Post-traumatic carotid cavernous fistula of late manifestation report

Późna manifestacja pourazowej przetoki szyjno-jamistej – opis przypadku

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Słowa kluczowe: przetoka szyjno-jamista, wytrzeszcz, uraz głowy.

Abstract

Carotid cavernous fistula (CCF) is an abnormal connection between the carotid artery and the cavernous sinus. Carotid cavernous fistula of traumatic aetiology occurs more commonly. The characteristic triad of symptoms covers the following: pulsating exophthalmos, a humming sound within the skull, and dilation and tortuosity of conjunctival and episcleral veins. The diagnosis of CCF may constitute a diagnostic problem in the situation when the symptoms occur several weeks after injury, it may overlap with other post-traumatic changes, and the dominant symptom may be cranial nerve palsy. The lack of a correct diagnosis and adequate causative therapy creates the risk of not only loss of vision, but also of life. We present a case of a patient who developed fistula symptoms a few weeks after an injury. The diagnosis of post-traumatic CCF was confirmed by imaging examinations. The application of transluminal embolisation led to the resolution of the majority of symptoms.

Streszczenie

Przetoka szyjno-jamista to nieprawidłowe połączenie pomiędzy tętnicą szyjną a zatoką jamistą. Najczęściej spotyka się przetokę szyjno-jamistą o etiologii urazowej. Charakterystyczna triada objawów obejmuje: pulsujący wytrzeszcz gałki ocznej, silny szum w głowie, poszerzenie i krętość naczyń żylnych spojówki i nadtwardówki. Rozpoznanie pourazowej przetoki szyjno-jamistej może stanowić problem w sytuacji, gdy objawy pojawiają się po kilku tygodniach od urazu, nakładają się na inne zmiany pourazowe lub podstawowym objawem jest porażenie nerwów czaszkowych. Brak prawidłowej diagnozy i odpowiedniego leczenia przyczynowego stanowi ryzyko nie tylko utraty wzroku, lecz także życia. Przedstawiamy przypadek pacjenta, u którego objawy przetoki wystąpiły po kilku tygodniach od urazu. Rozpoznanie pourazowej przetoki szyjno-jamistej potwierdziły badania obrazowe. Zastosowane leczenie metodą przeznaczyniowej embolizacji spowodowało wycofanie się większości objawów.

Introduction

Carotid-cavernous sinus fistula (CCF) is an abnormal connection between the cavernous sinus and the internal carotid artery, or the branches of the external or internal carotid arteries [1].

Fistulas are classified based on anatomy, haemodynamic properties, or aetiology. Anatomical fistulas are divided into direct and indirect types. Direct fistulas are a connection between the internal carotid artery and the cavernous sinus. Indirect fistulas concern connections between the branches of the internal or external carotid arteries and the cavernous sinus [1]. Commonly used classification of fistulas, according to Barrow based on angiographic images, divides CCF into four types: direct fistulas are type A, while types B, C, and D are indirect fistulas [2].

The type of fistula is translated into its haemodynamic properties. High-flow and low-flow fistulas are distinguished. High-flow fistulas are those where carotid arterial blood out-flowing under high pressure from the damaged internal carotid artery enters directly the cavernous sinus, which in most cases results in a rapid occurrence of symptoms. Low-flow fistulas, which are the connections between the cavernous sinus and the branches of the carotid internal or external arteries, are characterised by a slow blood flow and subtle clinical symptoms [1].

Based on aetiology, CCFs may be divided into post-traumatic and spontaneous [1]. Post-traumatic CCFs constitute 70–90% of cases, concern primarily young males, and are the result of road accidents, mainly motorcycle accidents [3]. These fistulas are most often unilateral, although there occur cases of bilateral injury of the internal carotid artery in its intracavernous segment [4]. Post-traumatic fistula usually develops as a result of basal skull fracture, especially of the sphenoid bone; sporadically the development of a fistula is the result of injuries penetrating through the eye socket [5].

Spontaneous carotid-cavernous fistulas occur considerably more rarely, and are usually caused by rupture of an aneurysm of the internal carotid artery in its intracavernous segment, or the same artery when it is atherosclerotically changed, or weakened as a result of concomitant connective tissue disorder (Ehler-Danlos syndrome) or fibromuscular dysplasia [1, 3, 8].

