

Indirect Carotid Cavernous Fistula : A Case Report

Introduction Carotid Cavernous Fistula (CCF) is a rare condition that refers to an abnormal connection between the cavernous sinus and carotid arteries. Indirect CCF mostly occur in older female patients. Indirect CCF can resolve spontaneously without surgical intervention. External manual compression of the carotid artery has been described as the treatment of choice in CCF, especially for indirect CCF.

Purpose To report a case of indirect CCF resolving with external manual carotid compression

Case Report A 61-years-old female with proptosis, lagophthalmos, episcleral injection, pain, headache, redness of the left eye, limitation of eye movement since 2 weeks before came to Cicendo Eye Hospital. His visual acuity was 0.3 on the left eye, contrast, and color sensitivity were within normal limit. She had hypertension and has been treated with antihypertension medication. She denied any history of trauma before. Her CT-Angiography revealed Type B CCF according to Barrow Classification. The patient planned to undergo Digital Subtraction Angiography (DSA) by Neuro-surgery Department. She was instructed to perform an external manual carotid compression while waiting for DSA. After 2 weeks the ocular motility and her symptoms were improving.

Conclusion Indirect type CCF can occur spontaneously and could be a sight-threatening condition especially in older female. Diagnosis confirmation should be done early for appropriate therapy of the patients. Patients with indirect CCF can resolve spontaneously with external manual compression.

Keyword carotid cavernous fistula, indirect, external manual compression

I. INTRODUCTION

Carotid Cavernous Fistula (CCF) is a rare but treatable cause of visual loss. Carotid Cavernous Fistula refers to an abnormal connection between the cavernous sinus and carotid arteries. Based on the etiology CCF can be caused by a spontaneous or acquired disease with the most common etiology is trauma. CCF can be classified according to the etiology, hemodynamic behaviors, or angioarchitecture of the vessels involved. Another classification of CCF according to Barrow classification can be divided into four types from type A, B, C, and D. Type A CCF can also be classified as direct, whereas type B, C, and D can be classified as indirect CCF. The incidence of traumatic CCF is more than 70% and usually occurred in young male patients

following closed head injury. Spontaneous CCF was reported for the remaining 30%, typically in older female patients.¹⁻³

Vision loss, intracerebral hemorrhage, subarachnoid hemorrhage, epistaxis, or cranial nerve palsies are complications of CCF. The goal of the therapy is to preserve flow and prevent further complications. Management of CCF includes endovascular embolization with coiling or liquid agents. The success rate of this intervention was 90% although recurrence may occur. Conservative management includes external manual compression especially in low risk, low flow, indirect CCF. Vision loss in CCF can be preventable with appropriate treatment.¹⁻³ Herein, we reported an improving case of indirect carotid cavernous fistula treated with external manual compression.

II. CASE REPORT

A 61-years-old female came to the Neuro-ophthalmology outpatient clinic Cicendo Eye Hospital with swelling and protrusion of the left eye for 2 weeks before admitted to the hospital. The complaint is also accompanied by limitation of eye movement, headache, pain, redness, tearing, and blurred vision on the left eye. She denied any double vision and history of trauma before. She has a history of hypertension and currently on antihypertensive medication. There was no history of diabetes mellitus, dyslipidemia, or other systemic diseases. She also had a history of wearing thick glasses in the right eye.

Physical examination revealed consciousness was fully alert with 138/88 on blood pressure and other vital sign was within normal limit. Other neurological examination was within normal limit. An ophthalmology examination revealed hypertropia of the left eye and 7⁰ exotropia, ocular motility on the right eye was within normal limit whereas in left eye was limited with -3 to nasal, inferonasal and inferotemporal, -2 to temporal and -1 to superonasal and superotemporal direction. Ocular bruit was negative on the left eye. Uncorrected visual acuity on the right eye (RE) was 1/60 and 0.3 on the left eye (LE) with no improvement on pinhole examination.

