Direct Carotid-Cavernous Fistula in a Filipino Female Presenting With Simultaneous Orbital/Ocular, Cavernous and Cortical Symptomatology Without History of Trauma: A Case Report

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ABSTRACT

Carotid-cavernous fistula (CCF) is a rare and dangerous neurological disorder that arises due to an abnormal communication between the internal carotid artery (ICA) or the external carotid artery (ECA) and their branches and the cavernous sinus. It can either be a direct fistula (high-flow with acute symptoms) most commonly resulting from trauma (70-90%) or an indirect fistula (low-flow with insidious symptoms) secondary to hypertension, atherosclerosis and collagen vascular disorders. The shunting of arterial blood into the venous system leads to venous hypertension causing various clinical manifestations depending on the venous drainage patterns and the shunt flow. Increased anterior, posterior and superior venous drainage results to orbital/ocular, cavernous and cortical symptomatology, respectively. This paper aims to present a case of 58-year old Filipino female with a 2-day history of sudden, severe headache, vomiting and blurring of vision followed by decrease in sensorium and sudden proptosis and chemosis of the left eye. Patient had no co-morbidities, history of trauma, surgeries, facial skin infections or prior febrile illness. The left eye had exophthalmos, subconjunctival hyperemia, scleral edema/chemosis and ocular bruit. Neurologic examination showed a stuporous patient with multiple cranial nerve deficits (impaired direct and consensual pupillary reflex left, complete ptosis left, sluggish corneal reflex left, impaired oculocephalic reflex left), right hemiplegia and meningeal signs. Cranial Computed Tomography (CT) Angiogram revealed an acute parenchymal hemorrhage in the left frontotemporal lobe with subarachnoid component, with engorged left cavernous sinus and dilated left superior ophthalmic vein. Digital Subtraction Angiography (DSA) was done revealing a direct type of left carotid-cavernous fistula with massive ICA shunting to the cavernous sinus, superior ophthalmic vein and inferior petrosal sinus. The clinical and radiographic evidence were consistent with a Direct/Type A CCF. Unique in this case was a patient with no history of trauma presenting with simultaneous orbital/ocular, cavernous and cortical symptomatology – a clinical picture of CCF that has never been documented in any literature nor included in any classification system. The presence of all three symptomatology can be explained by a direct/high-flow fistula that resulted to increased anterior, posterior and superior venous drainage as documented in the DSA. In addition, spontaneous intracranial hemorrhage in CCF is exceptionally rare and it is the most daunting symptomatology of this disease. With that, this specific case may pave the way to a new classification scheme and determine its corresponding treatment approach.

Keywords: *Carotid-cavernous fistula, cavernous sinus, orbital/ocular, cavernous, cortical*

INTRODUCTION

The cavernous sinus is a large, irregular space measuring 2 cm long and 1 cm wide located on each side of the sphenoid sinus, sella turcica and pituitary gland, which forms a network of intercommunicating venous channels enclosing the internal carotid artery and the abducens nerve[1, 2]. Each cavernous sinus may also be considered as a confluence of sinuses because anteriorly it receives the two ophthalmic veins and posteriorly it empties into the superior and

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inferior petrosal sinuses, through which it is connected with the transverse sinus and the bulb of the internal jugular vein[1], respectively.

A carotid-cavernous fistula (CCF) is a rare sight- and life-threatening disorder that arises due to an abnormal communication between the internal carotid artery (ICA) or the external carotid artery (ECA) and their branches and the cavernous sinus[3,8]. The simplest classification divides CCFs into direct and indirect – (1) direct fistulas are high-flow fistulas with acute onset of symptoms resulting from trauma (70-90%), a defect in the ICA wall, or rupture of a cavernous ICA aneurysm; and (2) indirect fistulas are spontaneous, low-flow fistulas with insidious onset of symptoms occurring secondary to hypertension, atherosclerosis, neurofibromatosis and collagen vascular disorders such as fibromuscular dysplasia or Ehlers–Danlos syndrome, comprising the majority of CCFs encountered in clinical practice[3, 4, 5, 10]. The angiographic classification developed by Barrow et al. divides CCFs into 4 types: Type A CCFs are direct connections between the cavernous ICA and cavernous sinus, while Types B, C, and D CCFs are indirect connections between the cavernous sinus and branches of the ICA (type B), branches of the ECA (type C), and branches of the ICA and ECA (type D)[5].

