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Direct spontaneous high-flow carotid-cavernous fistula in a 9-month-old child after nasolacrimal duct repair

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ABSTRACT

Successful treatment of direct spontaneous carotid-cavernous fistula in a 9-month-old child and review of the literature devoted to this issue are reported in the manuscript. Spontaneous high-flow carotid-cavernous fistula is extremely rare in children. Detailed diagnosis of the fistulae includes CT-angiography and cerebral subtraction angiography. Previous surgeries (nasolacrimal canal repair in this case) may be considered as an etiological factor of carotid-cavernous fistula. Transarterial transfemoral endovascular embolization of the fistula with microcoils ensured closure of abnormal communication and complete regression of clinical manifestations in our case.

Keywords: carotid-cavernous fistula, spontaneous fistula, lacrimal canal repair, endovascular embolization, childhood.

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Abbreviations

CCF — carotid-cavernous fistula
CT — computed tomography
MRI — magnetic resonance imaging
ICA — internal carotid artery
CS — cavernous sinus
ECA — external carotid artery
DAVF — dural arteriovenous fistula

Congenital spontaneous direct carotid-cavernous fistula (CCF) is extremely rare in childhood and infancy. In the vast majority of cases, high-flow CCF is a consequence of traumatic brain injury. We report successful endovascular treatment of direct spontaneous CCF in a 9-month-old child. Moreover, the review of this pathology in children is presented.

Case report

The child was born from the 3rd pregnancy (the 3rd delivery). Childbirth was urgent at 39 weeks. Child's weight at birth was 4100 g, Apgar score 8—9. The child was on natural feeding. Cranial trauma, hereditary diseases and connective tissue diseases were absent.

The parents noted tears in the left eye at 3 months of age. Left-sided contrast-enhanced dacryocystography was performed. Contrast enhancement included lacrimal sac and superior part of the vertical segment of nasolacrimal duct. There was no leakage of the contrast agent into the nasal cavity. Attempts to rinse the nasolacrimal canal were ineffective. Considering no positive changes, nasolacrimal canal probing was carried out on 08.08.2017. The procedure was ineffective too. Laser reconstruction of the left nasolacrimal canal was performed щп August 22, 2017. Spontaneous removal of the bougie occurred 3 weeks later. The desirable effect of surgery was not achieved. Ophthalmologist recommended follow-up within 1 month. Left eyelid edema, conjunctival swelling, and moderate vein enlargement on the skin of the eyelid and face have occurred since October 2017. Ophthalmologist confirmed anatomical integrity of the left orbit. Subsequent negative changes developed in December 2017 (left-sided exophthalmos, tortuosity of the veins of both eyelids and upper left facial half, aggravation of conjunctival hyperemia on the left).

MR-angiography revealed abnormal enlargement of the left cavernous sinus within parasellar area on the left. Moreover, dilated veins were observed in the projection of the left orbit (**Fig. 1a, b**).

CT-angiography revealed fistula between the cavernous segment of the left internal carotid artery (ICA) and cavernous sinus (CS). The dimensions of cavernous sinus were 9×8×6 mm. There was venous enlargement in the left orbit and mediobasal parts of the left temporal lobe (**Fig. 2a, b**).