Refractometry on the right eye revealed S-9.75 C-7.25 x 152 and on the left eye was S+2.00 C-0.25 x42. Intraocular pressure (IOP) was 12 mmHg and 28 mmHg, respectively. Hertel examination revealed 16 mm on the right eye and 19 mm on the left eye.



Figure 1. Anterior Segment and Ocular Motility of the Left Eye

Anterior segment examination was within normal limit on the right eye. There was chemosis, episcleral injection, crockscrew appearance on conjunctiva, clear cornea. Light reflex revealed Grade I Relative Afferent Pupillary Defect (RAPD) on the left eye. There was lens opacities with NO3NC3 on both eyes according to The Lens Opacity Classification System (LOCS). Funduscopy examination on the right eye showed defined border optic disc with peripapillary atrophy and decreased of foveal reflex. The left eye showed optic disc swelling, artery to vein (A/V) ratio about 1:4 and tortousity of retinal blood vessel. Amsler grid, Ishihara color plate and contrast sensitivity was within normal limit on the left eye. Other neurological examination was within normal limit.

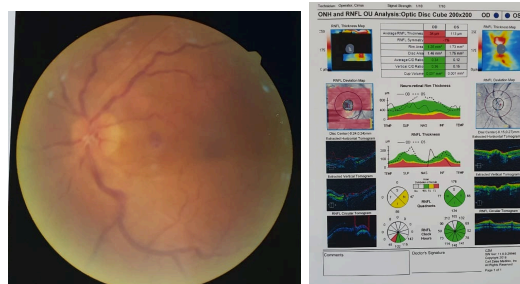


Figure 2. Fundus Photography and Optic Disc OCT of the Left Eye

Ocular Computed Tomography (OCT) of optic disc showed slightly increased Retinal Nerve Fiber Layer (RNFL) thickness on the left eye. Humphrey 30-2 examination showed low test reliability due to fixation loss. Computed Tomography (CT) Angiography revealed enlargement of left internal carotid artery in cavernous with abnormal connection from cavernous and enlargement of left superior ophthalmic vein suggested left carotid cavernous fistula B Type according to Barrow Classification. Contrast enhanced Computed Tomography (CT) Scan showed proptosis of the left eye with enlargement of left extraocular muscle.

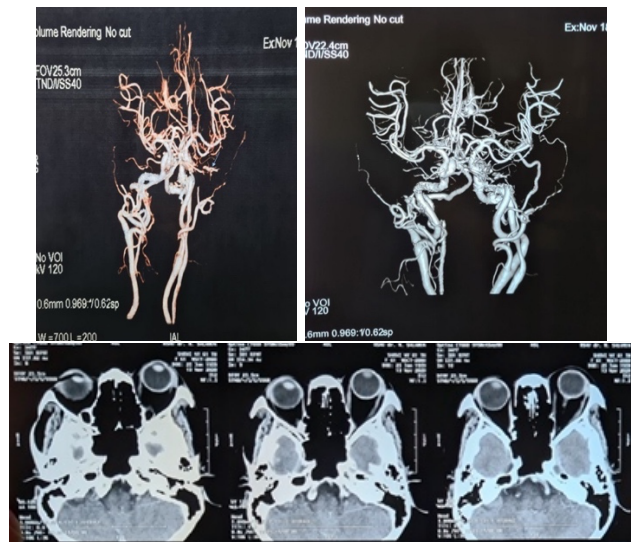


Figure 3. Computed Tomography Scan and CT Angiography of the Patient

The patient was diagnosed with Left Indirect Carotid Cavernous Fistula, Bilateral Immature Cataract, Suspected with Impending Central Retinal Vein Occlusion of LE, Secondary Glaucoma LE, RE High Myopia and Suspected with RE Macular Atrophy. Patient was consulted to Glaucoma unit and Neuro-surgery Department for further evaluation. The patient was treated with oral Citicholine tablet 1x1000mg, artificial tears eye drop 6x1 drop LE, Timolol Maleate 0.5% eye drop 2x1 drop LE, and advised to do external manual carotid compression. Neuro-surgery department are

planning to do Digital Subtraction Angiography (DSA) to this patient. This patient also plan to consult to Vitreoretina Unit.