In CCFs, arterialized blood is shunted into the venous system leading to venous hypertension as well as increased arterial pressure and decreased perfusion[4,5]. The symptomatology of CCF can either be orbital/ocular, cavernous or cortical depending on the venous drainage patterns and shunt flow as follows – (1) increased anterior venous drainage (ophthalmic veins) results in an increase in intraocular pressure leading to orbital congestive symptoms; (2) increased posterior venous drainage (petrosal sinuses) results in cranial nerve deficits such as ophthalmoplegia, diplopia, ptosis, or anisocoria encompassing the cavernous symptoms; and (3) retrograde superior drainage into the superficial middle cerebral vein may result in cortical manifestations such as cerebral hemorrhage, seizures or venous infarctions[1, 3, 7, 10].

Angiography gives the definitive diagnosis of CCF. First-line modalities include color Doppler ultrasonography, computed tomography (CT) angiography, and magnetic resonance (MR) angiography; however, the gold-standard is Digital Subtraction Angiography (DSA) due to its superior capability to accurately localize lesions for endovascular management[5, 8]. The main goal in the treatment of CCFs is to preserve flow in the ICA while occluding the fistula. Endovascular management is the current treatment modality of choice for CCFs. Transarterial embolization is the preferred access method for direct CCFs and transvenous embolization is preferred for indirect CCFs[5].

This paper aims to present a case of a direct carotid cavernous fistula presenting with simultaneous ocular/orbital, cavernous and cortical symptomatology, which has not been documented in any available literature.

CASE PRESENTATION

This is the case of a 58-year old Filipino female, single, "ukay-ukay" vendor who presented with a sudden onset of severe headache two days before admission while staying at home with two bouts of vomiting and blurring of vision in the left eye. Patient then became non-conversant due to severe pain and was noted to have right-sided weakness upon arrival at the Emergency Room. She was admitted and initially managed as a case of Cerebrovascular disease, Lobar hemorrhage, left frontotemporal lobe with subarachnoid component, and was noted to have decreasing sensorium over the next hours. On the first hospital day, the patient was noted to have sudden proptosis and chemosis of the left eye, hence there was a reconsideration of the patient's underlying condition. Patient is a nulligravid with no known co-morbidities, no history of trauma or surgical procedures, no prior spectacle or

contact lens use, no skin infections on the face and no preceding febrile illness prior to the onset of symptoms such as rhinitis, sinusitis, ear infection or dental caries.

Vital signs were as follows: blood pressure of 180/100, heart rate of 85, respiratory rate of 20 and temperature of 36.8 C. Physical examination of the right eye was normal but the left eye was noted to have exophthalmos, subconjunctival hyperemia, scleral edema/chemosis, and no discharges but with ocular bruit (Figure 1). Both eyeballs were soft on finger tonometry. Neurologic examination showed a stuporous patient with no eye opening on the left upon painful stimuli, no verbal output and was unable to follow commands. Higher cortical functions could not be assessed. Funduscopic findings on both eyes revealed positive red orange reflex, clear media, arteriovenous (AV) ratio of 2:3 on the right and 1:3 on the left, distinct disc borders, no hemorrhage or exudates and no papilledema. Other cranial nerve findings revealed an impaired direct and consensual light reflex (left), complete ptosis (left) with impaired horizontal and vertical oculocephalic reflex (left) and sluggish corneal reflex (left). Other cranial nerve functions were not assessed. Patient had no spontaneous movement of the right upper and lower extremities, with grimace upon deep nailbed pressure of the right extremities. There was hyperreflexia on the biceps, triceps, brachioradialis, quadriceps and ankles reflexes (right) with positive Babinski sign (right). Nuchal rigidity and Kernig's sign were also elicited.

