DEPARTEMEN OF OPHTHALMOLOGY

MEDICAL FACULTY PADJADJARAN UNIVERSITY

NATIONAL EYE CENTRE CICENDO EYE HOSPITAL

BANDUNG

Case Report	:	Dural Arteriovenous Fistule with Compressive Optic Neuropathy and Secondary Glaucoma, a case report
Presenter	:	Dewi Kania Maemunah
Consultant	:	Bambang Setiohadji, MD

Approved by

Neuro Ophthalmology Unit Consultant

Bambang Setiohadji, MD

Dural Arteriovenous Fistule with Compressive Optic Neuropathy and Secondary Glaucoma, a case report

Abstract

Introduction

Dural carotid-cavernous fistulae (CCF) are characterized by a communication between the cavernous sinus and one or more meningeal branches of the internal carotid artery, external carotid artery, or both. The fistula may occur spontaneously. Patients with CCF generally present with varied complaints, including unilateral visual loss, proptosis, chemosis of conjunctiva, and raised intraocular pressure.

Purpose

To report a case of dural arteriovenous fistule with compressive optic neuropathy and secondary glaucoma.

Case Report

A 42 years old male, presenting with proptosis and suffered pain of left eye about five weeks ago. Previous history was persistent redness & painless of left eye since ten months, with no bruit. Visual acuity (VA) was 1.0 in the right eye and closed to face finger counting in the left eye. Esotropia in primary gaze and restrictive on ocular movement to all of direction with RAPD grade II on the left eye, raised intraocular pressure 21mm Hg and conjuctival injection. Funduscopic examination showed unilateral optic disc swelling and turtousity of retinal vessels on the left eye. Magnetic Ressonance Angiography of the left eye revealed enlarged of superior ophthalmic vein and there was communication between meningeal medial artery and left superior ophthalmic vein. He was diagnosed with Dural arteriovenous fistule with compressive optic neuropathy and secondary glaucoma of the left eye. The patient was treated with intravenous methylprednisolone 4x250 mg, ranitidine 2x50 mg, mecobalamine 1x500 mcg, timolol maleat and latanoprost eyedrop. The patient was reffered to neurosurgery department for Digital Sustract Angiography (DSA).

Conclusion

Dural CCF usually become symptomatic spontaneously. Complications include vision loss caused by optic neuropathy and raised episcleral venous pressure that may produce increased intraocular pressure.

Keywords

dural arteriovenous fistule, compressive optic neuropathy, secondary glaucoma

I. Introduction

Carotid-cavernous sinus fistula (CCF) is an abnormal communication between the internal or external carotid arteries and the cavernous sinus. Carotid-cavernous sinus fistulae occur because of traumatic or spontaneous rents in the walls of the intracavernous internal carotid artery or its branches. This results in

short-circuiting of the arterial blood into the venous system of the cavernous sinuses. [1,2]

These lesions may be classified based on the following: etiology (traumatic vs spontaneous), velocity of blood flow (high vs low), and anatomy (direct vs dural, or internal carotid vs external carotid).[2]

Direct carotid-cavernous sinus fistulae, which represent 70-90% of all carotid-cavernous sinus fistulae in most series, are characterized by a direct connection between the intracavernous segment of the internal carotid artery and the cavernous sinus. These fistulae usually have high rates of arterial blood flow and most commonly are caused by a single traumatic tear in the arterial wall. [2,3]

Dural carotid-cavernous sinus fistulae are characterized by a communication between the cavernous sinus and one or more meningeal branches of the internal carotid artery, external carotid artery, or both. The clinical presentation of a CCF is influenced by the type and size of the fistula as well as its location, blood flow rate and drainage route. Posterior drainage usually produces no ocular symptoms, although some patients may experience a cranial neuropathy, such as facial paresis or ocular motor nerve paresis. Ocular signs and symptoms typically arise as the drainage shifts to an anterior route via the superior and inferior ophthalmic veins. Because of their lower flow rate, dural CCFs usually produce less severe symptoms than those from a direct CCF. Objective and subjective bruits also are less common with dural fistulas.

II. Case Report

A 42 years old male, presenting with proptosis and suffered pain of left eye about five weeks ago. He also complaint nausea, vomit and headache. He didn't hear bruit in left eye. He was complaint with persistent redness & painless of left eye since ten months, he get antiglaucoma agent but not relieved by medicines. No history of trauma, hypertention and diabetes mellitus.