Clinical symptoms and the dynamics of their manifestation are directly related with the aetiology, localisation, and haemodynamic properties of the fistula.

Post-traumatic fistulas are most often direct, high-flow fistulas equivalent to type A, according to the classification by Barrow [2]. Increased pressure inside the cavernous sinus caused by the inflow of arterial blood leads to changes in vascular out-flow, which is translated into the pace of development and the scope of the clinical symptoms observed. Physical symptoms usually occur on the same side as the fistula, but may also be bilateral or even contralateral, as a result of vascular connections. The disease manifests itself within several days or weeks in the form of a classic triad: pulsating exophthalmos, dilation and tortuosity of conjunctival and episcleral veins, and a humming sound within the skull [1].

A case of a patient is presented who developed full symptoms of fistula several weeks after injury – the use of imaging diagnostics, i.e. computed tomography (CT) and magnetic resonance (MR), allowed the confirmation of the carotid-cavernous sinus fistulas. Treatment with transluminal embolization led to the resolution of the majority of the symptoms.

Case report

A 59-year-old male was afflicted with craniocerebral trauma as a result of a road accident. During hospitalisation in the Surgical Ward at the Provincial Hospital, skull fracture, the presence of intracerebral haematoma, and brain concussion were diagnosed using imaging CT examination (CT scan) of the head. Conservative treatment was applied. The patient was discharged home in a good general state, without deviations in the neurological assessment. Three weeks after the injury, the patient reported to the Hospital Emergency Unit of the Provincial Hospital due to exophthalmos of the right eyeball. In ophthalmological examination an oedema of the tissues of the right eye socket was diagnosed, drooping of the upper eyelid, and limited mobility of the right eye (RE) in all directions; examination of the fundus of the right eye did not show any abnormalities. Steroid therapy was applied. In the check-up CT scan of the skull performed two days later, the evolution of post-traumatic changes was observed and fissure fracture of the right temporal bone passing through the base of the middle cranial fossa covering the sphenoidal sinus. After a check-up at the Ophthalmology Consultation Department the patient was referred in emergency mode to the Maxillofacial Surgery Ward. During hospitalisation, the patient reported a humming sound in the eye socket region. In physical examination, changes were observed concerning the right eyeball, these were pulsating exophthalmos, drooping of the eyelid, loss of mobility of the eyeball and, dilated pupil poorly responding to light.

Computed tomography of the eye sockets with contrast was performed in emergency mode. The examination revealed a widened lateral straight muscle of the right eyeball, a wider non-uniform right optic nerve in the posterior segment of the eye, expansion of the right ophthalmic vein, hyperdensive cavernous sinus on the right side (Figures 1 and 2). In laboratory tests performed, no increase was observed in the concentration of inflammatory markers (C-reactive

