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Case Report

Endovascular Management of Spontaneous Indirect Carotid-Cavernous Fistula

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Abstract:

Carotid cavernous fistulas (CCF) are abnormal communications inducing shunting of blood between the carotid arterial system and the cavernous sinus (CS), resulting in flow reversal in the veins draining through the CS.[1] CCFs can be classified as spontaneous or traumatic according to the etiology and direct or indirect (dural) according to the anatomical features[2]. The authors have dealt a case of a spontaneous indirect Carotid-Cavernous Fistula in a young male of 30 on the right side. The patient presented with headache and bulging of right eye. On examination there was pulsatile proptosis on the right side with conjunctival congestion. CT angiography revealed CCF on the right side with dilated, tortuous right ophthalmic and common facial vein. DSA revealed indirect Carotid-Cavernous Fistula fed by right middle meningeal artery (MMA), accessory meningeal artery and branch of right ophthalmic artery(OA) drained by right ophthalmic vein with abnormal communication via right common facial vein. Patient underwent transarterial embolization with ONYX^R . DSA revealed total obliteration of fistula. Headache subsided and proptosis reduced immediately after embolization on procedure table with intact neurology. Almost two months down the follow up revealed complete recovery of proptosis and conjunctival congestion with no further headache.

Key Words: *Carotid-Cavernous Fistula, Meningeal Artery, Ophthalmic vein Proptosis, Embolization, .ONYX*

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Introduction:

Carotid cavernous fistulas(CCF) are abnormal arteriovenous anastomoses between the internal carotid artery (ICA) and the cavernous sinus (CS). CCFs can be classified as spontaneous or traumatic according to the etiology and directorin direct (dural) according to the anatomical features[2]. Inadirect CCF, arterial blood flows to the CS through the shunt in the intracavernous part of the ICA, causing high blood flow and high pressure. On the other hand, indirect (dural) CCFs result from the connection of the meningeal branches of the ICA with the CS, causinglow bloodflowandlowpressure[2,3]. CCF results in flow reversal in the veins draining through the CS. Flow may be directed anteriorly (ophthalmic venous system), posteriorly(inferior petrosal sinus (IPS), superior petrosal sinus, or the basilar plexus), laterally (sphenoparietal sinus), contralaterally (inter-CS), or inferiorly via the pterygoid plexus through the vein of the foramen

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rotundum and the vein of the foramen ovale. Commonly, a multidirectionalrevision of drainage occurs [1].The main symptoms of CCFs include orbital bruit which can be heard by the patient and objectively determined by the physician, proptosis, chemosis , arterialization of the episcleral veins with the typical corkscrew-like aspect, eyelid edema, diplopia, visual impairment with different degrees and elevated intra ocular pressure , headache ophthalmoplegia, redness of conjunctiva. The treatment modality used depends on classification, angiographic complexity, arterial size, risk of neurological and visual impairment, and operator preferences. Spontaneous closure of indirect CCF occurs in 20–60% of cases[4]. Indications for endovascular treatment of low-flow indirect fistulas include decreasing visual acuity or field,optic disc swelling, ocular hypertension, proliferative retinopathy,headaches, cortical drainage, intraparenchymalhaemorrhage or cosmetic deformity[5,6]. In contrast , direct CCFs should almost always be closed as spontaneous closure is unlikely and ocular and neurologic compromise is common [7]. Various methods have been described for the treatment of indirect CCF including transarterial and transvenous embolization. Transarterial feeder vessel embolization may be performedbut is frequently inadequate for curative treatment. Thustransvenous embolization is generally considered to be a more effective treatment for indirect CCF. ONYX is a new liquid embolic agent that has been found to be useful in treating CCF . However, the use of Onyx in a transvenous approach has limitations. Injecting Onyx against the flow of the fistula is technically challenging; the tendency for the compound to follow the direction of flow increases the risk of embolization of a normal draining vein and migration to an unwanted site.

