

# Atypical Presentation of Direct Carotid Cavernous Fistula

Fizzah Farooq<sup>1</sup>, Mehvash Hussain<sup>2</sup>, Muhammad Munir Quraishy<sup>3</sup>  
<sup>1-3</sup>Dow University of Health sciences and civil Hospital, Karachi

## ABSTRACT

An atypical case of carotid cavernous fistula (CCF) is presented which was challenging to diagnose even with investigations. A 50-year old female, presented with complaints of increasing chemosis, diplopia and proptosis of her right eye following a fall from stairs. Her IOP was 32 mm Hg in right and 18 in left eye. Absence of other classic clinical signs like bruit/thrill of carotid cavernous fistula led to difficulty in diagnosing. There was absence of radiological evidence like enlargement of superior ophthalmic vein on MRI, MRV and MRA. We started symptomatic treatment for increased IOP and exposure keratopathy. Right sided CCF diagnosis was confirmed on CT angiogram and Digital Subtraction Angiography. Coil embolization was done which resulted in symptomatic and clinical relief. Carotid cavernous fistula with absence of typical signs is sometimes challenging to diagnose and treat. Importance of Digital Subtraction Study and CT Angiogram is highlighted.

**Key Words:** Carotid cavernous fistula, CT angiogram, Digital Subtraction Angiography.

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*Correspondence:* Fizzah Farooq  
Down University of Health sciences and civil Hospital,  
Karachi  
Email: Fizzah.farooq@hotmail.com

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## INTRODUCTION

Carotid cavernous fistula is a rare complication, which occurs due to abnormal communication between carotid artery and cavernous fistula.<sup>1</sup> Anatomically, it is categorized into direct and indirect. The cause of fistula can be iatrogenic, spontaneous or secondary, for example trauma.<sup>2</sup> It is a rare sight threatening complication of trauma. This case is an atypical presentation of direct carotid cavernous fistula following history of fall. The unusual presentation of this case will assist in eliminating confusion regarding CCF and will help in early diagnosis and prompt treatment since such cases are an ocular emergency.

## Case Presentation

A 50 year old housewife and a known case of

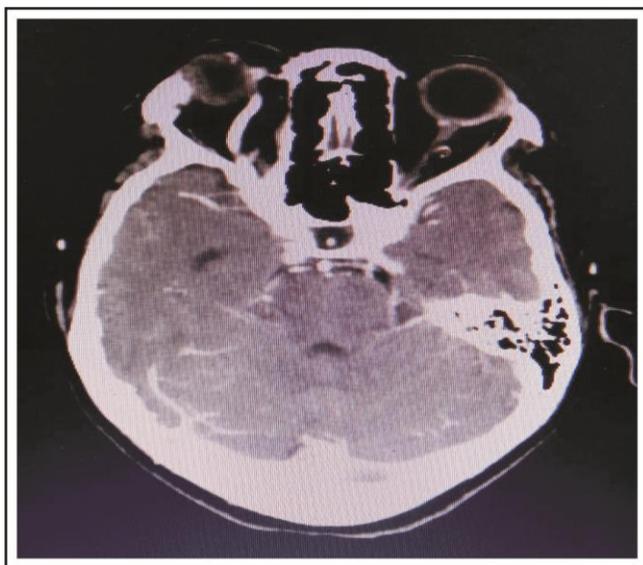
hypertension, reported to our outpatient department with complaints of gradually increasing diplopia, proptosis and chemosis in her right eye after falling from stairs in her house 25 days back. She also had complaints of blurring of vision, which was sudden and painless and uniform over day and night. Twenty five days back she slipped from stairs at her home which was followed by nasal bleed and altered level of consciousness. She was immediately taken to trauma center, on her way she regained her consciousness but had multiple episodes of non-projectile vomiting. On presentation in the emergency, her vital signs were normal. Her GCS was 15/15. She was given symptomatic treatment and CT brain was advised. CT scan showed frontal intra cranial bleed for which she was kept under observation for 2 – 3 hours. After 2 hours her CT was repeated, which showed resolved bleeding so she was discharged. In our department, her best corrected visual acuity was 6/12 in right and 6/6 in left eye. There was binocular diplopia in all gazes with right lateral gaze restriction. Pupillary light reflexes, cover/uncover and colour vision were normal. On anterior segment examination right eye was congested (Figure 1) with intra ocular pressure (IOP) of 32 mm Hg. Fundus was unremarkable with

normal color, contour and ratio of disc ratio. Macular reflex was good. Bruit, whooshing sound and pulsation were absent over the eyes.



