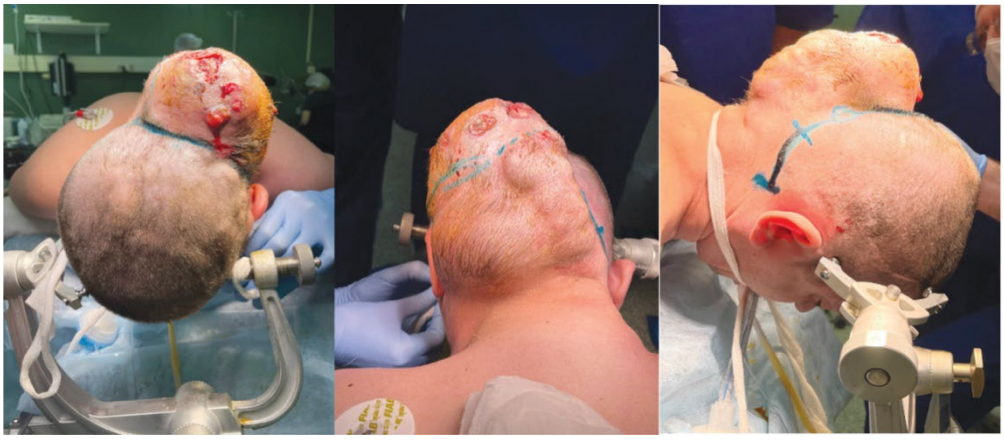

Fig. 13. Intraoperative photographs. General view of extracranial arteriovenous malformation and skin incision lines

Preparation for the intervention began with planning the incisions and corresponding skin flaps, taking into account the location of infected trophic ulcers and thinned skin over the AVM nodes. The prospect of performing autodermoplasty or microsurgical autotransplantation of a complex of tissues was considered, but left as a reserve in case it was impossible to cut out local skin flaps of sufficient area and mobility. Sufficient thickness of the skin over the vessels outside the main AVM node was taken into account, removal of individual vessels was planned while preserving the thickness of the skin over them.

The markings showed that there were enough of the patient's own tissues to perform plastic surgery of the defect.

Surgical access was performed using a T-shaped incision: along the upper and lateral edges of the AVM in the direction from right to left, and from approximately its middle - a perpendicular incision to the level of the posterior arch of the C1 vertebra (Fig. 14). The formed triangular skin flaps ultimately allowed access to the entire AVM.

Ligation and coagulation of small afferent vessels along the perimeter of the AVM were performed. During mobilization from the skull, it turned out that the caverns of the AVM were tightly fixed to the aponeurosis and immobile, partially with areas of coarse fibrosis and ossification. During isolation of the thrombosed fistula, it ruptured, tissue stratification and bleeding occurred. Stopping the bleeding with clamps and vascular clips was unsuccessful. Digital pressure was applied until the end of AVM mobilization. After successful hemostasis (from large venous collectors), the afferent arteries were isolated, coagulated or ligated. Acute intraoperative blood loss was 4 liters over 15 minutes, a blood transfusion was performed. Then 2/3 of the AVM was excised to provide access to its remaining part. Hemodynamic parameters were stable, mean arterial pressure was within 90/60 mm Hg. Isolation and ligation of afferent and efferent vessels was continued. After radical excision of the EAVM, a hole in the occipital bone with a diameter of up to 10 mm corresponding to the sinus drainage was visualized, with moderate venous bleeding, which was stopped using hemostatic materials. Acute massive blood loss was compensated in full, taking into account the hemostasis system parameters, hemodynamic, metabolic, and electrolyte parameters. Intraoperative reinfusion of autoerythrocytes that passed through the leukocyte filter of the *Cell Save* r device was performed. The recorded blood loss, according to *Cell Saver data* , was 4160 ml, the recorded reinfusion was 1015 ml of autoerythrocytes.

The altered skin, trophic ulcers and AVM were removed in a single block (Fig. 15).

The scalp defect was repaired using displaced skin flaps. The excess width of the left skin flap was not excised, as in this case its width would have been insufficient to supply the distal part with blood. The flap edge was de-epidermized, a fold was formed, which was invaginated into the wound, and then sutures were applied. The duration of the open surgical intervention was 13 hours.

After the operation, the patient was transferred to the neurosurgical intensive care unit, where he was transferred to spontaneous breathing without hemodynamic, metabolic, or electrolyte disturbances. The patient was treated in the intensive care unit for the next 5 days. After transfer to the neurosurgical unit, he was activated and dressings were applied. According to the surgical CT and CT angiography data, no pathological or residual areas of AVM were detected (Fig. 16).