Case Report:

A 30 year old male presented with headache and bulging of right eye for 2 months. There was no history of trauma. On examination there was pulsatile proptosis on the right side with conjunctival congestion (Fig. 1.A,B,C). CT angiography revealed CCF on the right side with dilated, tortuous right ophthalmic and common facial vein (Fig.2 A,B,C). DSA revealed indirect Carotid-Cavernous Fistula (Barrow type D) fed by right middle meningeal artery (MMA), accessory meningeal artery and branch of right ophthalmic artery (OA) drained by right ophthalmic vein with abnormal communication via right common facial vein(Fig. 3 A,B,C)

Transarterial embolization of fistula with ONYX was performed . In this endovascular procedure guiding

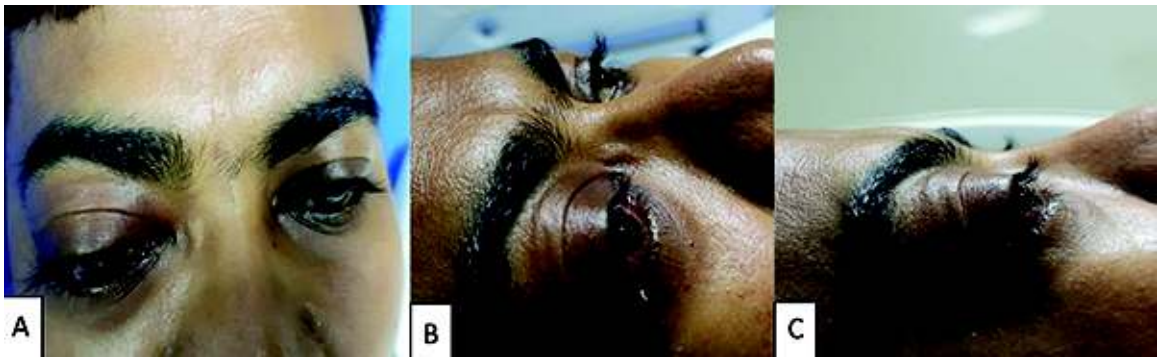

Fig.-1: *Proptosis of right eye with conjunctival congestion (A+B+C)*

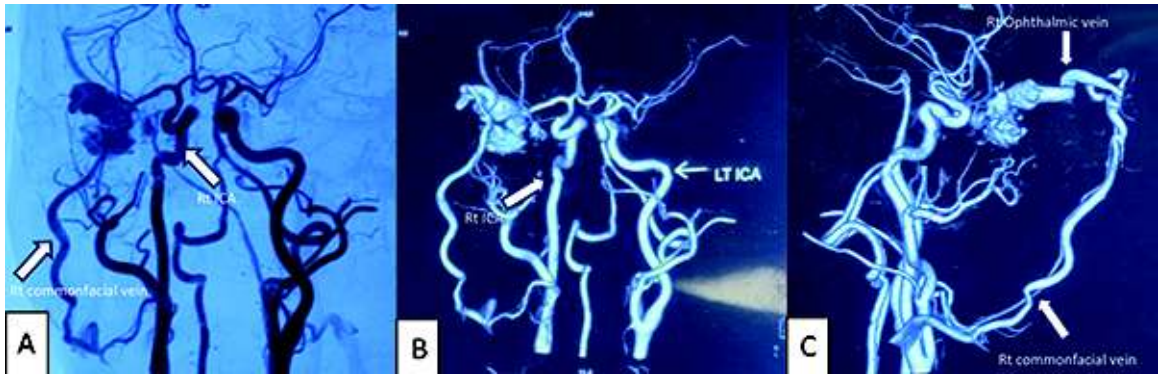

Fig.-2: *CTA reveals right sided CCF with dilated tortuous ophthalmic and common facial vein (A+B+C)*

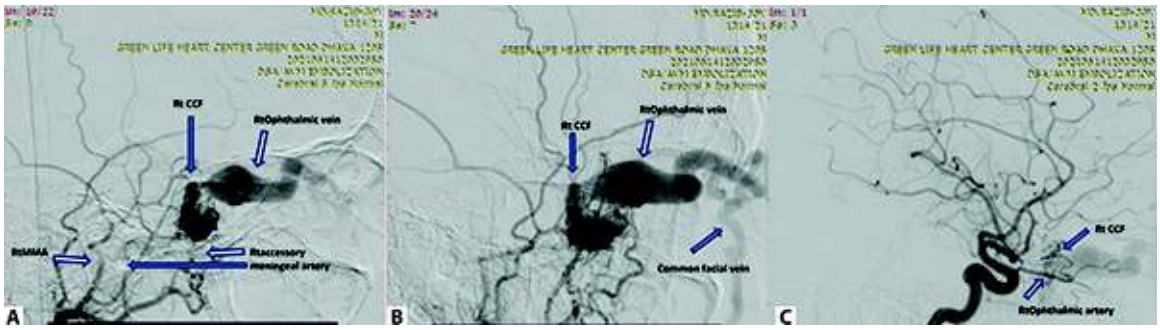

Fig.-3: *DSA reveals right sided indirect CCF (Barrow type D) (A+B+C), fed by right MMA (A) ,accessory meningeal artery (A) and branch of right OA (C), drained by right Ophthalmic vein (A+B) with abnormal communication with right common facial vein (B)*