Upon admission, patient underwent Plain Cranial CT scan revealing an acute parenchymal hemorrhage in the left frontotemporal lobe with subarachnoid component and exophthalmos of the left orbit (Figure 2). Cranial CT Angiogram done revealed an engorged left cavernous sinus and dilated left superior ophthalmic vein with no evidence of thrombus, aneurysm or malformation (Figure 3).

The above-mentioned findings were suggestive of a fistula between the left internal carotid artery and left cavernous sinus, hence a Digital Subtraction Angiography (DSA) or Four-vessel angiography was done that revealed a direct type of carotid-cavernous fistula in the left, creating a fistulous pouch measuring 20 mm in diameter and with massive ICA shunting to the cavernous sinus, superior ophthalmic vein and inferior petrosal sinus (Figure 4 – A and B). Medical decompression and blood pressure control

Figure 1. Orbital congestion of the patient manifested as exophthalmos, subconjunctival hyperemia and scleral edema/chemosis.

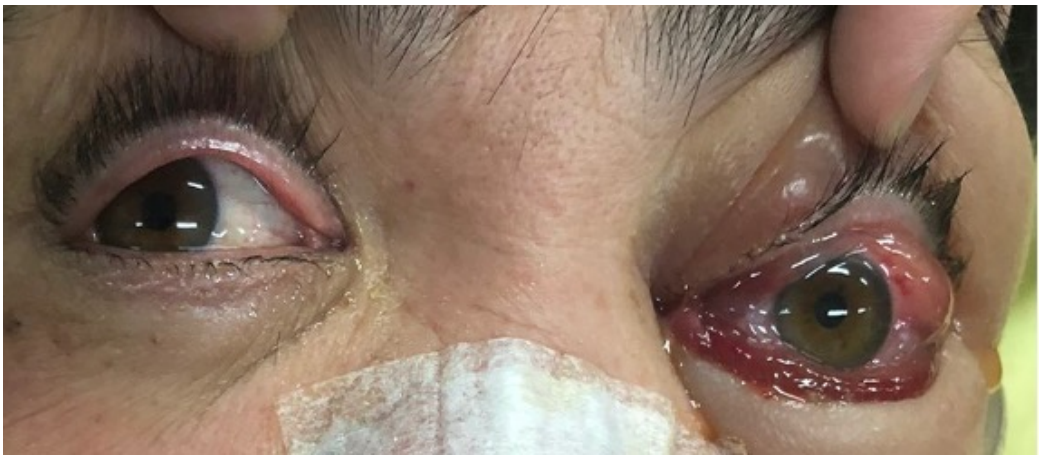


Figure 2. Plain Cranial CT scan. Images A-C shows acute parenchymal hemorrhage in the left frontotemporal lobe amounting to approximately 30 cc with perilesional edema and subarachnoid component in the cortical sulci, left frontotemporoparietal lobe, interhemispheric fissure and left sylvian fissure with compression of the left lateral and 3rd ventricles with rightward midline shift of 0.9 cm. Image D shows exophthalmos of the left orbit.