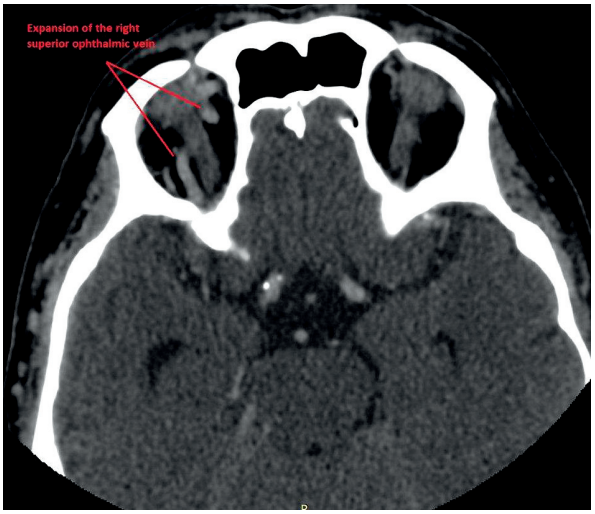

Figure 1. Computed tomography. Dilatation of the right superior ophthalmic vein

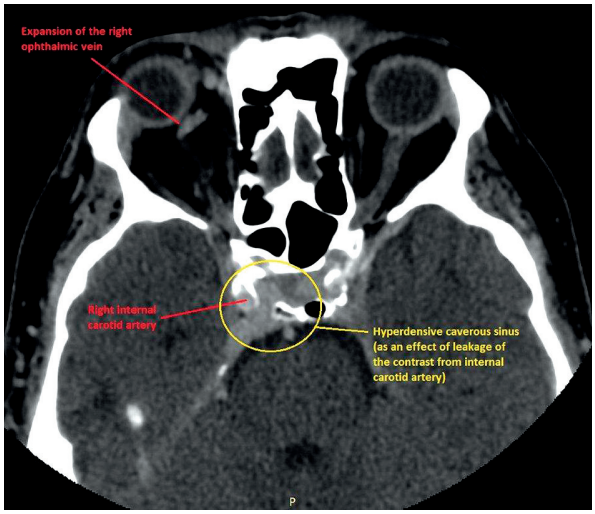

Figure 2. Computed tomography. Hyperdensive cavernous sinus

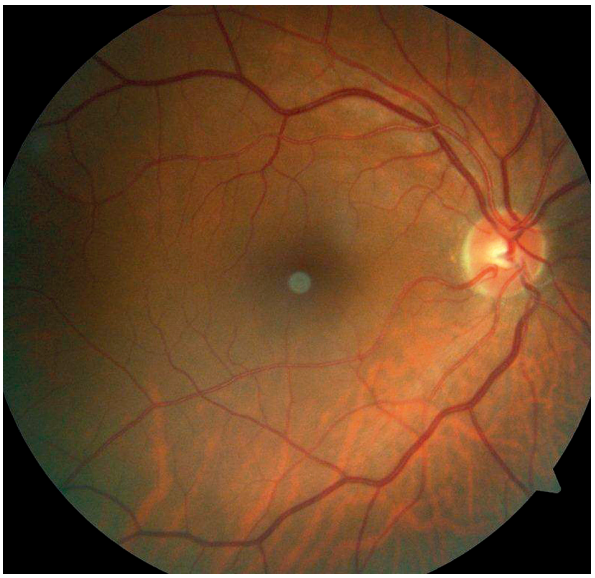

Figure 3. Fundus of the eye. Slightly dilated veins

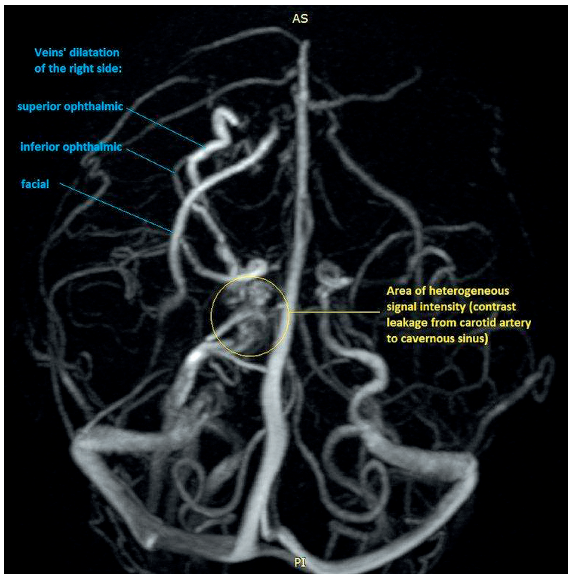

Figure 4. Angio-MR. Contrast leakage from right internal carotid artery to cavernous sinus

protein (CRP) 9.5 mg/l) and coagulation parameters (D-dimer 708 ng/ml). Steroid therapy was implemented, antithrombotic treatment and antibiotic therapy, without improvement of the clinical state. The patient was transferred to the Regional Hospital for ophthalmological consultation. In the physical examination a normal acuity of vision was observed (Vo.d.s = 1.0), a raised intraocular pressure in the right eye (To.d = 24 mm Hg), normal pressure in the left eye (To.s = 14 mm Hg); deviations from normal concerned the right eyeball: pulsating exophthalmos, loss of mobility of the eyeball, drooping of the upper eyelid, extravasation and oedema of the conjunctiva, dilated pupil lazily responding to light; in the image of the fundus of the eye, apart from slightly dilated veins, no important abnormalities were found (Figure 3). The patient reported humming in the skull, which disappeared while pressing the right carotid artery. Data collected while taking medical history, and clinical symptoms observed during examination, suggested the presence of the carotid cavernous fistula. The patient had MR angiography performed which showed dilation of the ocular vein, intraocular blood vessels, cavernous sinus, and expanded face vessels on the right side (Figure 4). The MR was equivalent to the image of the carotid cavernous fistula. The patient was transferred to the Clinic of Neurosurgery where an embolisation procedure was performed obtaining total closure of the fistula. After the procedure, a decrease was observed in oedema and exophthalmos, persistent palsy of nerve III in the form of loss of mobility of the eyeball, drooping eyelid, and expanded pupil of the right eye. In an ophthalmological check-up examination performed 3 months after embolisation, resolution of