catheter 6F Envoy was navigated till the bifurcation of ECA. Microcatheter Marathon^{TM} .013 in. over microwire CHIKAI^{TM}.010 in was navigated through accessory meningeal artery (Fig. 4).

ONYX^{TM} was injected slowly and waited until reflux stops and forward flow establishes and pressure cooker effect takes place (Fig.5)

Then ONYX was injected further and embolization of fistula along with mentioned feeders and proximal ophthalmic vein was done (Fig.6)

Post procedure DSA revealed complete embolization of right sided indirect CCF and patency of normal vasculature (Fig.7)

Headache subsided and proptosis reduced immediately after embolization on procedure table with intact neurology (Fig. 8).

Almost two months down the follow up revealed complete recovery of proptosis and conjunctival congestion with no further headache.

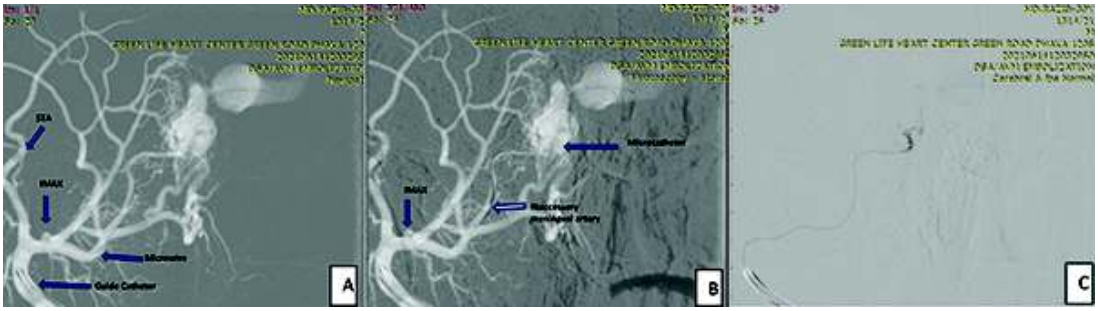

Fig.-4: *Road map shows Guide catheter Envoy at the bifurcation of ECA (A+B) , microcatheter Marathon^{TM} .013 in (B) over microwire CHIKAI^{TM} .010 in (A+B) all the upto fistulous point ;visualization of fistula by shot through microcatheter (C) just before embolization.*

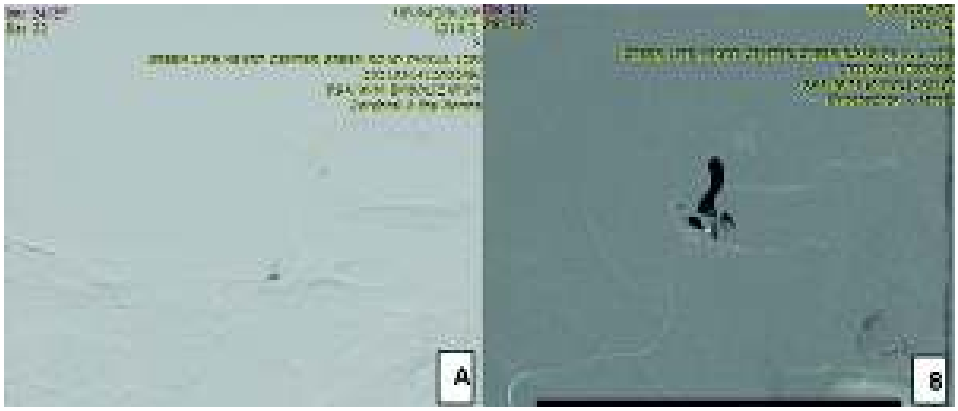

Fig.-5: *Injection of ONYX^{TM} with reflux (A); forward flow of ONYX^{TM}and establishment of pressure cooker effect (B)*