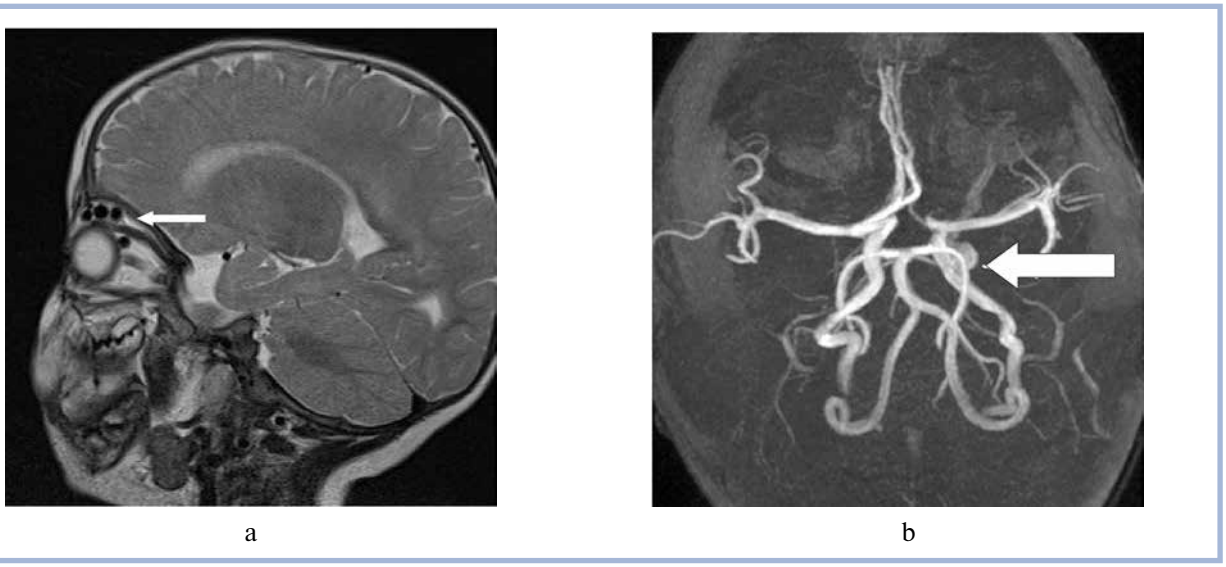
According to selective cerebral angiography, there was a direct high-flow CCF type A by D.L. Barrow [1] with predominant blood drainage into superior ophthalmic vein (**Fig. 3**).

Transfemoral transarterial endovascular embolization of the cavernous sinus by microspirals was preferred in this child. The surgeon determined this surgical approach. The intervention was carried out under endotracheal anesthesia. Nine microspirals were sequentially placed into the aneurysm (**Fig. 4**). Control angiography confirmed no contrast enhancement of the fistula, major vessels were patent (**Fig. 5**). Regression of clinical manifestations of CCF was observed in early and late postoperative period (**Fig. 6, 7**).

The feature of this report is congenital obstruction of nasolacrimal duct in the patient's older brother. The disease was cured at the age of 3 months. There were no clinical manifestations of CCF in the older brother.

Discussion

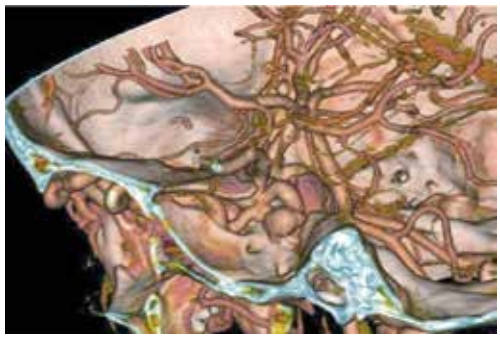
Abnormal communication between the CS and intracavernous segment of ICA (carotid-cavernous fistula)



a b

Fig. 1. **Preoperative MRI.**

a — MRI of the brain, enlarged superior ophthalmic vein in the left orbit (arrow); b — 3D TOF MRI, enlarged vein within the cavernous sinus on the left (arrow).



a



b

Fig. 2. **Preoperative CT-angiography.**

a — CT of intracranial vessels, enlarged superior ophthalmic vein within the cavernous sinus on the left is visible; b — CT of extracranial vessels, enlarged superior ophthalmic vein with multiple anastomoses is visible.