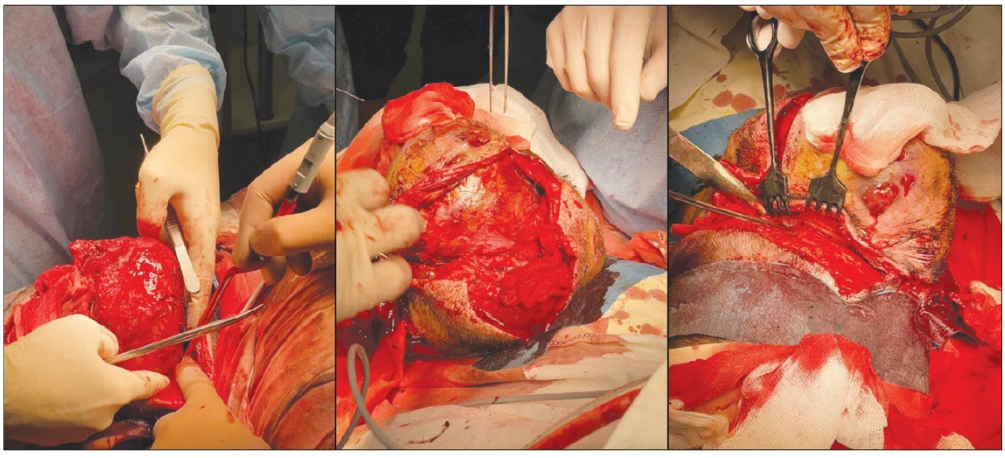

Fig. 14. Stages of skin mobilization and arteriovenous malformation isolation

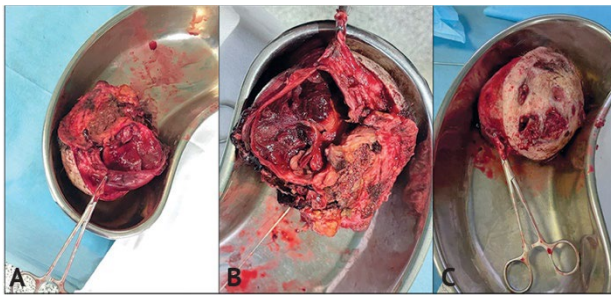

Fig. 15. Intraoperative images of the removed extracranial arteriovenous malformation. A, B — internal surface, C — external surface of the extracranial arteriovenous malformation with trophic ulcers of the skin

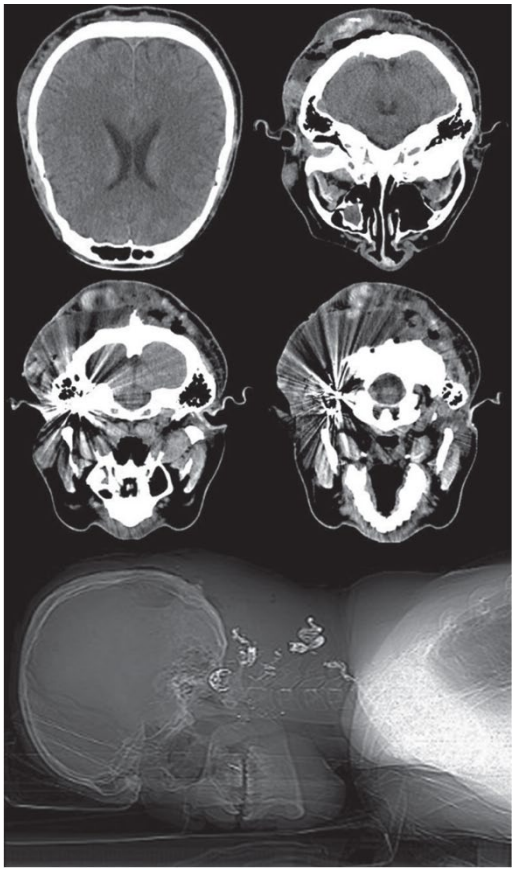

Fig. 16. Computed tomography of the patient's head one day after surgery

The wound healed by primary intention (Fig. 17).

In 1 month after operations patient discharged. Subsequent control studies were performed 6 months after discharge. According to the computed tomography and magnetic resonance imaging of the head, no pathological vascular formations were detected (Fig. 18).

The postoperative scar after 6 months is healthy, without signs of inflammation, without trophic disorders. There are no neurological symptoms (Fig. 19).