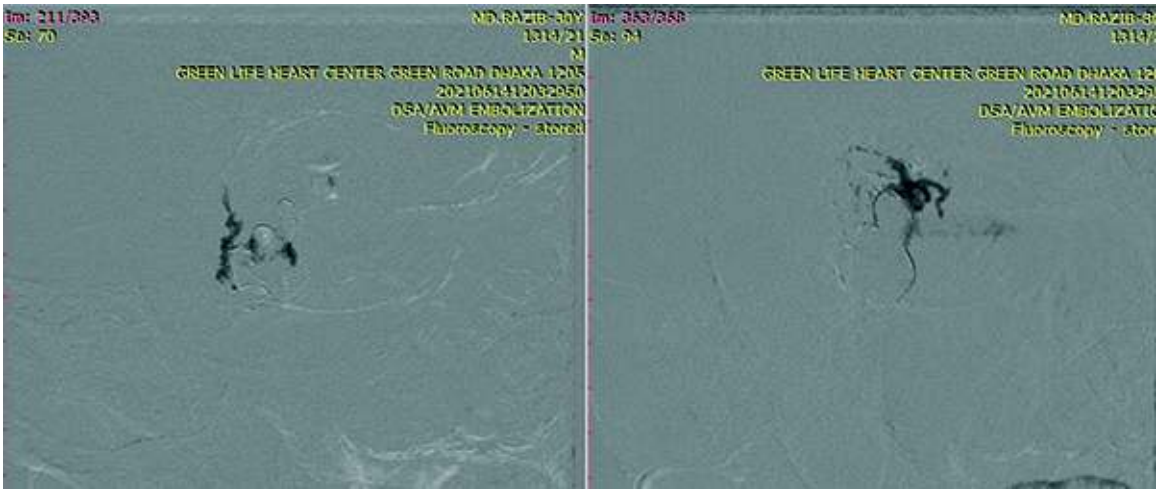

Fig.-6: *Forward flow of ONYX^{TM} and complete embolization of CCF (A+B).*

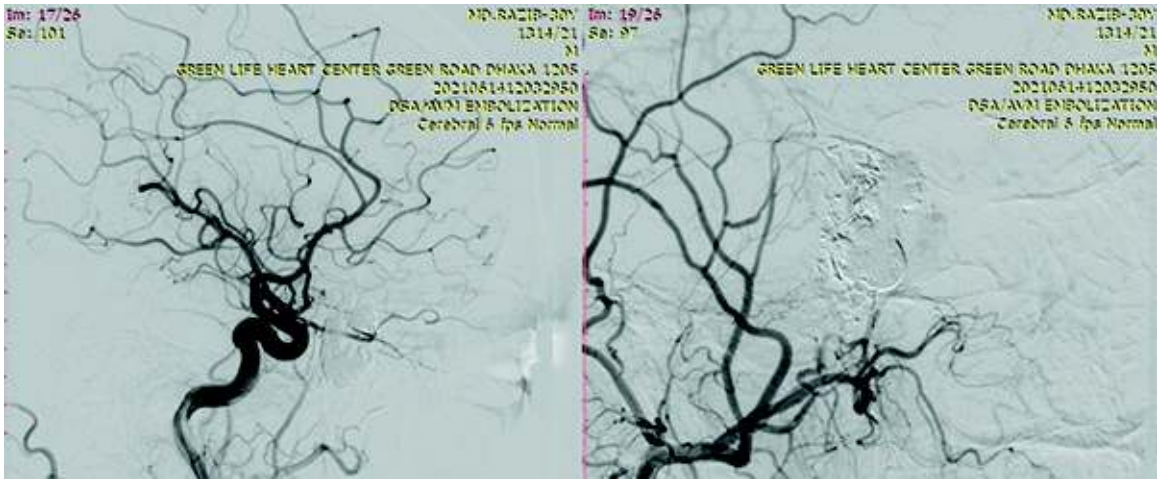

Fig.-7: *Post embolization DSA revealed complete embolization of CCF with normal vasculature (A+B).*

Fig.-8: *Post procedure clinical status -reduction of proptosis (A+B)*

Discussion

CCF accounts for 20% to 40% of all intracranial DAVFs[8]. CCFs may develop spontaneously or secondary to trauma, ruptured cavernous carotid aneurysm, venous thrombosis, or genetically weakened arteries. No cause is identified for the majority of non-traumatic CCFs; however, several predisposing factors have been identified including sinus thrombosis, atherosclerosis, hypertension,

diabetes, phlebitis, sinusitis, and pregnancy[1,9,10,11].