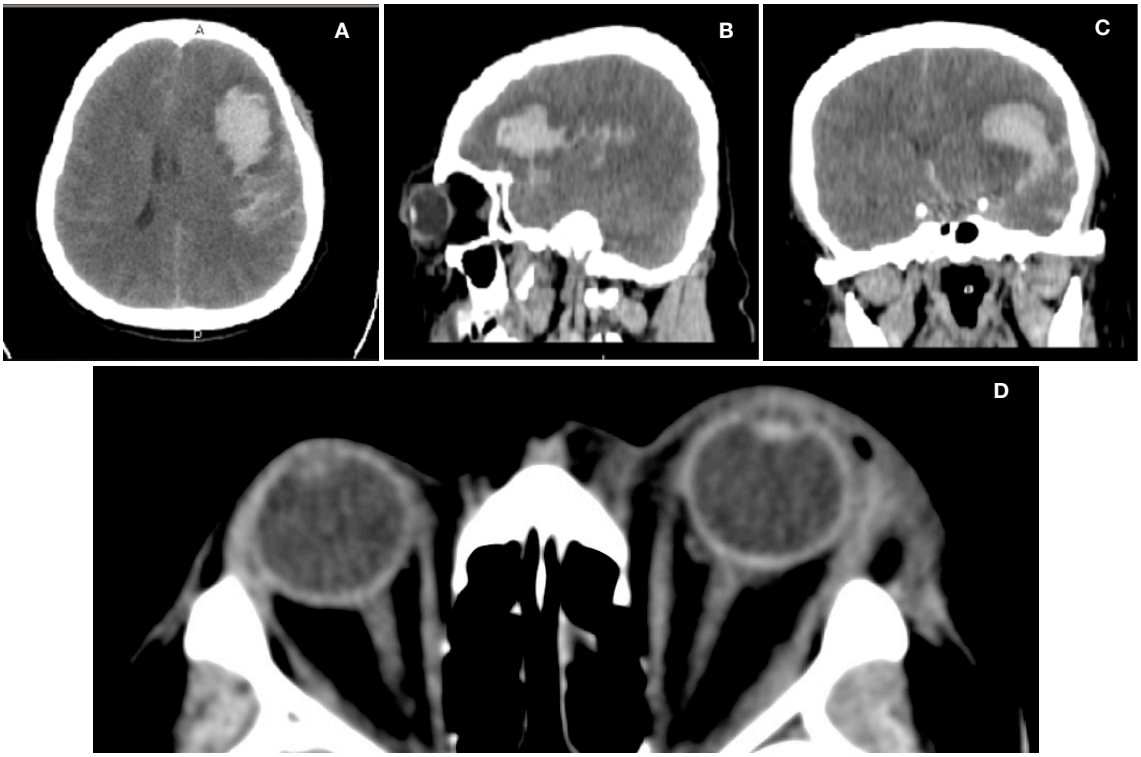
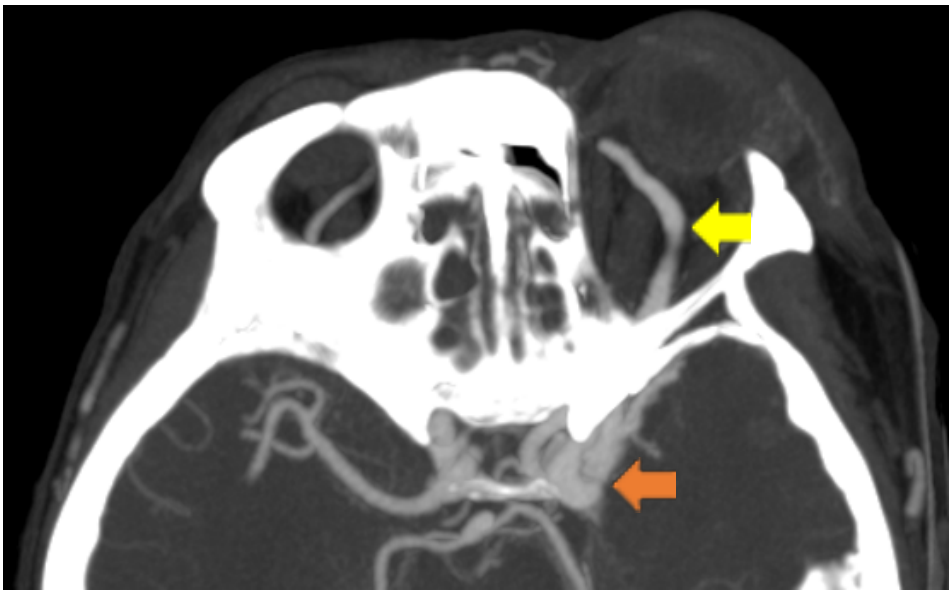


Figure 3. Cranial CT Angiography. The left cavernous sinus is engorged (orange arrow) while the left superior ophthalmic vein is prominent (yellow arrow) with relative exophthalmos of the left orbit.



were done and the patient was referred to another institution for surgical intervention.