Fig. 3. **Preoperative selective cerebral angiography, direct and lateral projections.**

Enlarged superior ophthalmic vein (arrow).

is classified into 4 types (A, B, C, D) by D.L. Barrow [1]. Type A is a high-flow fistula with direct shunt through the ICA wall defect. Other variants include indirect shunt from the meningeal branches of ICA and/or external carotid artery (OCA) into cavernous sinus. These types are united under one name — dural arteriovenous fistulae (DAVF) into CS. DAVF are the most common among spontaneous fistulae.

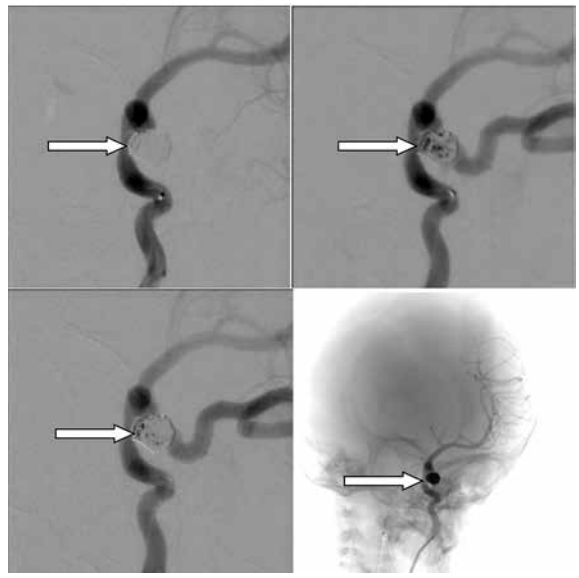


Fig. 4. **Stages of endovascular surgery — consecutive introduction of microspirals into the fistula.**

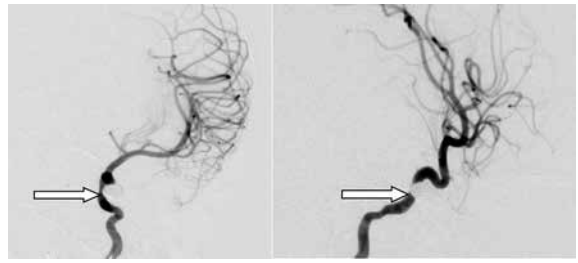


Fig. 5. **Direct selective cerebral angiography after surgery.**

The microspirals placed in the cavity are indicated by the arrow.

The vast majority of direct fistulae have a traumatic genesis and are more common in young men. Spontaneous CCF type A are generally rare and extremely rare in children. Few case reports of congenital cavernous fistula especially type A are reported in the literature. One of the first mentions of this pathology is in the book "Annals of Surgery" dated 1924 [2]. The authors describe spontaneous exophthalmos in 38 patients including 2 (5.3%) children. In one case, a 13-year-old boy suddenly developed a right-sided pulsatile exophthalmos, headache, vomiting, chemosis and eyelid vein dilation. Examination of the ocular bottom revealed swelling and unclear borders of the optic disc. Ligation of the right ICA and right external jugular vein was performed. The patient died 9 days later. Autopsy verified arteriovenous fistula between the cavernous sinus and cavernous segment of ICA. Spontaneous bilateral pulsatile exophthalmos was observed in another 5-year-old patient. Carotid artery compression was not successful. Ligation of the left carotid artery was ineffective too. Further fate of the patient was



Fig. 6. **A child in 2 days after surgery. Moderate swelling of the upper left eyelid.**



Fig. 7. **A child in 3 weeks after surgery. No asymmetry of the eye fissures, no eyelid edema.**

not described. It is not clear what fistulae were diagnosed in the described patients.

A.W. Biglan et al. (1981) reported over 30 cases of spontaneous CCF including a 7-week-old infant with DAVF into CS [3]. Y. Konishi et al. (1990) reported 12 cases of CCF in children. The authors emphasized no direct fistulae in these patients [4].