High-flow CCF classically presents with a triad of subjective bruit (which may also be heard on auscultation of the globe), proptosis, and conjunctival chemosis[9]. Other symptoms include blurred vision, headache, diplopia, and ocular/orbital pain all of which were frequently observed in our series[4,12,13,14,15,16].

Indirect, low-flow fistulas oftenpresent with insidious and progressive proptosis, conjunctival injection and raised IOP[9,17,18].

Aggressive neurological symptoms are rare due to the benign venous drainage pattern, but can occur in association with dangerous venous drainage patterns, including cortical venous reflux (hemorrhagic infarction), deep venous drainage (hemorrhage, edema), and thrombosis of the central retinal vein (blindness) Ophthalmic complications secondary to CCFs occur as a result of congestion from impaired venous drainage of the orbit to the cavernous sinus. Clinical presentation is variable due to differences in fistula size, location, rate of flow and location in the cavernous sinus [19].

CT and MRI are often employed for initial diagnostic imaging. Previously reported CT findings include proptosis, and enlargement of the ipsilateral CS, SOV, and extraocular muscles (EOMs). In addition to the above

findings, MRI demonstrates abnormal flow voids in the CS and orbital oedema[9]. In our study, CT, CTA, and MRI were all observed as initial medical imaging modalities followed by DSA. DSA remains the gold standard for the definitive diagnosis and classification of CCFs and is recommended for the planning of endovascular treatment. Indirect fistulas are most commonly supplied by branches of the ECA including the internal maxillary, middle meningeal, accessory meningeal and ascending pharyngeal branches, and less commonly by cavernous segment branches of the ICA[18]. Previous reports identify type D DAVFs as the most common indirect CCF,followed by type C and B[20]. In our case , Barrowtype D fistula was encountered.

The goal of treatment for indirect fistulas is to reduce CS pressure by interrupting the fistulous communications without redirecting flow to cortical

structures [1,9,17]. Indirect fistulas often involve multiplebranches of the meningeal arteries which can be difficultto treat with endovascular techniques [6]. However Indirect CCF can be treated using a transarterial or a transvenous approach. The transvenous approach is most commonly used for indirect fistula because the transarterial approach is a more complex and less effective technique. Thecombination of transarterial treatment with embolicagents such as Onyx has resulted in higher successrates[6]. Unlike direct CCFs, which are rarely asymptomatic and are treated urgently,indirect CCFs may resolve spontaneously due to thrombosis in up to 60% of cases[2,19]. Over all, conventional endovascular therapy remains the mainstay of treatment.

A wide variety of embolic agents are available for indirect CCF embolization, including polyvinyl alcohol particles, coils, N-butyl cyanoacrylate (NBCA), and DMSO solvent materials such as Onyx, Squid, and precipitating hydrophobic injectable liquid. Of those, coils and DMSO solvent materials such as Onyx are the most commonly used agents, with each having advantages and disadvantages.

Coils are placed in a controlled manner and are more easily deployed in the desired position than is Onyx. However, coils are associated with a lower rate of complete obliteration. Moreover, coils are more thrombogenic than Onyx and can lead to progressive thrombosis causing mass effects.

Onyx embolization is less controlled than coil placement, however, this material is associated with a higher rate of complete obliteration [21]. Furthermore, because Onyx is less thrombogenic than coils, paradoxical worsening occurs less frequently after Onyx embolization. Nevertheless, paradoxical worsening can occur after Onyx embolization [22], possibly caused by CS thrombosis and swelling, or due to the angiotoxic effect of DMSO [23].

In this case author has used ONYX because it is less thrombogenic and has a higher rate of complete obliteration. However when using Onyx, it is essential

to allow time for the material to create a plug around the tip of the microcatheter, which enables the forward flow of Onyx. Author report the successful endovascular transarterial embolization of indirect CCF with ONYX. No unintended ONYX migration or procedure related complication was observed and fistula was completely occluded.

Conclusion:

Carotid-cavernous fistulaare abnormal communication between the carotid artery and cavernous sinus[2]. There are different options of management including conservative, surgical and endovascular depending on presentation, severity and types- direct or indirect. Recently author has dealt with a case of indirect CCF in a 30 year old male who presented with spontaneous onset of headache, proptosis and redness in right eye. DSA revealed indirect Carotid-cavernous fistula (Barrow type D). It was successfully treated with transarterial embolization with ONYX.

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