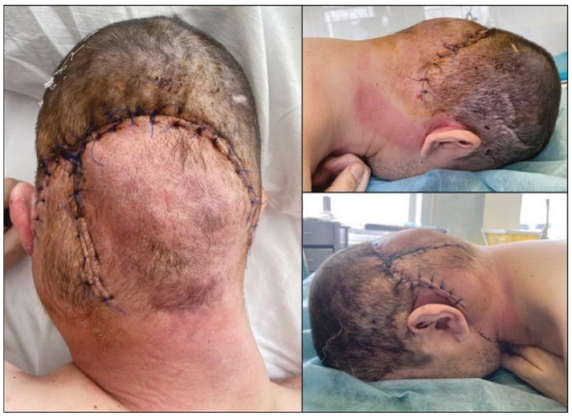

Fig. 17. View of the wound in the early postoperative period. Different projections of the wound

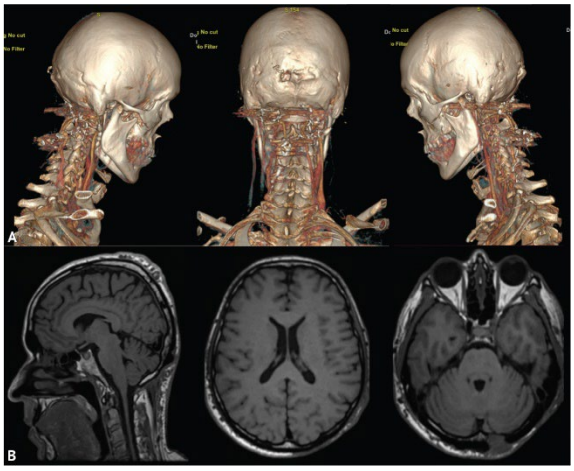

Fig. 18. Control studies 6 months after surgery. A — 3D reconstruction of CT angiograms; B - magnetic resonance imaging of the brain

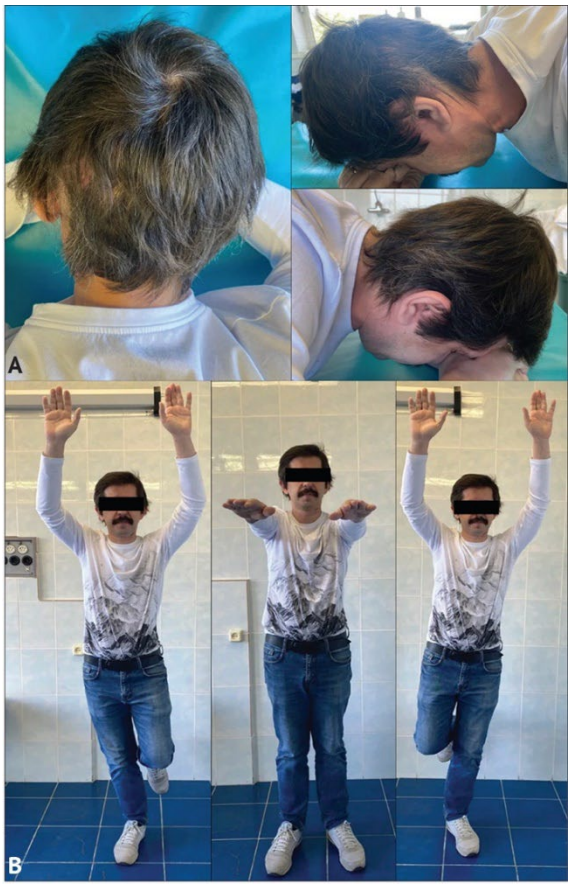

Fig. 19. Patient after 6 months (A, B)

DISCUSSION

The main clinical manifestations of intracranial AVMs are bleeding (58% of cases); in cerebral aneurysms this figure is 92%, convulsive syndrome - in 34% of patients [2]. Focal neurological deficit often develops only after hemorrhage and depends on the localization and volume of the intracerebral hematoma. Like cerebral AVMs, extracranial AVMs also tend to increase in size, which can be affected by head injuries or hormonal changes during puberty or pregnancy [1]. The main local manifestations of EAVMs vary from mild changes in skin color, soft tissue edema to the development of trophic ulcers with a high probability of erosive bleeding. The most common clinical manifestations include pain in the area of the formation, a feeling of pulsation [5]. Esthetic dissatisfaction with extracranial AVMs causes psychological problems.

Due to the rare occurrence of this pathology, the treatment tactics for extracranial AVMs have not been determined. *Lilje D. et al* . suggest taking into account 4 main criteria for choosing tactics in the treatment of patients with AVMs of the head and neck. The protocol includes dynamic and (or) angiographic visualization confirming the presence of AVM and allowing to distinguish it from other vascular formations; a detailed description of conservative treatment; the degree of devascularization of more than 99% for complete cure; observation time of at least one year with radiographic control after treatment [6].

According to most authors, the treatment tactics for extracranial AVMs should be based on determining the stage of the disease and individual characteristics of the angioarchitectonics. In order to assess the stage of the disease, Schobinger [4] proposed a clinical classification of the stages of peripheral AVMs (table).

According to the authors, a more aggressive approach to treatment (endovascular, surgical) should be considered when AVM progresses and moves to the next stages.