Patient then came to follow up 2 weeks later with improvement on her condition. Left eye ocular motility was improving with -1 in all direction. Uncorrected visual acuity in the LE was 0.3 with 0.63 on pinhole examination. Intraocular pressure (IOP) on the left eye was 13 mmHg on antiglaucoma therapy. Anterior segment examination revealed chemosis, episcleral injection, crockscrew appearance on conjunctiva. Light reflex revealed Grade I Relative Afferent Pupillary Defect (RAPD) on the left eye. Funduscopy examination on the left eye optic disc swelling, artery to vein (A/V) ratio about 1:4 and tortuosity of retinal blood vessel. Amsler grid, Ishihara color plate and contrast sensitivity was within normal limit on the left eye. The patient still advised to do the external manual carotid compression until DSA procedure. Prognosis in this patient *quo ad vitam dubia ad bonam, quo ad functionam dubia ad bonam, quo ad sanationam dubia ad bonam*.



Figure 5. Clinical Presentation in 2 Weeks Follow Up and Illustration of External Manual Carotid Compression

III. DISCUSSION

Carotid cavernous fistula is a rare condition caused by abnormal connection between carotid arteries and cavernous sinus. The ocular venous outflow originate

from arcades of retinal veins and drains into central retinal vein and choroidal veins. Episcleral venous plexus collects blood from uveal circulation and aqueous through Sclerom canal. These veins empty mainly to the superior ophthalmic vein. Superior and inferior ophthalmic veins then drains blood into cavernous sinus. Cavernous sinus is a venous channels located at middle cranial fossa, bilaterally to the sella turcica. Internal Carotid Artery (ICA) enters the cavernous sinus via foramen lacerum and surrounded completely by venous blood. The nerves of the cavernous sinus are Cranial Nerves (CN) III, IV, V1, V2, VI and sympathetic plexus around the internal carotid artery. Cranial nerves III, IV, V1 and V2 are attached in the lateral wall of cavernous sinus. The CN VI enters the cavernous sinus and surrounded by venous blood, like internal carotid artery. Carotid cavernous fistula can cause increased intercavernous pressure and leads to compression in adjacent structure. The patient can present with total ophthalmoplegia due to injury of CN III, IV, VI as seen in this patient.⁴⁻⁷

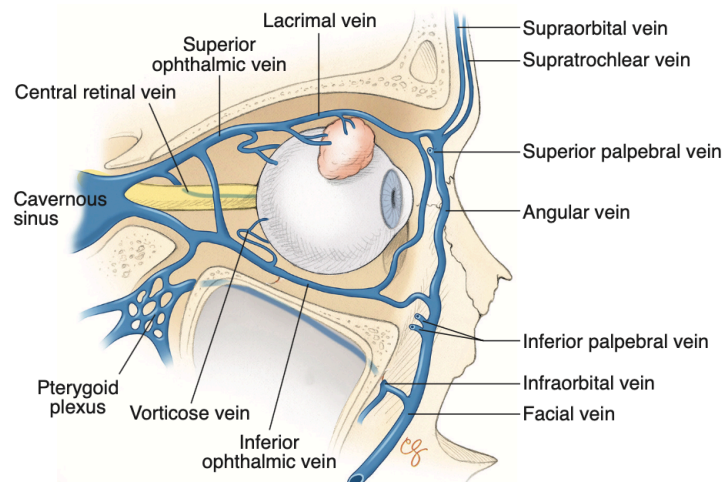


Figure 6. Ocular Venous Drainage

Source : Cantor LB⁷

Carotid Cavernous Fistula can be classified by their cause (spontaneous or traumatic), haemodynamic behaviours (high or low flow), and angioarchitecture (direct and indirect). Barrow et al. classified CCF by arterial system involved into 4 types including type A, B, C, and D. Type A or direct CCF is a fistula between