Vital signs examination was within normal limit; blood pressure was 120/80 mmHg, respiration rate was 20x/m, heart rate was 80x/m, and temperature was 36 ^{o}C. Initial ophthalmologic examination showed visual acuity (VA) using

Snellen chart to be 1.0 in the right eye and Closed to face finger counting in the left eye. Eye position in primary gaze was exotropia about 15^0. Ocular movement was restrictive abduction and adduction of the left eye upon left gaze on a scale of -2, and restrictive up gaze and down gaze on scale of -3 compatible with third nerve palsy and sixth nerve palsy (Figure 2.1).

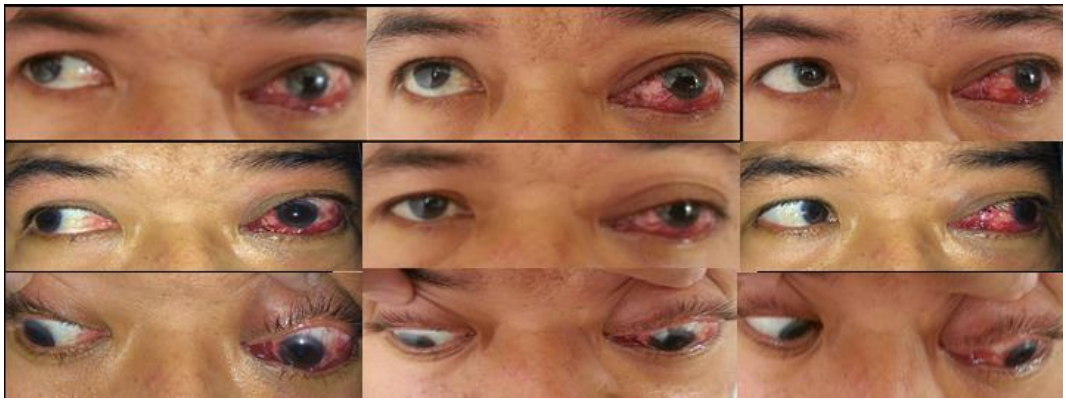

Figure 2.1 Restrictive in abduction, adduction, up gaze and down gaze

Anterior segmen on the right eye within normal limit except the pupillary reflex showed decrease in indirect examination. Colour test, Amsler grid test and contras sensitivity on the right aye was normal. External examination on the left eye showed proptosis and episcleral injection diffused as "crockscrew" sign with relative afferent pupillary defect, raised intraocular pressure 21mm Hg and conjuctival injection. Dilated funduscopic examination showed optic disc swelling with turtousity of retinal vessels (figure 2.2). Colour test, Amsler grid test and contras sensitivity test are difficult.

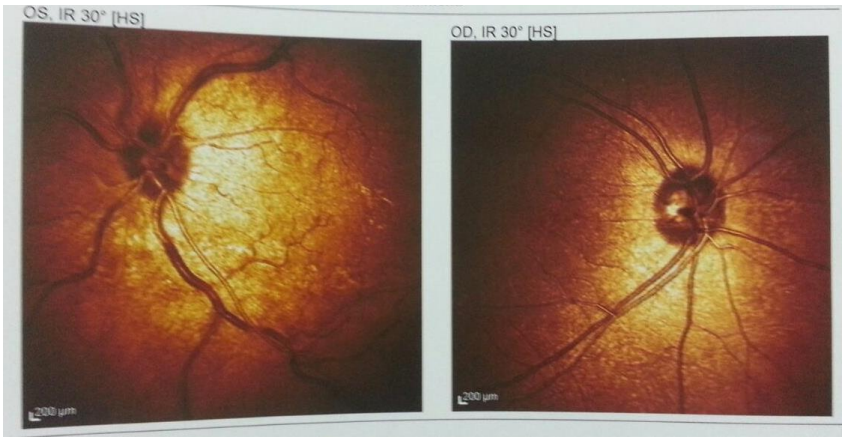

Figure 2.2 Fundus photograph showed swelling optic disc with turtousity of retinal vessels of left eye

Visual field examination using Humphrey 30-2 revealed poor reliability test but showed normal on the right eye meanwhile Humphrey 30-2 visual field on the left eye cannot identifiable (figure 2.3). Ocular computed tomography (OCT) revealed normal disc on right eye and swelling disc on the left eye (Figure 2.4).

