



## CAROTID CAVERNOUS FISTULAS IN POSTMENOPAUSAL WOMEN

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**SUMMARY** – During the last five-year period, eight patients were hospitalized at University Department of Ophthalmology, Sestre milosrdnice University Hospital, for elaboration and therapy of spontaneous indirect carotid cavernous fistulas. The aim of this report is to present our experiences in the diagnosis, treatment and follow up of patients with carotid cavernous fistulas. All patients were postmenopausal women, mean age  $69.75 \pm 7.3$  years, without any trauma in their history. The most common symptoms were proptosis, low vision and chemosis. Ultrasound, computerized tomography, magnetic resonance imaging and angiography were used as diagnostic methods. Endovascular balloon occlusion was the treatment of choice in all patients except for one with spontaneous closure of the fistula, whereas another one refused this mode of treatment. The first follow up examination at one month after therapy showed improvement of ocular status. There was no pain, bruit, chemosis, pulsations of the globe, headache or double vision, while periorbital edema, proptosis, dilatation of retinal veins and intraocular pressure were reduced. During the six-month follow up period, no recurrence was recorded. Spontaneous fistulas described in this study tend to be dural shunts. These shunts have a predilection to develop in postmenopausal women, possibly due to atherosclerosis, hypertension and alterations in blood coagulation associated with hormonal changes.

**Key words:** *Carotid cavernous fistula; Dural shunt; Postmenopausal women*

### Introduction

Carotid cavernous fistula (CCF) results from an abnormal communication between the carotid artery system and the cavernous sinus. Carotid cavernous fistulas can be spontaneous or traumatic and are classified as direct or indirect. Direct fistulas occur in 70%-90% of all cases and are caused by tears in the intracavernous portion of the internal carotid artery arising from trauma or from rupture of a preexisting aneurysm<sup>1,2</sup>. They are high-flow fistulas and the onset can be dramatic with severe pulsatile proptosis, bruit, and poor vision due to ocular

hypoxia. Indirect fistulas (dural shunts) occur when thin walled meningeal branches of the internal or external carotid arteries rupture spontaneously within the cavernous sinus. Since they are low-flow, the onset may be subtle with mild proptosis, redness and ocular hypertension.

Although the etiology of CCF does not belong to the field of ophthalmologists, clinical presentation and complications do. It is in fact a rare condition, especially dural spontaneous fistulas, and its clinical presentation can be easily overlooked. Many patients with low-flow CCF are misdiagnosed and treated erroneously for infections and inflammation, often for months to years, as it was the case with few of our patients.

Dural shunts are frequent in hypertensive patients, after minor trauma or on straining. Postmenopausal hy-

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pertensive women are at a particular risk because the carotid artery intima-media thickness was shown to strongly and positively correlate with age in women aged 55 years and older but not before<sup>1,2</sup>.

Estrogens modulate arteriolar stiffness, baroreceptor sensitivity, and blood pressures, which all are key factors that play an etiologic role in the development of a major vascular risk factor of hypertension<sup>3</sup>. Diagnosis is accomplished through ultrasonography, neuroimaging and arteriography, which is the gold standard in identifying CCF with vessel involvement. As this is a rare condition and 20%-50% of indirect carotid cavernous fistulas will close spontaneously, it is often a dilemma whether we should treat the patient with indirect fistula and how long we should wait before treatment.

There is a common attitude that lesions that present with worsening proptosis, pial venous drainage, deteriorating vision, epistaxis, increased intracranial pressure, glaucoma and ophthalmoplegia require urgent or semi-urgent treatment. Transvenous endovascular therapy (balloon occlusion or coil embolization) is indicated for high- and low-flow dural CCFs that require repair.

## Patients and Methods

This was a retrospective study that included all CCF patients hospitalized at University Department of Ophthalmology, Sestre milosrdnice University Hospital, during the period between January 2003 and January 2008. All patients were postmenopausal women aged 51-80 (mean age  $69.75 \pm 7.3$ ) years. All patients had a spontaneous onset of CCF. There was no trauma in their history.

Ophthalmologic examination included visual acuity testing and refraction, slit lamp examination, fundus examination and applanation tonometry using Goldmann's applanation tonometer, Goldmann visual field, Hertel exophthalmometry, extracranial doppler of carotid arteries and power doppler.

The diagnosis of CCF included clinical examination, radiological evaluation using ultrasonography, contrast-enhanced computerized tomography (CT), magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA). CT scanning and MRI are preferred radiological modalities. CT findings included enlargement of cavernous sinus, enlargement and tortuosity of the superior ophthalmic vein (greater than 4 mm in diameter), enlargement of extraocular muscles and proptotic eye. MRI findings included findings similar to those on CT, i.e. abnormal flow voids in the affected cavernous sinus, decreased MRI signal and involved cav-

ernous sinus, dilated intercavernous sinuses and intercavernous vessels, lateral wall convexity of the cavernous sinus and dilated superior ophthalmic vein.