cavernous sinus and ICA. Type B to D included in indirect CCF, type B are dural fistula between meningeal branches of ICA and cavernous sinus, whereas type C are abnormal connection between meningeal branches of ECA and the cavernous sinus, and type D are both branches of internal and external carotid artery. Majority of Type A are high-flow lesion, it has acute onset of symptoms and rarely resolving spontaneously.^{2,3,5,6} This patient had type B CCF based on CT Angiography examination in which indirect CCF.

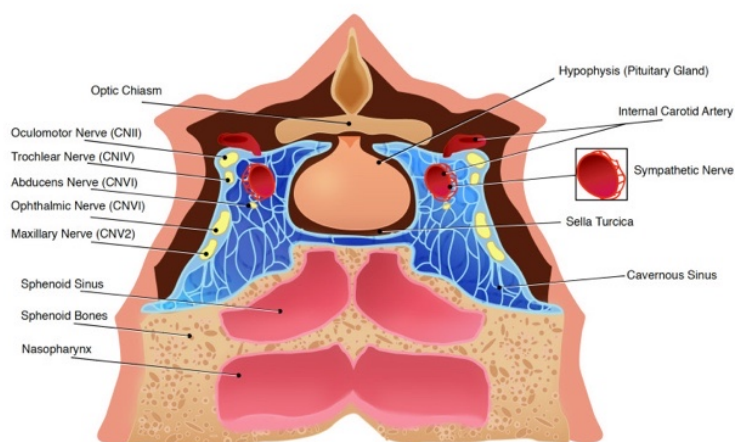


Figure 7. Cavernous Sinus Anatomy

Source : Massa⁴

Direct CCF occur mostly after head injury, whereas indirect or spontaneous CCF occur secondary to spontaneous rupture of dural arteries wall which passed through the sinus. Indirect CCF occur mostly in elderly women and associated with hypertension, vascular diseases, atherosclerosis, pregnancy, and connective tissue disorder. It has been postulated that indirect CCF occur due to ruptured of dural arterial wall, however the exact mechanism still poorly understood.^{6,8,9} This patient has risk factor of indirect CCF such as elderly women with hypertension.