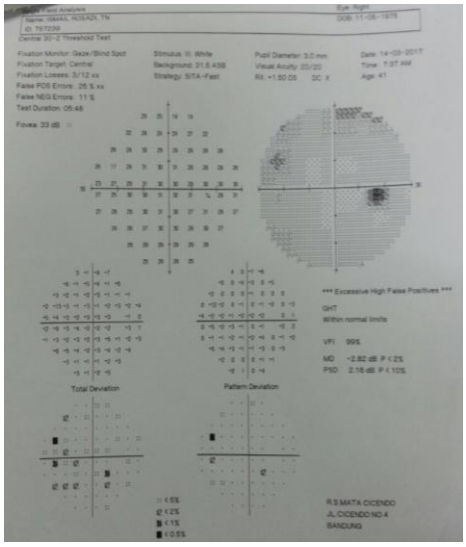

Figure 2.3 poor reliability test with unspesified visual field defect on the right eye

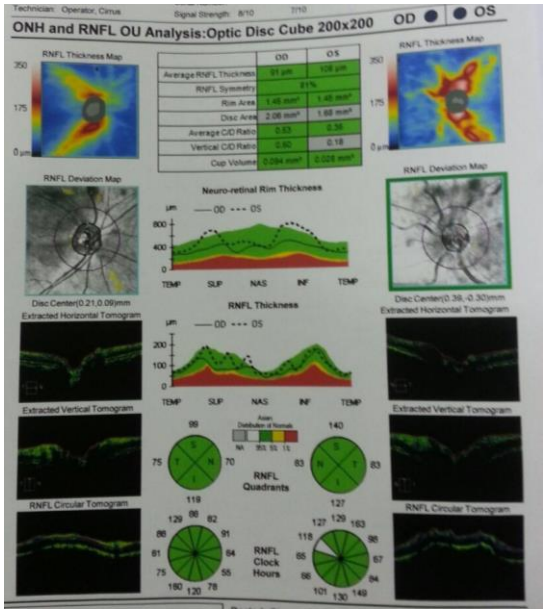

Figure 2.4 Ocular computed tomography (OCT) revealed normal disc on right eye and swelling disc on the left eye

Orbital-head Computed Tomography (CT) scan and Magnetic Ressonance Imaging (MRI) showed proptosis and enlarged of all the extraocular muscle of the

left eye. Magnetic Ressonance Angiography of the left eye revealed enlarged of superior ophthalmic vein and there was communication between meningeal medial artery and left superior ophthalmic vein.

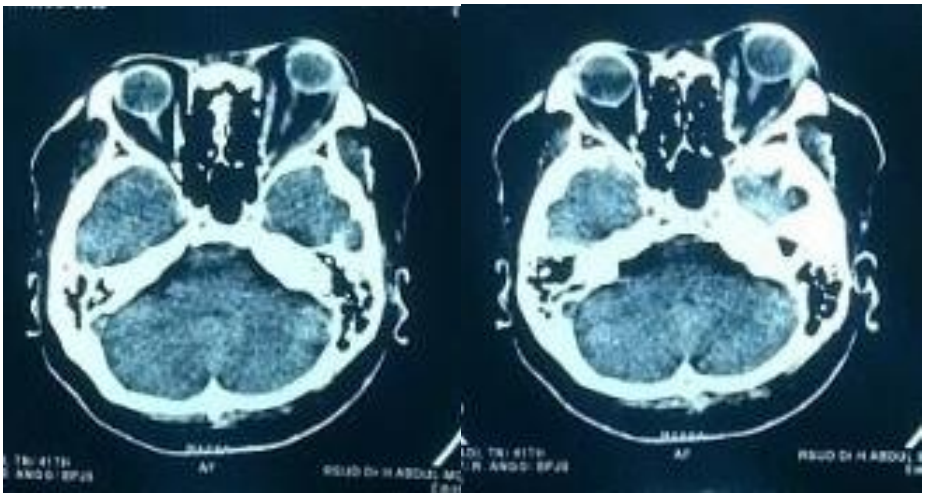

Figure 2.5 Orbital-head CT scan showed proptosis and enlarged of all the extraocular muscle of the left eye

Based on the history, clinical features and neuroimaging, the patient was diagnosed Dural Carotid-Cavernous Fistule with Compressive Optic Neuropathy and Secondary Glaucoma of the left eye. The patient was treated with intravenous methylprednisolone 4x250mg for 3 days continuing to oral methylprednisolone 1mg/kg body weight, ranitidine 2x 50mg, mecobalamin 1x500 mg, prostaglandine analogue eye drop once a day and Timolol maleat eye drop twice a day on the left eye. Pastient was reffered to neurosurgery departement and planned Digital Substract Angiography (DSA). The prognosis was dubia because need cloosed observation in non-ocular symptoms.

