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CASE REPORT

POSTERIOR ISCHEMIC OPTIC NEUROPATHY SECONDARY ON DIRECT CAROTID CAVERNOUS FISTULA: A RARE CASE REPORT

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Abstract

Background: Direct carotid-cavernous fistula (CCF) is a pathological arteriovenous connection between the internal carotid artery and the cavernous sinus, commonly resulting from craniofacial trauma. Posterior ischemic optic neuropathy (PION) is an uncommon but vision-threatening complication of traumatic CCF.

Case Report: A 45-year-old woman presented with a one-week history of left eye swelling, double vision, ocular pain, redness, tearing, and progressive visual loss following a traffic accident. Left eye examination showed 2 meters counting finger visual acuity, 25 mmHg intraocular pressure, ocular movement limitation, superior visual defect, decreased colour vision and contrast sensitivity. Anterior segment findings included proptosis, eyelid swelling, 180-degree inferior chemosis, conjunctival injection, dilated corkscrew episcleral vessel, positive rapid anterior pupillary defect. Significant improvement followed after endovascular treatment.

Discussion: The coexistence of direct CCF and PION is exceptionally rare. This case showed atypical visual loss without disc edema. The likely mechanism involves arterial steal and venous congestion. Early intervention led to partial visual recovery. Prompt recognition and ophthalmologic monitoring are essential to improve visual outcomes in CCF-associated PION.

Conclusion: Early collaborative management is vital in cases of traumatic CCF with PION to prevent disease progression, preserve visual function, and achieve optimal outcomes**.**

Keywords: Carotid Cavernous Fistula, Posterior Ischemic Optic Neuropathy, Visual Field Defect

INTRODUCTION

Arteriovenous fistulas are acquired vascular anomalies characterized by an abnormal direct connection between an artery and a vein, bypassing the capillary network. These lesions may arise due to trauma or degenerative processes and are broadly classified into two types: direct and indirect (dural) carotid-cavernous fistulas (CCFs).[1]

Both direct and indirect CCFs commonly present with ocular symptoms such as proptosis and elevated intraocular pressure. Additional manifestations may include diplopia, that resulting from cranial nerve palsy[1] or orbital venous congestion, retinal arterial or venous compromise, ischemic optic neuropathy | (ION), choroidal effusions, ocular pain (often due to exposure keratopathy from proptosis), ipsilateral headache, and in rare cases, cerebral venous infarction secondary to venous hypertension.[2]

We report a rare case of traumatic direct CCF complicated by PION, highlighting its atypical clinical presentation, diagnostic considerations, and management outcomes. This case highlights the critical need for early identification and coordinated multidisciplinary management to prevent permanent visual loss and other serious sequelae. Close ophthalmologic follow-up is imperative for ongoing evaluation of visual function, inform prognosis, and detect progressive changes, including optic nerve atrophy.

CASE ILLUSTRATION

A 45-year-old female was referred from the Neurosurgery Department to the Neuro-ophthalmology Outpatient Clinic at Dr. Soetomo General Academic Hospital with history of progressive swelling and proptosis of the left eye. The symptoms began approximately one week following a motor vehicle accident. The ocular complaints were accompanied by pain, redness, excessive tearing, restricted ocular motility, horizontal diplopia, and blurred vision in the left eye. The patient reported a decrease in visual clarity along with superior visual field loss in the affected eye. She noted that the diplopia improved when the fellow eye was closed. Additionally, she described a pulsatile "whooshing" sound in her left ear. The patient had no history of hypertension, diabetes mellitus, or other systemic diseases. She had history of head trauma sustained in a traffic accident approximately two weeks prior to presentation.

On physical examination, the patient was alert and fully oriented (compos mentis) with other vital signs were within normal limits. Visual acuity in the right eye was 4 meters counting fingers, which improved to 5/5 with best corrected visual acuity (BCVA) using a −2.50 dioptre spherical lens. In the left eye, visual acuity was 2 meters counting fingers and did not improve with pinhole. Intraocular pressure measured by non-contact tonometry was 18 mmHg in the right eye and 25 mmHg in the left eye. Ocular motility was normal in the right eye. In the left eye, significant restrictions were noted: approximately -3 limitation in nasal and temporal gaze, and -4 limitation in inferonasal, superonasal, superotemporal, and inferotemporal directions. Color vision testing using the Ishihara chart showed 38/38 correct plates in the right eye and 24/38 in the left eye. Confrontation visual field testing was normal in the right eye, while the left eye exhibited superior field loss. Hertel exophthalmometry revealed 16 mm for the right eye, 20 mm for the left eye, with a base of 110 mm.