The endovascular treatment was done by an Interventional Radiologist last November 5, 2019 at Delos Santos Medical Center. The patient would require 17 coils for the procedure estimated at 1 million pesos, but due to overwhelming financial cost the next best option was to do Internal Carotid Artery Sacrifice with the hope that the patient would not develop post-procedural ischemic stroke. Since the collateral circulation to the left cerebral hemisphere was intact via the patent anterior communicating and left posterior communicating arteries (Figure 4 – C & D), patient was deemed eligible for the procedure, hence Carotid-cavernous fistula closure with Trans-arterial coil (Figure 5 – A) plus Internal carotid artery sacrifice was done. The additional embolization using

Figure 4. Digital Subtraction Angiography. Image A demonstrates a normal blood flow from injection of contrast material from the right internal carotid artery into the right anterior cerebral artery and right middle cerebral artery coursing into their terminal branches. In Image B, the arterial phase of the left internal carotid arteriography shows massive shunting between the cavernous segment of the left internal carotid artery and the cavernous sinus creating a fistulous pouch measuring 20 mm in diameter. This fistula is noted to opacify the enlarged superior ophthalmic vein, superficial middle cerebral vein and the inferior petrosal sinus with drainage to the internal jugular vein. Flow to the left anterior and left middle cerebral arteries is compensated by flow from the right internal carotid artery via the patent anterior communicating artery (Image C) and the right vertebral arteries via the patent left posterior communicating artery after compression of the ipsilateral left common carotid artery (Image D).

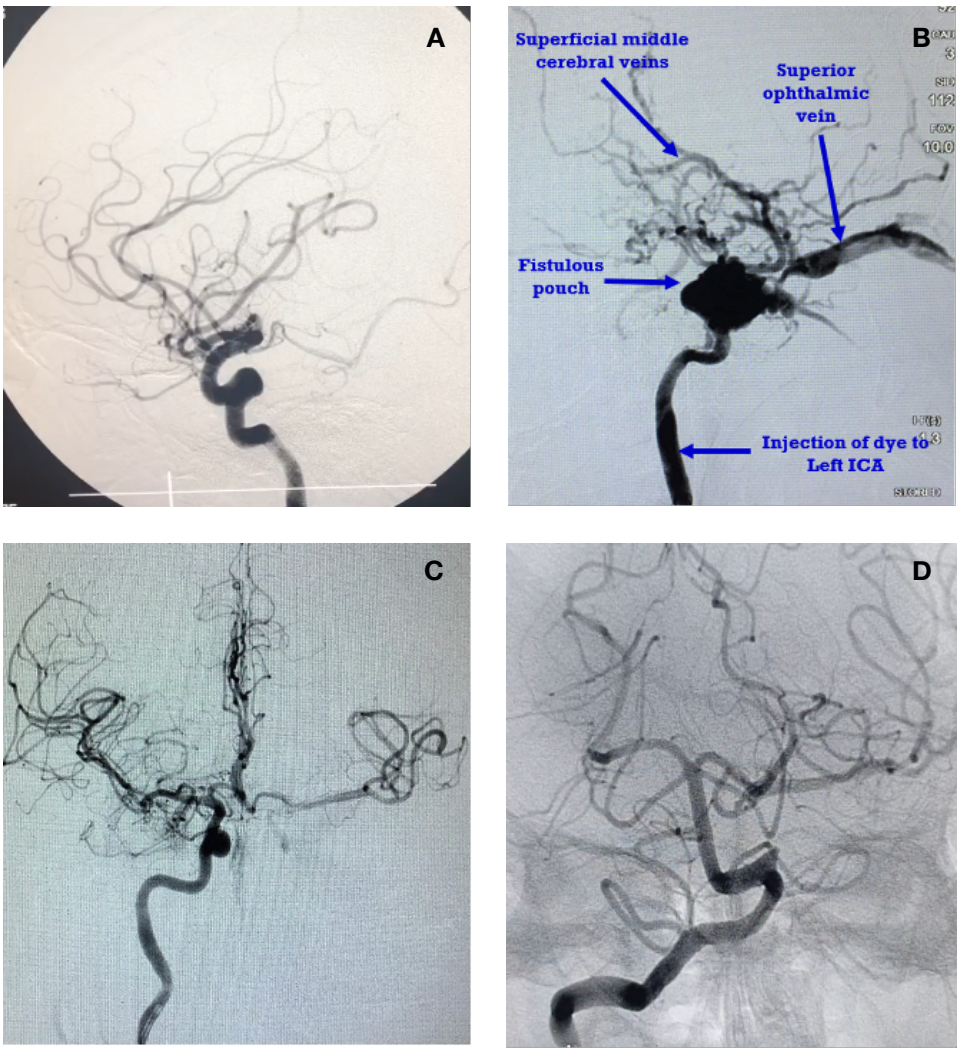
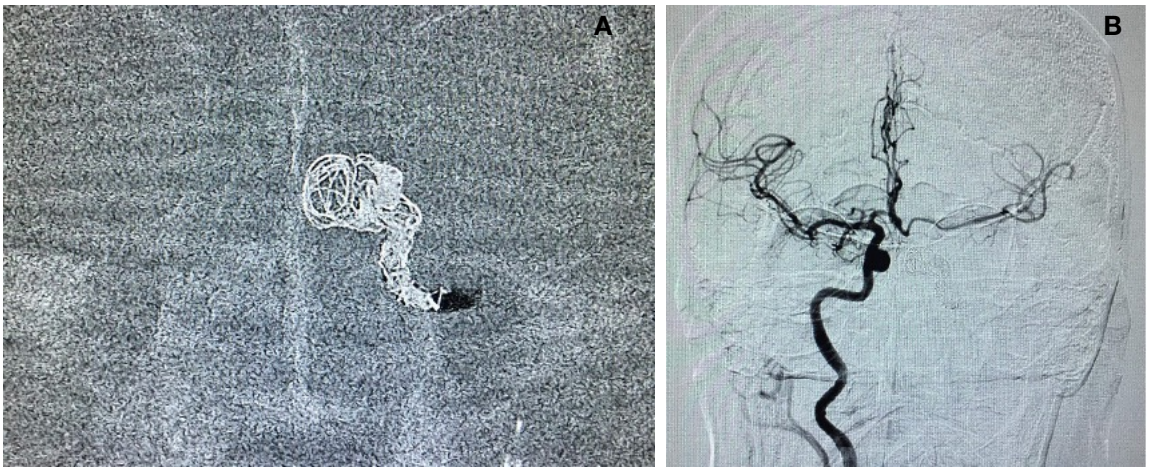