On third day follow up, visual acuity on the left eye increased to 0,08. External examination on the left eye showed proptosis and episcleral injection with relative afferent pupillary defect, intraocular pressure still 21 mm Hg. Dilated funduscopic examination still showed optic disc swelling with turtousity of retinal vessels. Therapy was continued with Metylprednisolone oral tappered.

III. Discussion

Carotid cavernous fistula (CCF) is an abnormal communication between the carotid arterial system (Internal Carotid Artery or External Carotid Artery) and the venous cavernous sinus.[3]

Barrow et al Classified CCF into four angiographic types based on arterial supply. Type A fistulas are direct communications between the internal carotid artery and the cavernous sinus. Types B, C and D are indirect shunts (dural), because fistulas to the cavernous sinus arise from dural arteries and not directly from the internal carotid artery. Direct CCF (Type A) usually occurs in young men secondary to trauma. Indirect CCF (dural CCF) usually arising from dural branches of either internal carotid artery (ICA) (Type B) or external carotid artery (ECA) (Type C) or both (mixed or Type D).[3,4]

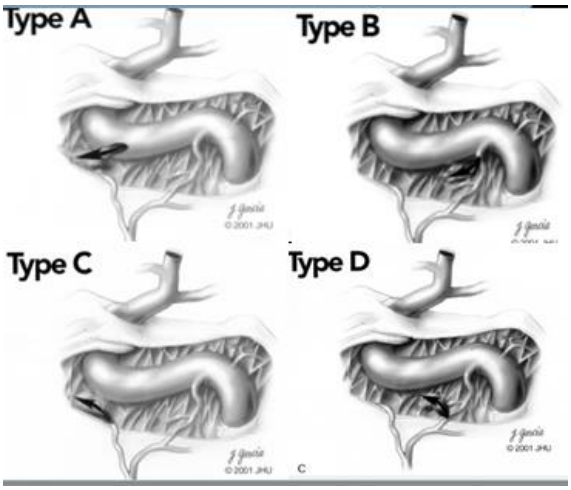

Figure 3.1 Artist's drawing of type of fistula[2]

In this case, a 42 years old man came with a chief complaint of presenting proptosis and suffered pain of left eye about five weeks ago. Ophthalmologic examination showed decreased visual acuity to closed to face finger counting, restrictive on ocular movement to all of direction with RAPD grade II on the left eye, raised intraocular pressure 21mm Hg and conjuctival injection. On dilated funduscopic examination showed unilateral optic disc swelling and turtousity of retinal vein on the left eye. This clinical features are suggestive to carotid-

cavernous fistule with no bruit as low flow or indirect of CCF with compressive optic neuropathy and secondary glaucoma.

Common clinical signs include proptosis, orbital bruit, chemosis, extraocular palsy, pulsating exophthalmos, ptosis, elevated intraocular pressure, anterior segment ischaemia and retinopathy. Patients often complain of a swollen red eye, orbital pain, diplopia, headache and progressive vision loss.[1-4]

The neuroimaging in this patient revealed suggestive of CCF on MRI. There were orbital congestion, thickened extraocular muscles, exophthalmus, dilation of superior opthalmic vein and meningeal media artery communicated with superior ophthalmic vein on MRA.

The abnormal communication results in high-pressure arterial blood entering the low-pressure venous cavernous sinus, which interferes with the normal patterns of venous drainage and which compromises blood flow into the cavernous sinus and globe.[3,5,6]

Indirect CCF have a more gradual onset over days, weeks, or months, a less fulminating course and generally lower flow like in this case that revealed no bruit that suggestive indirect CCF. Patients with an indirect type fistula generally present with varied complaints, including unilateral visual loss, visual loss, proptosis, elevated intraocular pressure, optic disc edema, and dilated and tortuous retinal vessels.[2]

The blood supply to the region of the cavernous sinus is provided by interconnecting branches of the internal and external carotid arteries, and it is from these vessels that dural CCF often called dural arteriovenous fistula. Such fistulas usually are separated anatomically into three types: (a) shunts between meningeal branches of the internal carotid artery and the cavernous sinus, (b) shunts between meningeal branches of the external carotid artery and the cavernous sinus, and (c) shunts between meningeal branches of both the internal and external carotid arteries and the cavernous sinus.[2,3]

Dural CCF usually become symptomatic spontaneously. The pathogenesis of these fistulas is controversial. Newton and Hoyt speculated that spontaneous

dural CCF form after rupture of one or more of the thin-walled dural arteries that normally traverse the cavernous sinus.[2]

Complications include vision loss and raised episcleral venous pressure may produce increased intraocular pressure that occasionally can be quite high. Visual loss, although less frequent than in patients with direct carotid-cavernous fistula, occurs in 20% - 30% of patients with dural CCF. It may be caused by compressive optic neuropathy, chorioretinal dysfunction, or uncontrolled glaucoma. In this patient there was compressive optic neuropathy and secondary glaucoma that may be caused by raised episcleral venous pressure.[2,7]

Funduscopy examination in this patient revealed swelling of the optic nerve head and there was turtousity of retinal vessel suggestive compressive optic neuropathy caused by increased episcleral venous pressure and raised intraocular pressure. Methylprednisolone was given to control the swelling of the optic nerve and administered antiglaucomatous topical medication.