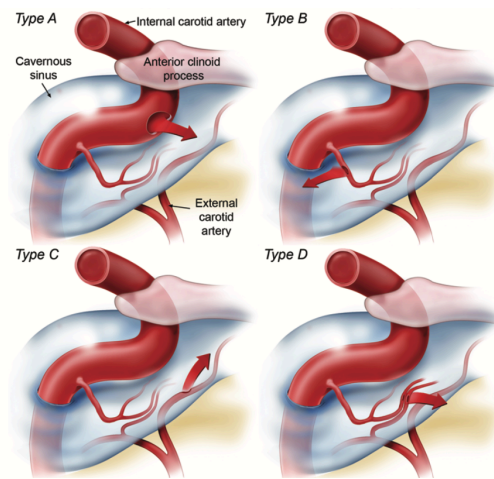


Figure 7. Type A to D Barrow Classification of CCF

Source : Peng T¹⁰

Clinical presentation of CCFs is variable depending on the arterial and venous outflow involved. Superior and inferior ophthalmic vein involved in CCF may demonstrate proptosis or pulsatile exophthalmos. Extraocular muscle enlargement, conjunctival vessel arteriolization, chemosis, eyelid edema also occur due to venous congestion in the orbit. Decreased visual acuity may occur from combination of following mechanism including exposure keratopathy, venous stasis retinopathy, central retinal vein occlusion with macular edema or choroidal detachment due to venous congestion. Glaucoma can developed because of increasing in episcleral venous pressure. Increased of venous pressure and vascular compromise may cause ischemic optic neuropathy. Ocular ischemia may also cause neovascular glaucoma. The engorged cavernous sinus may compress cranial nerves and cause diplopia in posterior or inferior venous drainage involvement. Compression of CN V1 and V2 may cause decreased of facial sensation in ipsilateral face.^{2,5,6} This patient presented with headache, chemosis, proptosis, conjunctival arteriolarization, secondary glaucoma and retinal vein tortuosity suggested as impending central retinal vein occlusion as clinical presentation of CCF. Ophthalmoplegia associated with CN III, IV and VI and ocular misalignment found in this patient probably caused by compression in the CN or vascular steal.

Diagnosis workup to confirm CCF was made by neuroanatomic and neurovascular imaging. First line modalities are CT or Magnetic Resonance Imaging (MRI) with angiography since both are non-invasive and quick. These examination provide imaging include proptosis, dilatation of vessel, extraocular muscle enlargement, cavernous sinus enlargement, and skull imaging. Digital Subtraction Angiography (DSA) is the current gold standard in CCF diagnosis and should be performed before any intervention. This examination provide information on associated vascular injuries, flow velocity and detect haemodynamic processes.^{2,3,6} We performed CT Angiography on this patient in which revealed type B CCF and also planning to do the DSA by Neurosurgery Department.

Therapeutic intervention for CCF treatment include conservative management, endovascular intervention, open surgery and radiosurgery. The goal of treatment is to preserve flow in the internal carotid artery while occlude the fistula. Conservative management can be done in low risk, low flow or indirect CCF. Patients with indirect CCF exhibit spontaneous fistula closure in 20-60% patients. Direct CCF rarely resolved spontaneously and usually require treatment. Emergency treatment in CCF needs to be done in patient with progressive proptosis, visual decline, haemorrhage, and increased intracranial pressure. Transarterial or transvenous embolization remained the first-line treatment modality in CCF, over 90% cases was successfully cured with this approach. Although has high successful rate it also associated with risks of thrombosis and reopening of the fistula.^{3,5,6}

External manual carotid compression can promote resolution of the CCF. The patient instructed to compresses the ICA on the involved side using contralateral hand. The use of contralateral hand if ischaemia occurs, the hand will release its pressure on the artery due to hemiparesis. This procedure success rate reported to be 35% with resolution occurring between 2 weeks to 7 months after initiation of the procedure. External manual carotid compression is not without risks, the contraindication of this procedure are patient with atherosclerotic disease or patient with significant risk of stroke occurrence as the result of insufficient carotid flow and

embolic complication. Arrhythmias, heart block and syncope can occur as a result of stimulation in the carotid nerve.^{11,12} This patient instructed to perform external manual carotid compression and presented with resolving of her condition in the follow up period without any complication.

Carotid cavernous fistula can cause complication such as loss of vision, intracerebral haemorrhage, subarachnoid haemorrhage, epistaxis, or cranial nerve palsies especially in high flow fistula. Patients with direct or indirect CCF usually has reassuring visual prognosis unless there is evidence of optic nerve or retinal ischemia before treatment. Embolization therapy has high successful rate and provide resolution of bruit and IOP. Proptosis, chemosis and ocular misalignment usually improved within weeks of CCF closure and resolve within 3 months. Recurrence may occur in young patients the risk is lower with complete occlusion of the fistula.^{6,13}

IV. CONCLUSION

Carotid cavernous fistula is a rare condition and can be sight threatening. Ophthalmologist should be familiar with the symptoms. A thorough evaluation should be done to help differentiation of the diagnosis. An appropriate examination should be decided for diagnosis confirmation. Treatment should be done early to preserve vision and prevent other CCF complications rather than waiting for spontaneous thrombosis. Long term ophthalmologic follow up needs to be done because of the possibility of recurrence of the condition.

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