Figure 5. Digital Subtraction Angiography. Image A shows the coil mass placed at the Carotid-cavernous fistulous pouch, post-embolization. Post-embolization, there was intact flow to the left anterior and left middle cerebral arteries via the patent anterior communicating after injection of dye into the right internal carotid artery (Image B).



liquid embolic allowed the use of only 4 coils. Repeat Digital Subtraction Angiography immediately after the embolization still showed an intact collateral circulation (Figure 5 – B). There was noted drastic improvement of the patient's ocular and neurological symptoms from the time of admission until the succeeding weeks (Figure 6).

DISCUSSION

Carotid-cavernous fistulas are acquired vascular lesions representing an abnormal connection between the ICA or ECA and the cavernous sinus. The patient in this case was diagnosed with a direct/Type A CCF consistent with the various clinico-radiographic evidence, but what is unique in this case was the presence of simultaneous ocular/orbital, cavernous and cortical findings in a patient with no history of trauma (causes 70-90% of all direct CCFs). The ocular/orbital signs and symptoms of the patient such as blurring of vision, exophthalmos, subconjunctival hyperemia, scleral edema/chemosis, ocular bruit and retinal venous congestion (AV ratio of 1:3) were the result of increased venous drainage to the left superior ophthalmic vein. Increased pressure in the cavernous sinus itself resulted to various cranial nerve deficits such as impaired direct and consensual reflex (optic and oculomotor nerves), complete ptosis and sluggish corneal reflex (trigeminal nerve) and impaired horizontal and vertical oculocephalic reflex (oculomotor, trochlear and abducens nerves). The ocular/orbital, cavernous and cortical signs and symptoms occurring simultaneously in one patient has never been documented in any literature nor included in any classification system of CCF symptomatology at present. In the retrospective studies done by Thomas et al. (29 patients from 2005 to 2013) and Leone et al. (94 patients from 1994 to 2018), they have classified their patients' clinical presentation under "ocular/orbital only", "cavernous only", "ocular/orbital and cavernous", "cortical" and "asymptomatic", but none of their subjects presented with all three symptomatology [11].