Orbital sonogram findings demonstrate signs similar to those on CT scans and MRI. We found dilated tortuous veins which were prominent on B-scan echograms. With the A-scan method dilated ophthalmic vein was evident. MRA was very useful in identifying fistulas as well as particular vessel involvement, which was of crucial importance for the treatment.

All patients were referred for endovascular balloon occlusion treatment using detectable silicon balloon delivered by a flow-control guide catheter. After endovascular balloon had been placed in the desired location, an angiogram was performed to confirm occlusion of the fistula before deployment. Treatment was done in six patients. In one patient, the fistula closed spontaneously while waiting for the treatment, and another one refused this mode of treatment.

## Results

Eight patients with spontaneous carotid cavernous fistula were included in the study. Hypertension was found in six women. Fistulas were mostly right sided (six cases). Orbital signs found on examination are shown in Table 1.

Conjunctival injection with tortuous perilimbal episcleral vessels is one of the most prominent external signs. It was found in all our patients, in association with chemosis of the conjunctiva. Proptosis of the involved eye was present in five cases. In one case, the protrusion was bilateral from the start, almost equally on both sides, suggesting large intracavernous venous communications. In one case the protrusion was bilateral, but much more severe on the side of the fistula. In addition, protrusion manifested later on the contralateral eye.

Table 1. Orbital signs found on examination

| Symptoms  | Eyes |
|---|------|
| Conjunctival injection, tortuous episcleral veins | 8    |
| Chemosis  | 6    |
| Proptosis   | 5    |
| Objective visual loss                             | 5    |
| Periorbital flush, edema, venous dilatation       | 5    |
| Pulsations of globe                               | 1    |
| Audible bruit                                     | 2    |
| Dilated retinal veins                             | 4    |

Table 2. Symptoms reported by patients at onset

| Symptoms                            | Eyes |
|-------------------------------------|------|
| Visual impairment or blurred vision | 6    |
| Diplopia                            | 2    |
| Headache                            | 2    |
| Ocular or retro-ocular pain         | 3    |
| Swishing noise in head              | 3    |

Minimal and maximal protrusion as assessed by exophthalmometer was 2 mm and 11 mm, respectively. The duration of the disease seems to have little impact on the size of proptosis. Retinal hemorrhages and papilledema were not recorded.

Pulsation of the globe, 'tonometer sign', was observed in two patients. It is caused by pulsations of the globe which then hits the applanation probe while measuring intraocular pressure with Goldmann tonometer. In four patients there was evident periorbital flush and edema of the lids. Visual loss was present in six patients, and we recorded no late visual loss. There was no case of blindness or severe visual loss. After treatment, visual acuity improved. The symptoms reported by patients at onset are listed in Table 2.

Vision impairment or blurred vision was the most common complaint, always on the side of the fistula. Headache, usually on the side of the fistula, was reported by two patients. Swishing noise in the head was present in three patients, and one patient felt swishing noise in her chest. Only in two patients we were able to hear an audible bruit with a stethoscope. Diplopia was present in two patients. Involvement of the following cranial nerves was recorded: abducens, trigeminal and facial nerve in one case each.

Damage to the trigeminal and facial nerves as evidenced by sensation or by lagophthalmos presents a constant threat to the globe where proptosis is already a serious problem. The development of secondary high intraocular pressure was observed in 5 cases.

## Discussion and Conclusion

Many excellent summaries about carotid cavernous fistulas have already appeared, however, we hope that a review of the present series may accentuate the problem itself. Orbital venous pathologies encompass a broad range of entities including tumors, shunts, congenital anomalies, aneurysms and obstructive lesions. Patients may present with a variety of clinical findings which may include a combination of tumefaction, vascular engorgement, orbital pulsation and exophthalmos, depending on the relationship between the lesion and the vascular system. An extensive clinical and radiological evaluation in such patients is necessary.

Suspicion of CCF is crucial for diagnosis. All our patients with low-flow CCF were misdiagnosed and treated for infections and inflammation for months to a year. Although all accumulated figures from the literature show that approximately 25% of carotid cavernous fistulas are spontaneous and 75% are traumatic, in our patients the etiology was 100% spontaneous. CCFs heal spontaneously in some 5% to 10% of patients. In our study, spontaneous closure of the fistula occurred in one (12.5%) patient. Another 10% perhaps are aided by cervical compression, probably leading to vascular thrombosis with relief of symptoms. The remaining 80% to 85% find very little or no relief without treatment. Only about 3% die from intracranial hemorrhage. We could