Anterior segment examination of the right eye was unremarkable. In the left eye, ocular bruit was present. Clinical signs included proptosis, upper eyelid edema, 180-degree inferior chemosis, conjunctival injection, episcleral vessel dilation, and the presence of corkscrew vessels **(Figure 1)**. A relative afferent pupillary defect (RAPD) was noted in the left eye.

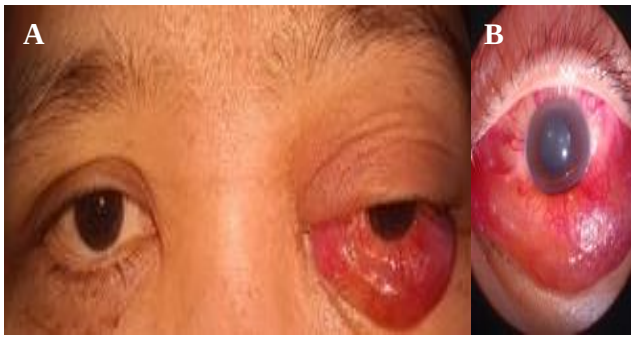

Figure 1. (A) Frontal view of left eye showing eyelid edema and chemosis. (B) Slit-lamp photo showing 180° inferior chemosis with corkscrew appearance

(Picture was taken with the patient's consent.
Courtesy: Ophthalmology Oupatient Clinic Dr Soetomo)

Posterior segment evaluation was normal in the right eye, whereas the left eye demonstrated vascular tortuosity. Humphrey visual field testing revealed a peripheral field defect in the right eye and an altitudinal field defect in the left eye. Spectral-domain optical coherence tomography (OCT) of the left eye showed normal retinal nerve fiber layer (RNFL) thickness and normal inner limiting membrane to retinal pigment epithelium (ILM–RPE) complex. However, there was focal thinning of the ganglion cell–inner plexiform layer (GCL–IPL), particularly in the nasal and superior quadrants **(Figure 2)**.

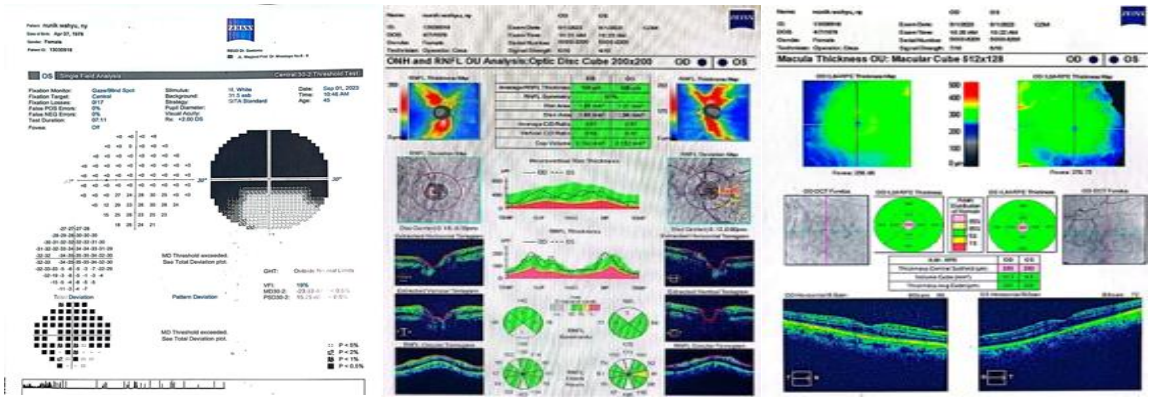

Figure 2. Humphrey Perimetry showed altitudinal visual field defect and Optical Coherence Tomography (OCT) showing normal RNFL and ILM-RPE thickness on the left eye.x

(Courtesy: Ophthalmology Oupatient Clinic Dr Soetomo)

Based on the clinical presentation and examination findings, the patient was diagnosed with a suspected traumatic carotid-cavernous fistula (CCF) in the left eye, accompanied by posterior ischemic optic neuropathy and complete ophthalmoplegia. To confirm the diagnosis and further assess the extent of vascular involvement, neuroimaging with magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) was arranged.

