Two Cases of Orbital Infarction Syndrome

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Orbital infarction syndrome is defined as ischemia of all intraorbital and intraocular structures. It is a rare disease caused by rich anastomotic vascularization of the orbit. It can occur secondary to different conditions, such as, acute perfusion failure, systemic vasculitis, orbital cellulitis and vasculitis. It results in orbital and ocular pain, total ophthalmoplegia, anterior and posterior segment ischemia, and acute blindness. We report here upon two cases of orbital infarction with similar presentations but with different causes, namely, mucormycosis and as a postoperative complication of intracranial aneurysm, discuss the possible mechanisms of orbital infarction, and present a review of the literature on the topic. The prompt recognition of clinical pictures and rapid diagnosis is essential for the early treatment of orbital infarction, since its progression is very rapid and it can be even fatal.

Key words: orbital infarction syndrome, mucormycosis, intracranial aneurysm

INTRODUCTION

Global orbital infarction is a rare disorder resulting from ischemia of the intraocular and intraorbital structures due to hypoperfusion of the ophthalmic artery and its branches. This syndrome can occur after common carotid occlusion, caused by, giant cell arteritis, myelofibrosis, orbital mucormycosis, and surgical complications.

In order to assist in the diagnosis of orbital infarction, we report upon two cases of orbital infarction syndrome with different causative mechanisms.

CASE REPORTS

Case 1

A 61-year-old woman experienced a sudden ocular pain, her upper eye-lid swelling had been worsening for a period of 7 days. The patient was diagnosed as having diabetes 3 years previously, but did not receive any treatment. Her vital signs were normal and mental status alert. Visual acuity was 8/20 (OD) and 10/20 (OS), which could not be corrected. The lids showed mild swelling, but pain and ecchymosis were absent. The pupils were mildly dilated, and IOPs were 22 mmHg (OD) and 21 mmHg (OS).

The patient visited the outpatient department on the following day, visual acuity had deteriorated to negative light perception in her right eye, the eyelid swelling had worsened, and ptosis was also present. Extraocular motility was limited in all directions. Conjunctival injection, swelling and proptosis had worsened. The pupils were mildly dilated, and a cherry-red spot was detected on fundus examination. Intraocular pressure showed no change. Orbital MRI revealed hypertensive linear lesions along the right optic nerve in the orbital fat and the optic canal. Findings typical of preseptal cellulitis were present. The cavernous sinus was normal, but severe

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Fig. 1. Initial finding of MRI (Case 1): post gadolinium DTPA enhanced T1-weighted axial image with fat suppression reveals streaky enhancing lesions along the right optic nerve (arrows). Note right preseptal periorbital soft tissue swelling and proptosis. Cavernous sinuses are symmetric in both sides. Enhanced thickened mucosal linings were seen in both the anterior ethmoid sinuses and the right maxillary sinus (not shown), suggesting sinusitis.

Paranasal sinusitis was present (Fig. 1). Fluorescein angiography showed delayed choroidal phase. The patient was diagnosed as having paranasal sinusitis, right orbital cellulitis and right orbital apex syndrome, and admitted for antibiotics therapy. On the 2nd day, paranasal sinus drainage was performed via fronto-ethmoidectomy and middle meatal antrostomy. On the 3rd day, the orbital swelling had worsened, and extended to the forehead and the lower cheek. Petechia was also observed. Ptosis had also worsened and the extraocular muscles became fixed, while the pupils were fully dilated and fixed, thereby giving an appearance of “cadaveric eye” (Fig. 2). Electrolyte examination showed decreased sodium/potassium levels (129/3.4) and the body temperature was 38.3°C. Mucormycosis in the paranasal sinus was diagnosed, and the patient was transferred to the internal medicine department for systemic amphotericin B therapy. The following day, the patient showed a drowsy mentality, while swelling in the left lid was also observed. The skin in the swollen area was slightly violet in color. Orbital MRI examination was requested. Ischemic findings without enhanced contrast were observed in the right orbit, ethmoid sinus, nasal septum and cavernous sinus, which were in contrast with that of the left side. Also, hyperintensive linear lesions that were absent in the initial study were observed in the left orbital fat and soft tissue (Fig. 3). The patient showed progressive electrolyte imbalance and fever, and was transferred to the intensive care unit. Facial color changes suspicious of right facial necrosis were observed, and the patient deteriorated into a
comatose state (Fig. 4). Culture of the paranasal sinus drainage fluid confirmed mucormycosis. Seven days after admission, the patient was discharged due to a request from her relatives.