It is important to distinguish between direct (type A) and indirect (Types B-D) fistulas because of the prognostic implications. Urgent treatment is usually needed for direct and high flow fistulas in which endovascular embolization is mostly applied. Indirect, slow flow CCFs usually close spontaneously without treatment.[2]

In the old days, CCF was treated with surgical operations with significant morbidity and mortality, In the modern era, neuroendovascular therapy offers a safe and effective treatment for patients with CCF and has replaced open surgery as the treatment of choice. The definite therapy is obliteration of the fistulous connection with restoration of normal arterial and venous flow.[8,9,10] In this case, the patient was reffered to neurosurgery departement and planned with Digital Substract Angiography that still a gold standard for diagnosis, valuable for categorization of CCF and verification of their drainage patterns and allows for the planning and performance of the therapeutic procedure.

After the successfully closure of the fistula, almost all symptoms and ocular signs disappear or improve. Ocular pulsation, eyelid edema, conjunctival chemosis, conjunctival vessels dilation, stasis retinopathy, optic disc swelling and

raised intraocular pressure disappear immediately. Exophthalmia decreases but it may not disappear entirely.[7,11]

IV Conclussion

Dural CCF usually become symptomatic spontaneously. Non-invasive Magnetic Ressonance Imaging and Angiography is helpful for diagnosing CCF. Complications include vision loss caused by optic neuropathy and raised episcleral venous pressure that may produce increased intraocular pressure. Methylprednisolone intravenous was an appropriate treatment for compressive optic neuropathy caused by CCF. The definitive treatment was depend on location of communicate between arteriovenous fistule. neuroendovascular therapy offers a safe and effective treatment for patients with CCF.

REFFERENCES

1. Karaman E, Isildak H, Haciyev Y, Kaytaz A, Enver O. Carotid-cavernous fistula after functional endoscopic sinus surgery. J Craniofac Surg. 2009 Mar. 20(2), hal. 556-8
2. Miller, NR.; Newman, NJ., Walsh & Hoyt's Clinical Neuro-Ophthalmology, Edisi ke-6. Lippincott Williams & Wilkins, 2005; hal. 2264-96
3. Kumar K, Carotid-Cavernous Fistula; Diagnosis with Magnetic Resonance Imaging. European Journal Of Pharmaceutical and Medical Research 2016; 3(3),hal. 313-20.
4. Barrow DL, Spector RH and Braun IF. Classification and treatment of spontaneous carotid-cavernous sinus fistulas. J Neurosurg 1985; 62, hal. 248-256.
5. Low HL, Sawlani V and Redfern RM. Carotid-cavernous fistula after blunt traumatic to the neck. British Journal Neurosurgery 2006; 20: hal. 254-9
6. Hu WY and Hudon M. Neuroimaging highlight: traumatic carotid-cavernous fistula. Can J Neurol Sci., 2000; 27, hal. 71-72
7. Petraru D, Costin D, Mihalache G, Direct carotid-cavernous fistula and glaucoma: case reports, Romanian Neurosurgery, 2011; XVIII 1: hal. 82 – 7
8. Cheng KM., Neuro-endovascular Therapy of Carotid-Cavernous Fistula. The Hongkong Medical Diary. 2006; 11(12). hal.13-4
9. Murthy, Mohan C, Gupta P, P. Bedi, T. Prabhakar, Caroticocavernous Fistula - A Case Report, Indian Journal Anaesthesia. 2005; 49 (3) : hal. 220 – 2
10. Halbach VV., Hieshima GB., Higashida RT., Reicher M, Carotid Cavernous Fistulae: Indications for Urgent Treatment, ANJR; 1987 (8): hal. 627-33
11. Espeela PHB., Guéritb JM., Goffettec P, Bilateral carotid-cavernous fistula following head trauma: Possible worsening of brain injury following balloon catheter occlusion? Clinical Neurology and Neurosurgery ,2006; 108(6), hal.576-9