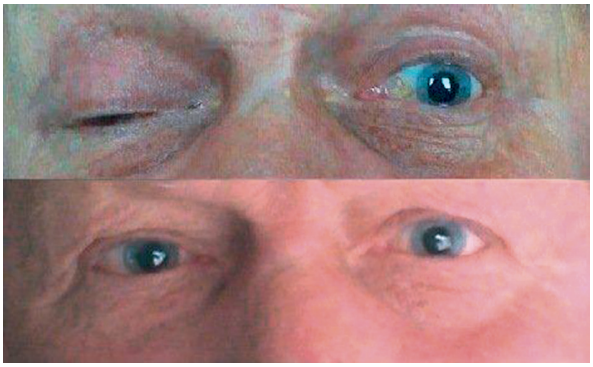

Figure 5. Patient before and after treatment

exophthalmos was observed along with full mobility of the eyeball, normal acuity of vision (Vo.d.s = 1.0), normalisation of intraocular pressure (To.d. = 14 mm Hg, To.s = 12 mm Hg), and the resolution of changes concerning the anterior and posterior segments related with blood stasis. The pupil remained slightly expanded. Examination of the visual field did not show any deficits. The patient mentioned regression of the humming sound heard previously and did not report any complaints on the part of the organ of vision (Figure 5).

Discussion

Knowledge of the symptomatology of the carotid cavernous fistulas, despite their rare occurrence, has important implications with respect to further management, treatment and prognoses.

Post-traumatic changes and symptoms of CCF may have a similar clinical image or co-occur, which

considerably hinders diagnosis. Damage of the bones of the eye socket, development of orbital haematoma, retrobulbar haematoma, or orbital oedema may cause exophthalmos imitating the exophthalmos present in the direct carotid cavernous fistula. In the majority of cases, lack of pulsation of the protruding eyeball, as well as the lack of a humming sound heard by a patient, are decisive. In the case of fractures concerning the upper eye socket bone, there may occur pulsation of the eyeball, resulting from the transmission of pulsation of the cerebrospinal fluid; however, such a pulsation is not accompanied by vascular murmur [1]. Post-traumatic oedema of the conjunctiva, as well as subconjunctival haemorrhages, may imitate the symptoms of vascular stasis. In the presented case, oedema and haematoma in the ocular region observed in the patient directly after the accident were the result of injury, and the patient did not present the triad of symptoms characteristic of carotid cavernous fistula.

Exophthalmos of the right eyeball, which developed 3 weeks later, and the symptoms of palsy of the oculomotor nerve in the form of drooping of the eyelid, expanded pupil, ophthalmoplegia of the right eye, obliged the expanding of imaging diagnostics.

In the differential diagnostics of the causes for exophthalmos with accompanying palsy of mobility of the eyeball, the following should be considered: superior orbital fissure syndrome, cavernous sinus thrombosis, thyroid eye disease, and cranial nerves palsy due to other intracranial changes. Superior orbital fissure syndrome and cavernous sinus thrombosis, as well as carotid cavernous fistula, may be the result of trauma [1, 7]. Typical symptoms of these units are similar to those occurring in the case of carotid cavernous fistula. Exophthalmos, ophthalmoplegia related with the damage of the cranial nerves III, IV, VI, oedema occurring in stasis and extravasation of the conjunctiva may cause diagnostic difficulties. With respect to the differential diagnostics based on clinical symptoms, the type of exophthalmos is important, which, in the case of carotid cavernous fistula, is characterised by pulsation of the eyeball, as well as murmur in the head reported by the patient, typical exclusively of carotid cavernous fistula. Laboratory tests (concentration of D-dimers), which are performed in the case of suspicion of stasis of the sinus, do not allow the making of a diagnosis, and may only suggest the direction of diagnostic management. Neuro-imaging examinations performed with the vascular option are of primary importance in making the diagnosis [8].