**Case 2**

A 32-year-old man experienced a sudden headache with vomiting. Computed tomography (CT) showed subarachnoid blood at the basal cistern; cerebral arteriography demonstrated a ruptured right anterior choroidal artery aneurysm. The size of intracranial aneurysm was $3 \times 4$ mm and directed posteriorly. The aneurysm was clipped via a right fronto-temporal craniotomy. Two hours after surgery, the right eye developed proptosis and chemosis. The patient became alert 24 hours after surgery. Due to the progression of proptosis, on the first post-operative day the patient was referred for consultation to the department of ophthalmology.

On ophthalmic examination, the right pupil was 5 mm in diameter and nonreactive to direct or consensual light reflex with afferent pupillary defect. He had severe proptosis (right: 24 mm, left: 15 mm, base: 115 mm), chemosis and complete ophthalmoplegia in the right eye. Intraocular pressures (IOPs) seemed to be high by digital palpation. The patient’s visual acuity was no light perception in the right. The right fundus showed retinal edema throughout the posterior pole and mid-periphery, with cherry-red spots (Fig. 5). Immediate lateral canthotomy and 250 ml of intravenous mannitolization were under-

taken. Computed tomography showed retro-orbital soft tissue swelling (-100 HounsfieId unit) of a fat-like density, causing right proptosis, but no intraorbital hemorrhage or cavernous sinus abnormalities. The right communicating intraorbital-frontal lobe hole was suspected continuous to the right superior orbital fissure (Fig. 6). Retro-orbital soft tissue swelling ruled out emphysema or hemorrhage through the bone setting. Visual evoked potential and the electroretinogram showed no waveform. The exact cause of the soft tissue swelling was not known. Secondary intraocular pressure elevation developed due to soft tissue swelling within the confined space of the orbit. We diagnosed optic neuropathy, 3rd nerve palsy and ophthalmic artery occlusion of the right eye after the aneurysmal operation. The patient was treated with 250 ml of 25% mannitol twice a day and 500 mg of soulmelodol, starting on systemic acetazolamide. Twelve weeks after surgery, the right fundus showed advanced optic atrophy and pigmentary retinopathy, 30 prism exotropia in right eye. Fluorescein angiography confirmed global retinal and posterior ciliary nonperfusion.

**DISCUSSION**

The ophthalmic artery originates from the internal carotid artery and has numerous ocular and orbital branches, which include the central retinal and ciliary arteries.² Indeed, the ophthalmic artery system
Fig. 6. Proptosis of the right eye, -100 hounsfield units fat density and a right communicating intraorbital-frontal lobe hole were suspected continuous to the right superior oblique fissure after aneurysmal operation (case 2).

is not a terminal system. There are several anastomoses and a rich collateral circulation between the branches of the external carotid and the ophthalmic artery. As we have described above, global orbital infarction results from ischemia of all the intraocular and intraorbital structures due to hypoperfusion of the ophthalmic artery and its branches. Therefore, isolated ophthalmic artery occlusion alone does not cause this syndrome and clinical presentation can vary greatly. However, in addition to ophthalmic artery occlusion, the collateral blood flow may be compromised by other factors, which lead to the development of orbital infarction. Visual loss results from retinal infarction (due to hypoperfusion of the central retinal artery), anterior ischemic optic neuropathy and choroidal ischemia (both due to compromised posterior ciliary arteries). Ophthalmoplegia results from impaired blood supply to the extraocular muscles or to the ocular motor nerves, and anterior segment ischemia due to hypoperfusion of the long ciliary arteries.