Cortical signs were also the least observed of the symptomatology, with Thomas et al. showing 6.8% (2 out of 29 patients) and Leone et al. showing 8.5% (8 out of 94 patients) in their respective studies [3, 11]. The presence of intracerebral hemorrhage was the cortical component of this CCF, manifested as decreased sensorium and right hemiplegia. The subarachnoid component

from the left frontotemporal hemorrhage accounted for the meningeal signs elicited from the patient. Spontaneous intracranial hemorrhage is exceptionally rare and it is the most daunting symptomatology of CCF resulting from dangerous venous drainages of retrograde cortical venous drainage via the superficial middle cerebral vein and basal cerebral vein (basal vein of Rosenthal)[3, 6, 7, 9].

CONCLUSION

High index of suspicion from the different symptomatology of CCF is important in diagnosing this rare and dangerous disease. The diagnosis and management of CCFs have improved substantially in recent years but this case of a direct CCF with simultaneous orbital/ocular, cavernous and cortical symptomatology may pave way to a new classification scheme and its corresponding treatment approach. The approach done was unconventional to be more cost efficient and effective. Lastly, CCF with intervention can lead to good outcomes.

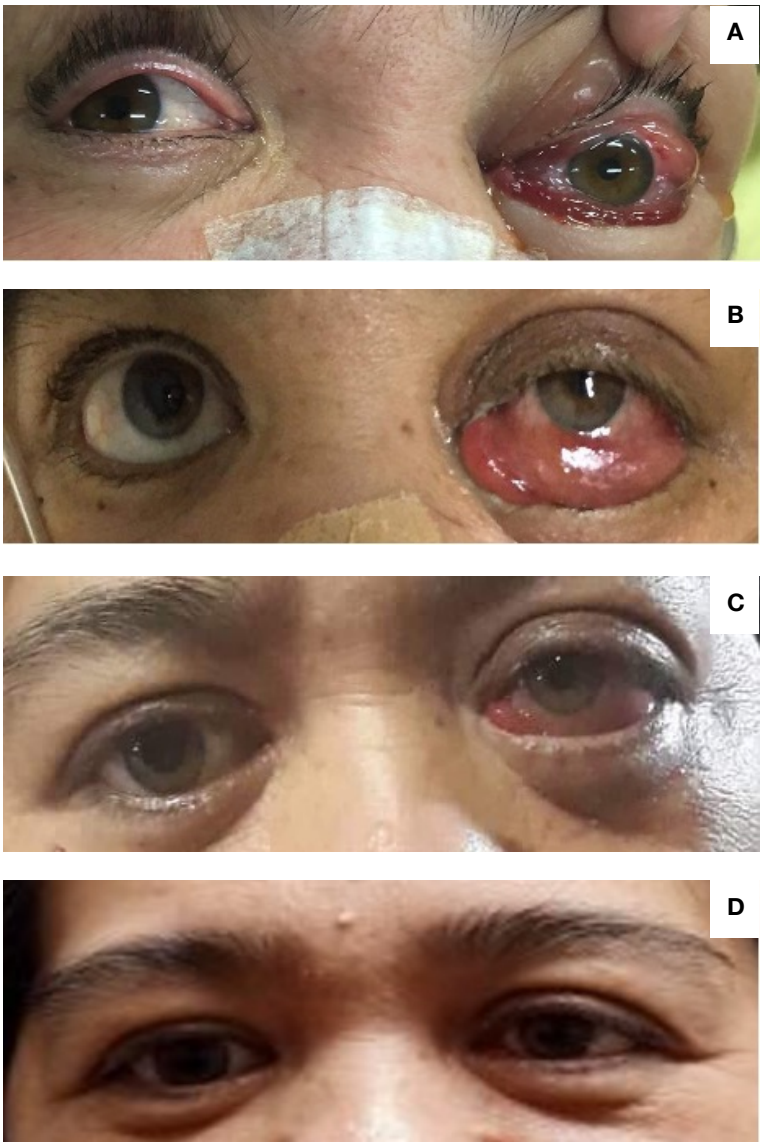


Figure 6. Sequential images showing the patient's clinical improvement from the time of admission until weeks following the CCF closure with trans-arterial coil with liquid embolization + ICA sacrifice.

A. Pre-embolization

B. 3 days post-embolization

C. 2 weeks post-embolisation

D. 8 weeks post-embolization

REFERENCES

1. Parent, A. Carpenter's Human Neuroanatomy, Ninth Edition. Department of Anatomy, Faculty of Medicine, University of Laval, Quebec, Canada. 1996.
2. Mohyuddin A. Indirect carotid cavernous fistula presenting as pulsatile tinnitus. 2000. Department of Otolaryngology, North Manchester General Hospital, Manchester, United Kingdom.