Imaging examinations are also among the indispensable instruments in the differential diagnostics of other causes of exophthalmos, or palsy of cranial nerves. In the case described, angiomas and orbital varices, metastatic tumours, or tumours infiltrating the eye socket from the adjacent region, lymphomas, mucous cysts, orbital pseudotumour, inflammation of the soft tissues of the eye socket, and thyroid orbitopathy in the course of Graves Basedow disease, were excluded based on the results of imaging examinations, laboratory tests, clinical symptoms, and medical history taking.

Murmur in the region of the eye socket reported by the patient, as well as the occurrence of pulsation of the eyeball, which contributed to persistent exophthalmos and palsy of nerve III, suggested the suspicion of carotid cavernous fistula.

Ophthalmological examination revealed pulsating exophthalmos, which was characterised by palpable pulsation of the eyeball accompanied by a murmur synchronised with the heartbeat, heard by a patient. Pressure of the cranial artery on the side of the exophthalmos discontinued the pulsation and murmur heard by the patient.

Oedema of the conjunctiva diagnosed in the anterior segment of the eyeball, and expansion of the conjunctival and episcleral veins were the result of vascular stasis. Increased intraocular pressure noted in the patient might have been the effect of an increase in pressure in the episcleral veins. An impaired outflow of a watery fluid from the eye related with an elevated pressure in the episcleral vein leads, and consequently to the development of intraocular hypertension.

Vascular stasis its related haemodynamic disorders lead to ischaemic changes. In the presented case, no features of oxygen deficiency of the anterior segment were diagnosed, which may be manifested by the oedema of the corneal epithelium, tyndallisation of watery fluid, atrophy of the iris, or the development of cataract [1].

With respect to the posterior segment, in ophthalmological examination, only the dilation of veins was observed (Figure 3). Other symptoms, not diagnosed in the case described, evidencing the disorders in blood flow are intraretinal extravasations, stasis of the central retinal vein occlusion, oedema of the optic disc nerve II, and haemorrhage to the hyaline body. In the case of direct fistulas, there may develop non-rhegmatogenous retinal detachment, as well as detachment of the choroid [4]. Retinal ischaemia results in an increase in the production of angiogenic factors and, in consequence, leads to vascular endothelial growth factor with respect to the posterior and anterior segment, an increase in intraocular pressure, and the development of neovascular glaucoma [1].

Frequent clinical manifestations of CCF include symptoms of cranial nerve palsy (III, IV, V1, V2, VI) sometimes dominant in the clinical image [6, 9]. Lesions of cranial nerves develop as a consequence of ischaemic neuropathy in the course of hypoperfusion. They are manifested in the form of eyeball movement disorders, i.e. ptosis, diplopia in the case of palsy of the nerves III, IV, and VI, or face pain concerning nerves V1 and V2 [8]. Nerve VI is the intracranial nerve, which was damaged because it lies free floating in the cavernous sinus [1].

Kim *et al*. described the case of a 32-year-old woman with bilateral fractures of a blow out type, double vision, and impaired control of movement of the left eye, which occurred directly after injury. Exophthalmos, oedema of the conjunctiva, or drooping of the eyelid were not observed in the patient. After reconstructive surgery concerning the bones of the eye socket no regression of disorders in the mobility of the left eye was observed. Angiography of carotid arteries showed that the cause of the isolated palsy of nerve VI was post-traumatic carotid cavernous fistula. After embolisation of the fistula a total regression of the symptoms was obtained [9].

The course of the post-traumatic carotid cavernous fistula may vary from isolated palsy of the intracranial nerves to intracranial bleeding and death.