Initial medical therapy was initiated by the Neuro-ophthalmology Division with the aim of controlling intraocular pressure, reducing inflammation, and providing neuroprotection. The patient was prescribed timolol maleate eye drops twice daily in the left eye, along with electrolyte-based artificial tears every four hours and chloramphenicol eye ointment three times daily. Systemic therapy included methylprednisolone 32 mg taken twice daily for seven days, followed by a once-daily dose for another seven days. To mitigate gastrointestinal side effects, omeprazole 20 mg was administered twice daily. Additionally, citicoline 500 mg twice daily was prescribed for neuroprotection, and mefenamic acid 500 mg every eight hours was given for analgesia.

In coordination with the Neurosurgery Department, the patient was scheduled for transfemoral cerebral angiography (TFCA) as a definitive diagnostic and therapeutic step. The patient underwent balloon embolization performed by the Neurosurgery Department. The procedure was conducted via a jailed microcatheter approach, and post-embolization angiography demonstrated complete occlusion of the carotid-cavernous fistula **(Figure 3)**.

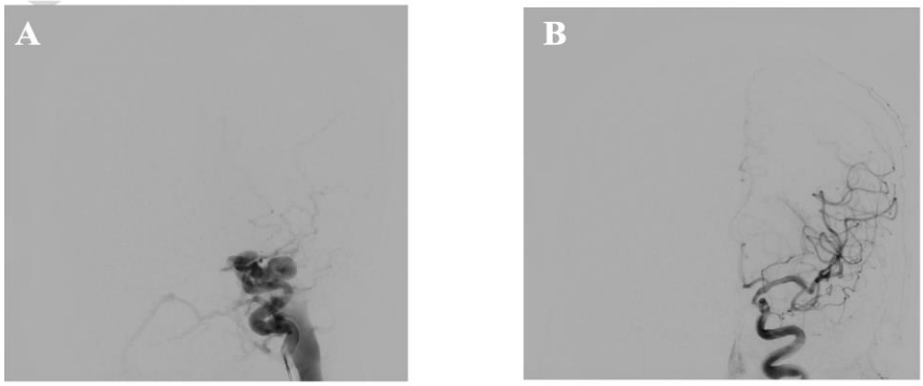

Figure 3. (A) Pre-balloon embolization of left ICA angiography (B) Post-balloon embolization revealed complete fistula occlusion. (Courtesy: Neurosurgery Department Dr Soetomo)

At the one-week follow-up in the outpatient clinic, the patient reported notable improvement in visual outcomes. She described improved vision, a reduction in diplopia, and resolution of the pulsatile tinnitus (previously perceived as a whooshing sound). On examination, visual acuity was 4 meters counting fingers, improving to 5/5 with -2.50 spherical correction in the right eye, and 5/20 pinhole 5/16 in the left eye. Intraocular pressure (IOP) had normalized, measuring 15 mmHg in the right eye and 10 mmHg in the left eye.

Ocular motility in the right eye remained full, while the left eye showed improved movement with a residual -3 limitation temporally and -1 in other directions. Color vision, assessed with Ishihara plates, was full (38/38) in both eyes. Confrontation visual field testing was normal in the right eye and showed improvement in the left eye, with remaining superonasal field loss. Exophthalmometry measurements were 16 mm – 110 mm – 16 mm, indicating resolution of the previously noted proptosis.

On subsequent anterior segment examination, the right eye remained within normal limits, while the left eye showed resolution of previous findings including absence of ocular bruit, proptosis, eyelid swelling, and corkscrew episcleral vessels. Only minimal chemosis and mild conjunctival injection were noted **(Figure 4)**. Relative afferent pupillary defect (RAPD) persisted in the left eye.