3. Thomas AJ, Chua M, Fusco M, Ogilvy CS, Tubbs RS, Harrigan MR, Griessenauer, CJ. Proposal of Venous Drainage-Based Classification System for Carotid Cavernous Fistulae with Validity Assessment in a Multicenter Cohort. 2015. Congress of Neurological Surgeons. https://doi.org/10.1227/NEU.0000000000000829
4. Chaudhry IA, Elkhamry SM, Al-Rashed, A, Bosley TM. Carotid Cavernous Fistula: Ophthalmological Implications. 2009. Middle East African Journal of Ophthalmology. doi: 10.4103/0974-9233.53862
5. Castro LN, Colorado I, Freitag AB, Rabinov J, Silvermann S. Carotid-Cavernous Fistula: A Rare but Treatable Cause of Rapidly Progressive Vision Loss. 2016. Department of Neurology and Department of Radiology, Massachusetts General Hospital, Boston, USA. DOI: 10.1161/STROKEAHA.116.013428
6. Miller NR. Dural Carotid-Cavernous Fistulas: Epidemiology, Clinical Presentation and Management. 2012. Wilmer Eye Institute, John Hopkins Hospital, USA
7. Griessenauer CJ, He L. Carotid Cavernous and Other Dural Arteriovenous Fistulas. 2017. Primer on Cerebrovascular disease, Second Edition. http://dx.doi.org/10.1016/B978-0-12-803058-5.00098-9
8. Martin S, Teo M, Bhattacharya J, Alakandy L. Carotico-cavernous fistula: An educational case. 2013. Internal Journal of Surgery Case reports. http://dx.doi.org/10.1016/j.ijscr.2013.07.002
9. Kiyosue H, Mori H, Sagara Y, Hori Y, Okahara M, Nagatomi H, Abe, T. Basal cerebral venous drainage from Cavernous Sinus Dural Arteriovenous fistulas. 2008. Department of Radiology, Faculty of Medicine, Oita University, Japan. doi: 10.1007/s00234-008-0486-3
10. Aralasmak A, Karaali K, Cevikol C, Senol U, Sindel T, Torak H, Ozdemir H, Alkan A. Venous Drainage Patterns in Carotid Cavernous Fistulas. 2013. Department of Radiology, Bezmialem Vakif University, Istanbul, Turkey, http://dx.doi.org/10.1155/2014/760267
11. Leone G, Reneiri L, Enriquez-Marulanda A, Dmytriw A, Nappini S, Lasia A, Buono G. Carotid Cavernous Fistulas and Dural Arteriovenous Fistulas of the Cavernous Sinus: Validation of a New Classification According to Venous Drainage. 2019. Department of Neuroradiology, Neurovascular Interventional unit, Careggi University Hospital. https://doi.org/10.1016/j.wneu.2019.04.220