Argo *et al*. described the case of a patient who died due to uncontrolled intracranial bleeding which occurred 60 days after injury; the results of autopsy showed the presence of carotid cavernous fistula [10]. Another example supporting the risk of massive bleeding in the course of CCF is the description of the case of a patient who had undergone a blunt trauma of the head, with massive bleeding from the nose. The patient required intraoperative transfusion due to undiagnosed CCF [11]. Massive intracranial bleeding as a complication of CCF is also reported by Hayashi *et al*., who described the case of a 45-year-old woman in whom carotid cavernous fistula, complicated with intracranial bleeding, developed after a car accident [12].

In the case of direct carotid cavernous fistulas, the prognoses vary. The majority of fistulas are not life threatening, and the major complications concern the organ of vision. Ocular complications of untreated fistula include lesions of the cornea induced by lagophthalmos, secondary glaucoma, occlusion of the central retinal vein, ischaemic neuropathy of the optic nerve, rhegmatogenous retinal detachment, and choroidal detachment [1]. It is estimated that 80–90% of patients with untreated carotid cavernous fistula are afflicted by permanent loss of vision [6]. Rare complications include the above-mentioned ischaemic strokes or haemorrhagic changes, which may be lethal [10]. In the case of treated carotid cavernous fistulas, the prognosis is good [13]. The pulsation of the eyeball and the murmur heard by patients usually disappear directly after embolisation. Oedema of the nerve disc, symptoms of venous stasis, or palsy of the cranial nerves usually resolved within several weeks or months [3]. An elevated intraocular pressure may return to normal directly after embolisation, or within several months after the procedure. Exophthalmos and decreased acuity of vision may not fully resolve [4].

Considering the considerable threat to vision, as well as the risk of development of haemorrhagic complications, including death, imaging examinations are especially important in the diagnostics of CCF, and the results of these examinations may be decisive.

In diagnostic management CT angiography, MR angiography and Doppler ultrasound examinations are of primary importance. An examination that enables a precise imaging of carotid cavernous fistula is digital subtraction angiography (DSA). Angiography is both a diagnostic and therapeutic instrument, because it may constitute an introduction to the procedure of intravascular embolisation. In the case of craniocerebral traumas, the first imaging examination is computed tomography, where the position of venous sinuses or thickening of posterior ocular muscles may suggest the presence of carotid cavernous fistula and direct further management [13].

The treatment of carotid cavernous fistulas consists of intravascular embolisation or performance of a classic neurosurgical procedure [5, 13]. The use of the method of intravascular embolisation is associated with greater safety. The goal of the treatment is closure of the lumen of the fistula together with the expanded cavernous sinus while maintaining or restoring the normal arterial blood flow. The use of the techniques of balloon angioplasty and the implantation of stents and coils led to the closure of the fistula in 75–85% of cases. When these methods are ineffective, cyanoacrylate glue is used for embolisation [14].

In the case of the presented patient, embolisation was performed, obtaining a total closure of the fistula and resolution of the majority of the symptoms; 3 months after the procedure only the pupil remained slightly expanded.

In literature, case reports are available in which spontaneous resolution of the carotid cavernous fistula occurred. A case was described of spontaneous resolution of the carotid cavernous fistula 6 weeks after the performance of angiography [15]. During diagnostic angiography there may occur irritation of the vessels and clamping of the carotid artery, which results in the closure of the fistula [4].

Gapsis *et al*. presented the case of a woman with bilateral post-traumatic fissures, in whom the symptoms spontaneously resolved after 21 months, leaving no remaining deficits within the region of the organ of vision [16]. However, due to a high blood flow, the closure of a direct carotid cavernous fistula is very rare.

Conclusions

Post-traumatic fistulas should be qualified for treatment in an emergency mode due to the high risk of loss of vision and haemorrhagic complications. Any delay in the implementation of causative treatment is risky for the patient. Early diagnosis is indispensable in order to prevent the development of severe complications, such as loss of vision, massive blood loss, intracranial haemorrhages, cerebral ischaemia, and death. It should be remembered that even full-blown symptoms of post-traumatic fistulas may develop within

the period of several weeks after trauma. Knowledge of the symptomatology, correct differential diagnostics, and adequate causative treatment allow the protection of the patient against severe complications of this disorder, and when undertaken early enough, may lead to the total resolution of symptoms.

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