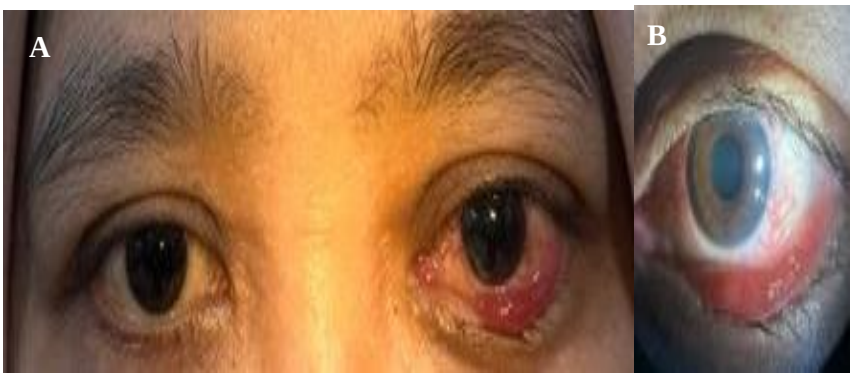

Figure 4. (A) Frontal view showing minimal inferior chemosis in the left eye. (B) Slit lamp showing 60° chemosis and conjunctival hyperemia(Picture was taken with the patient's consent. Courtesy: Ophthalmology Oupatient Clinic Dr Soetomo)

Posterior segment examination revealed a normal fundus in the right eye, while the left optic disc appeared slightly pale, suggestive of evolving optic neuropathy, with no evidence of vascular tortuosity.

Given the clinical improvement, treatment was adjusted accordingly. The patient was prescribed electrolyte-based artificial tears every 4 hours and chloramphenicol eye ointment every 8 hours in the left eye. Systemic therapy included methylprednisolone 16 mg once daily for 7 days, omeprazole 20 mg twice daily, citicoline 500 mg twice daily, and vitamin B complex once every 12 hours. Topical timolol maleate was discontinued due to normalized intraocular pressure.

DISCUSSION

Posterior ischemic optic neuropathy (PION) is a rare but important complication in patients with carotid-cavernous fistula (CCF).[3–6] Most cases of vision loss in direct CCF are attributed to damage or compression of the optic nerve resulting from elevated cavernous sinus pressure. This condition often presents with dilated retinal veins, and in more advanced stages, optic disc edema and retinal hemorrhages.[7]

However, our patient demonstrated a distinct clinical pattern. In this case, the patient experienced acute visual decline,[8,9] reduced color and contrast sensitivity, and a relative afferent pupillary defect (RAPD), accompanied by an altitudinal visual field defect on Humphrey Visual Field (HVF) testing, yet the initial fundus examination revealed no optic disc edema. This finding suggests a lesion located posterior to the optic nerve head, which is characteristic of PION.[9,10]

The diagnosis was further supported by the absence of papilledema despite signs of venous congestion, such as vascular tortuosity. A possible underlying mechanism is an arterial steal phenomenon due to posteriorly draining CCF, redirecting arterial flow from the orbit to the intracranial circulation.[4,11] Elevated capillary pressure from prolonged ophthalmic vein occlusion may reduce perfusion in the ophthalmic artery branches, resembling ischemic changes seen in central retinal vein occlusion. Additional contributing factors may include optic nerve ischemia from orbital congestion or transient hypoperfusion due to reduced internal carotid artery flow at the time of rupture or during fistula expansion.[3,5]

Although the visual outcome in PION is generally poor and irreversible,[3,9,10,12] early diagnosis and prompt treatment in this case appeared to prevent further deterioration and led to gradual improvement in ocular signs. Following neurosurgical TFCA,[13] serial HVF tests showed measurable visual field recovery,[6,14] indicating partial restoration of optic nerve function.

Interestingly, several weeks after intervention, the patient developed optic disc pallor, aligning with the expected delayed manifestation of optic atrophy often seen in PION.[10] While the primary management of direct CCF lies with neurosurgical intervention,[15] close ophthalmological monitoring remains essential to assess visual function, guide prognosis, and detect evolving signs such as optic atrophy.

CONCLUSION

This case highlights posterior ischemic optic neuropathy (PION) as a rare complication of direct carotid-cavernous fistula (CCF), marked by unilateral visual loss, normal optic disc but optic disc pallor is common 6-8 weeks later, and altitudinal visual field defect from Humphrey perimetry. The proposed mechanism, involving orbital venous congestion and an arterial steal phenomenon, provides new understanding of the pathogenesis of optic nerve ischemia. Early recognition and timely intervention led to partial visual recovery, challenging the typically poor prognosis of PION. This case underscores the critical role of ophthalmologists in early detection and supports the value of interdisciplinary collaboration to optimize visual outcomes in patients with CCF.

DECLARATION

ACKNOWLEDGMENTS

Ethics approval and consent to participate

The patient provided written informed consent for participation and publication of this case report.

Consent for publication

Not applicable.

Competing interests

The authors declare no conflict of interest.

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