Orbital infarction can occur secondary to various pathologic causes such as common carotid artery occlusion, giant cell arteritis, myelofibrosis, mucormycosis, trauma, and intracranial surgical complications. As seen in our first patient, it is not surprising for orbital infarction to occur in mucormycosis, because the hyphae have a marked tendency to invade and thrombose arteries. The first patient showed the characteristics of orbital infarction, but this was also associated with marked proptosis, orbital cellulitis, and the black eschar characteristic of mucormycosis. Although mucormycosis occurs in patients with DM or in those whose immunity is deficient, it can also occur in people with normal immunity. Clinically, mucormycosis in its rhino-orbito-cerebral form is the most prevalent form, but it is an opportunistic infection in the paranasal sinus of patients with DM or in those with an immunity deficiency, spreading to the orbit and brain. The organism produces obliterative arteritis and tissue necrosis. Although we had suspected mucormycosis in our first case, as she showed uncontrolled DM history and sinusitis on CT findings, and the clinical findings of orbital apex syndrome. This syndrome was confirmed eventually by nasal biopsy and the black eschar. Eventually, the patient lost her vision, her life was threatened, and even aggressive treatment with amphotericine B produced no improvement. Thus, although it has been reported that only 25% of mucormycosis progresses to blindness, rhinocerebral mucormycosis can cause not only orbital infarction but can also progress into a life-threatening condition.

The second case developed orbital infarction by a different mechanism. Zimmerman et al. reported that orbital infarction syndrome after intracranial aneurysm surgery has multiple factors, and patients with subarachnoid hemorrhage, increased intracranial pressure, anomalous arterial or venous circulation, or impaired orbital venous outflow seemed particularly vulnerable. However, in this case, there was no rise in intracranial pressure or any abnormality in the arteries or veins at the time of the aneurysmal operation. Frontotemporal or “pterional” craniotomy exposure requires a myocutaneous flap to be retracted anteriorly and inferiorly near the orbit and in some circumstances, this bulky flap could indirectly exert pressure on the globe, especially in individuals with “shallow” orbits or significant orbital congestion. Our patient was in the supine position during the operation, and therefore, the venous outflow obstruction that can occur in the dependent position could be excluded. Bone drilling is frequently performed for aneurysmal exposure or to secure a view of the pterional bone at the time of intracranial aneurysm operation. In our case, it may be that a hole adjoining the right superior orbital fis-
sure might have been made inappropriately during the bone drilling procedure. From the CT finding, the space between the right superior orbital fissure became wider compared to the left eye (Fig. 6). It was reported that the development of orbital emphysema, after an orbital fracture, is thought to involve a three-step process with a sino-orbital communication. A forceful expiratory effort creates a pressure gradient, which may force air into the orbit and orbital tissue, material such as fat may obstruct this communication, creating a one-way valve.\textsuperscript{12} Acute compartment syndrome with vascular compromise can then be induced due to the trapped intraorbital air mass.\textsuperscript{13} In case 2, the cause of the compression that formed in the retrobulbar space was not the air, but the soft tissue, similar to the finding of fat and that caused a swelling and pressure effect within the orbital cavity. During the intracranial aneurysm operation, saline is used continuously to irrigate the surgical site in order to reduce temperature during the drilling procedure and when the operation is almost complete, the surgical site is irrigated with saline solution. We suspect that saline entered the intraorbital space through the fronto-orbital communicating hole and caused edema of the soft tissue, but we don’t know the precise cause of the retrobulbar fat and soft tissue swelling. The swollen fat and soft tissue cause acute compartment syndrome, that in turn cause proptosis and the elevation of intraocular pressure. We were able to determine ophthalmic artery obstruction rather than central retinal artery obstruction since the overall swelling of the retina, pigmentary retinopathy and optic nerve atrophy were evidenced and light perception was lost. As the collateral arterial network evolves, the clinical picture may improve. However, when diagnosis is delayed, ophthalmologic problems including visual loss may occur, and even lead to a life-threatening level.

In conclusion, a diagnosis of global orbital infarction should direct investigations towards not only thromboembolic, inflammatory, and infectious arterial disease but also towards compressive arterial lesion. Prompt recognition of the disease progression and treatment are essential to allow visual recovery, and avoid life-threatening conditions.

REFERENCES