S.B. Yakovlev et al. (2008) analyzed follow-up data and treatment of 358 patients with direct CCF [5]. Traumatic fistula was observed in 317 (88,5%) patients, spontaneous fistulae – in 38 (10,6%) cases including 1 (0,3%) child with congenital fistula. We found 4 case reports of direct spontaneous CCF type A in available literature [2, 6—8].

M.D. Gossman et al. (1993) first described spontaneous CCF type A in a 5-year-old child [6]. The authors applied a combined treatment strategy. At the first stage, CS was occluded by a separable balloon-catheter that did not result total occlusion of the fistula. At the second stage, intravenous part of the left ICA was additionally clipped proximal to the left ophthalmic artery ostium. Extracranial part of the left ICA was embolized by a muscle and ligated.

T. Ansaar et al. reported direct spontaneous CCF type A in a 11-month-old child after surgical treatment of congenital obstruction of the nasolacrimal duct [7]. Endovascular treatment ensured complete occlusion of the fistula. It is noteworthy that the authors, as well as we, do not associate previous surgical treatment of nasolacrimal duct disease with the development of direct CCF. Transarterial approach is the most common in endovascular treatment of CCF. However, G.B. Mercado et al. (2011) preferred transvenous approach for embolization of direct spontaneous CCF in a 3-year-old patient [8]. The authors emphasized similar safety and effectiveness of transvenous and transarterial endovascular occlusion of CCF.

Conclusion

Spontaneous direct caroticocavernous fistula is extremely rare in childhood. There is no evidence of correlation between CCF and previous surgical treatment of the lacrimal duct obstruction. However, follow-up of patients with this disease is desirable for the timely diagnosis of CCF and angiographic examination. Endovascular intervention with separation of CCF by microspirals is currently preferred for this pathology.

Authors' participation:
Concept and design of the study — A.D.
Collection and analysis of data — A.N., T.I.
Writing the text — A.N.
Editing — A.D.

The authors declare no conflicts of interest.

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Commentary

The authors report a rare case of direct caroticocavernous fistula in a 9-month-old child after previous nasolacrimal duct surgery. There are few literature data on non-traumatic direct arteriovenous fistulae in children that makes this clinical case unique. The authors have formed an adequate complex of preoperative examination that ensured timely correct diagnosis. Endovascular closure of caroticocavernous fistula was preferred. It should be noted that reconstruction of major cerebral vessels is advisable in children considering growth of the organism. Transarterial occlusion of pre-fistulous part of cavernous sinus by microspirals ensured patency of internal carotid artery and occlusion of the fistula. The manuscript is perfectly illustrated by clinical data and neuroimaging results.

The authors discuss the key issue of correlation between previous nasolacrimal duct surgery as an etiological factor and development of the fistula. Various causes of spontaneous caroticocavernous fistulae are described in the literature. They include various conditions associated with vascular wall weakness. These are various types of collagenopathies, fibromuscular dysplasia, rupture of cavernous aneurysm. Certain cerebrovascular abnormality in this child may be suspected considering hypertrophy of the middle cerebral artery, which is clearly visible on baseline angiograms. Its diameter is similar to that of internal carotid artery. Rupture of small aneurysm of cavernous segment of internal carotid artery is less likely but possible although it is impossible to verify this fact in case of patent fistula. Thus, we cannot associate CCF with previous nasolacrimal duct surgery because these procedures could not be followed by injury of the orbital and skull base structures. So, I should agree with the opinion of T.R. Ansaar et al. too. The choice of transarterial or transvenous approach in the treatment of caroticocavernous fistula depends on localization and shape of the defect in ICA wall, anatomical peculiarities of cavernous sinus and state of outflow pathways. Nevertheless, the purpose of surgery is to occlude the fistula from the side of cavernous sinus and preserved patency of internal carotid artery regardless of the approach and endovascular implants. Combined approach may be used in the most difficult cases.

We can congratulate the authors with successful operation in such a small patient while this case report will be valuable for analysis of endovascular treatment of vascular diseases in children

S.B. Yakovlev (Moscow, Russia)