Table 3. First clinical examination before treatment

| Patient No. | Visual acuity | Intraocular pressure (mm Hg) | Hertel exophthalmometry | Fundus – dilated and tortuous veins |
|-------------|---------------|------------------------------|-------------------------|-------------------------------------|
| 1           | 0.2           | 21/18                        | 19/17                   | +                                   |
| 2           | 0.4           | 24/17                        | 22/16                   | +                                   |
| 3           | 0.7           | 20/16                        | 17/14                   | +                                   |
| 4           | 0.075         | 26/16                        | 25/18                   | +                                   |
| 5           | 0.05          | 25/17                        | 17/15                   | +                                   |
| 6           | 0.8           | 20/15                        | 14/15                   | –                                   |
| 7           | 1.0           | 19/17                        | 15/15                   | –                                   |
| 8           | 0.9           | 20/14                        | 14/14                   | –                                   |

Table 4. Clinical status one month after treatment

| Patient No. | Visual acuity | Intraocular pressure (mm Hg) | Hertel exophthalmometry | Fundus – dilated and tortuous veins |
|-------------|---------------|------------------------------|-------------------------|-------------------------------------|
| 1           | 0.6           | 19/17                        | 19/17                   | +                                   |
| 2           | 0.6           | 19/17                        | 20/16                   | +                                   |
| 3           | 1.0?          | 16/16                        | 16/14                   | +                                   |
| 4           | 0.3           | 17/17                        | 21/18                   | +                                   |
| 5           | 0.1           | 16/17                        | 15/15                   | +                                   |
| 6           | 1.0           | 16/16                        | 14/15                   | –                                   |
| 7           | 1.0           | 18/18                        | 15/15                   | –                                   |
| 8           | 1.0           | 18/16                        | 14/14                   | –                                   |

not confirm any symptom relief in our six patients before treatment. The longer the CCF remains patent, the more opportunity it has to increase in size, the more the injected vessels thicken, and the more fibrous changes occur in the abnormally distended orbital and periorbital tissues. The best results were therefore obtained in patients with early treatment<sup>2</sup>.

Our patients treated with transarterial embolization showed immediate and permanent occlusion of the fistula and improved visual acuity at six-month follow up. They all were very satisfied after the treatment, having no or minimal symptoms and improvement of ocular status (see Tables 3 and 4).

We recorded no potential complications of endovascular treatment like thromboembolic and ischemic events related to the balloon and catheter manipulation; formation of pseudoaneurysm secondary to balloon injury; and changes in arterial hemodynamics that can cause hemorrhage, edema and progression of ocular damage.

If a low-flow CCF is present with orbital symptoms that are worsening in time, it is best not to wait for spon-

taneous fistula closure but to treat the patient with endovascular balloon occlusion, which seems to be safe and effective treatment for CCF.

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## Sažetak

## KAROTIDNE KAVERNOZNE FISTULE KOD ŽENA U POSTMENOPAUI

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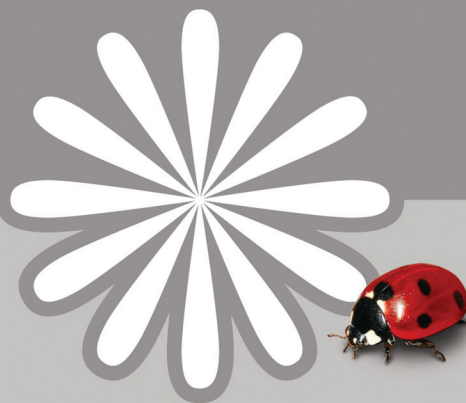
U razdoblju od posljednjih pet godina na Klinici za očne bolesti Kliničke bolnice "Sestre milosrdnice" osam bolesnica je hospitalizirano radi obrade i terapije kavernoze karotidne fistule. Cilj ove studije bio je prikazati naša iskustva u dijagnostici, terapiji i praćenju bolesnika s kavernoznim karotidnim fistulama. Svi pacijenti bile su žene u menopauzi srednje životne dobi  $69,75 \pm 7,3$  godina, bez ikakve traume u anamnezi. Najčešći simptomi bili su proptoza, pad vida i kemoza. Od dijagnostičkih metoda rabili smo ultrazvuk, magnetsku rezonanciju (NMR) i angiografiju. Endovaskularna balonska okluzija bila je terapija izbora kod svih bolesnica, ali je kod jedne došlo do spontanog zatvaranja fistule, dok je jedna odbila takvu vrstu liječenja. Na prvom pregledu, mjesec dana nakon terapije, došlo je do poboljšanja očnog statusa. Simptomi kao bol, šum, kemoza, pulsacije bulbusa, glavobolje i dvoslike nisu bili prisutni. Zamijećen je mnogo manji periorbitalni edem, proptoza i dilatacija retinalnih vena, te niže vrijednosti intraokularnog tlaka. Razdoblje praćenja bilo je šest mjeseci i nije zamijećena ponovna pojava fistule. Takve spontane fistule opisane u ovoj studiji su duralni šantovi s predilekcijom u postmenopauzalnih žena, najvjerojatnije zbog ateroskleroze, hipertenzije i promjena u zgrušavanju krvi povezanih s hormonskim statusom.

*Ključne riječi: Karotidno kavernoza fistula; Duralni šant; Postmenopauzalne žene*





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