Extra-intracranial AVMs, unlike intracranial ones, tend to continuously increase. This is stated by *ZouYun et al* . in a retrospective study including 446 patients (2011–2017) with AVMs. In 76.7% of patients, AVMs were localized extracranially (in the head and neck area). Thus, in children with stage I according to Schobinger, the risk of progression to adolescence was 41.9%, to adulthood — 80% [7]. The authors report the effective use of Bleomycin as a conservative component in the treatment regimen.

In a meta-analysis, *Lilje D. et al* . [8] analyzed data in *the PubMed* , *Embase* , *the Cochrane Library* , and *scholar.google.com databases* from 2000 to 2020 on interventional therapy for EAVMs. The results of the meta-analysis show that intra-arterial ethanol embolization is highly effective in the treatment of EAVMs of the head and neck located in bone tissue. The authors note that there is no standardized reporting system for the diagnosis, treatment, and outcome of extracranial AVMs in the modern literature. Despite a significant number of publications on this topic (n = 151), the authors concluded that to date there is no convincing evidence of an effective interventional treatment strategy.

Surgical treatment of AVM is the most effective. However, not all AVMs can be resected. AVM embolization is an alternative method that is less radical but more flexible in terms of repeated interventions. The combined treatment method

includes endovascular and open stages of surgery. Such interventions are conveniently performed in hybrid operating rooms. Radiosurgical irradiation (γ -irradiation) is considered an effective method for treating intracranial AVMs up to 3.5 cm in size.

Overall mortality among patients with intracranial AVMs can reach 30% and depends on the location and size of the pathology. Surgical mortality after endovascular embolization averages 4.3%, and after resection of small AVMs (*Spetzler–Martin* 1-2) — 1.7%, in the absence of treatment — 10.1%. The combined indicator "stroke + mortality" for any variant of the disease course is 30.7% [9]. Similar mortality data for patients with eAVMs are not available in the literature.

Most AVM recurrences occur within the first year after the intervention. According to *AS Liu* , 98% of AVMs after embolization and 86.5% after resection with or without embolization recurred within 5 years. The reasons for AVM re-expansion after treatment are probably similar to the reasons for its natural progression: an increase in the number of collateral vessels, vascular dilation, and/or the formation of a new vascular network. Partial resection or embolization of AVM are factors for AVM growth and rupture. Excision implies a traumatic impact, which in turn provokes neoangiogenesis and an increase in the size of the residual part of the vascular malformation. Embolization results in local hypoxia and an increase in hypoxia-inducible factor-1 〈, drug-resistant epilepsy (DRE), vascular wall fibrosis markers MMP-2 and MMP-9, and a decrease in the antiangiogenic tissue inhibitor of matrix metalloproteinase. AVM resection also causes local hypoxia and inflammation, which stimulates angiogenesis through the expression of DRE, basic fibroblast growth factor, and the production of MMP-2 and MMP-9 by platelets, neutrophils, and macrophages [1].

When determining the treatment tactics for EAVM, it is necessary to take into account the size and location of the AVM, the patient's age, and the stage of the disease according to the Schobinger classification. Patients with stage I arteriovenous malformations require dynamic observation by a physician. The decision to perform surgical intervention for stage II lesions depends on the symptoms and location of the AVM. The presence of stage III and IV arteriovenous malformations is an indication for surgery. Large AVMs affecting several areas of the head and neck are usually treated endovascularly. Treatment of such AVMs is often staged. Open resection of AVMs should be performed in the early stages of the disease (stages I and II according to Schobinger). For extensive lesions, combined treatment is recommended - embolization and excision [1].

Rapid increase in the size of the AVM against the background of conservative therapy or non-radical treatment methods becomes the reason for the patient to seek surgical help late. In this case, a combined treatment method and a multidisciplinary approach become a priority in choosing treatment tactics. The first stage is embolization of the formation, the second stage is removal. To reduce the exposure time between endovascular and open intervention and to prevent AVM progression during its partial embolization, it is convenient to use hybrid operating rooms. This strategy increases the duration of the operation, but allows for radical removal of the formation.

The mechanisms and pathophysiological features of extra-intracranial AVM growth require further study. Some drugs can slow AVM progression [10], so the search for new agents that cause AVM stagnation and the use of combined surgical methods in a hybrid operating room will improve disease outcomes.

CONCLUSION

In the presented clinical observation, we used the tactics of staged endovascular occlusions of arteriovenous malformation with its subsequent total removal in a hybrid operating room. Despite the maximum possible embolization of afferent arteries, such an intervention is associated with a high risk of intraoperative blood loss and complications. Careful preoperative planning and a multidisciplinary approach allowed us to perform a radical intervention with a positive result.

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