**Figure 1:** Left: Congestion at time of presentation. Right: Corneal ulceration after few days.

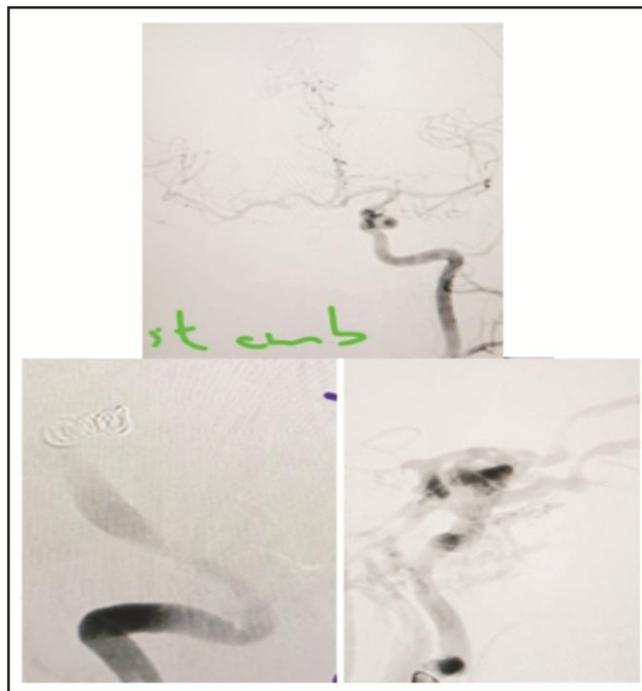
Patient was given symptomatic treatment to lower IOP and was advised CT brain and MRI/MRV/MRA to confirm diagnosis. All these investigations were normal. Despite treatment, chemosis and proptosis rapidly kept on increasing and led to exposure keratopathy followed by corneal ulceration and opacification. CT angiogram was suggested which showed early opacification of bilateral cavernous sinus and prominent superior ophthalmic vein on the right side (Figure 2).



**Figure 2:** CT angiogram showing dilated superior ophthalmic vein.

Digital Subtraction Angiogram Study was advised and patient was referred to interventional radiologist. Endo vascular approach was done and patient underwent embolization with coils post arterial

catheterization (Figure 3). Immediately after coiling, her symptoms and signs subsided but on subsequent follow up she developed neovascularization of cornea/iris which was found to be an inevitable complication of retinal hypoxia due to coil embolization as confirmed by interventional radiologist (Figure 4). We started her on topical Bevacizumab drops and within a month her neo vessels regressed.



**Figure 3:** Trans arterial coil embolization.

At her last follow up, her vision was improved with no diplopia and full extraocular movements. Corneal scar had shrunk to 2 clock hours with no neovascularisation. She had developed cataract. Our future plan is to do cataract surgery to improve vision (Figure 4).



**Figure 4:** Left image showing neo vessels, Right image shows regressed vessels at her last follow up.

## DISCUSSION

CCF is a very rare complication of trauma.<sup>1</sup> Although limited data is available regarding its incidence however, according to one report, it is 3.8% in basal skull fractures.<sup>2</sup> CCF is classified according to causes: spontaneous, iatrogenic or secondary like trauma due to penetrating injuries or road traffic accidents.<sup>3</sup> Thermodynamically it is classified into high flow or low flow. While anatomically it is divided into direct and indirect. In direct there is a direct communication between ICA and cavernous sinus. In indirect there is abnormal communication between meningeal branches and sinus.<sup>4</sup>

Vision loss is one of the most dreadful complications of CCF. Sight threatening complications may occur in direct CCF due to severe exposure keratopathy, corneal ulcerations, and possibly central retinal artery occlusion.<sup>5</sup> All these complications were seen in our case except central retinal artery occlusion which resulted in improvement in visual acuity after treatment. Up to 76% of direct CCF are traumatic.<sup>6</sup> Fall accounts for 10% of cause of direct CCF cases, which was the reason in our case too.

The differential diagnoses of CCF are vascular malformations of eyes, cerebral aneurysms, retro-orbital cellulitis, inflammation of orbits, retrobulbar hemorrhage, thyroid ophthalmopathy, lacrimal gland tumours, cavernous sinus thrombosis, and vasculitis.<sup>7</sup> We had excluded all these differentials before making our final diagnosis.

Direct CCF has presentation of proptosis (72%), chemosis (55%), bruit (80%), lateral gaze palsy (49%) and conjunctival injection (44%).<sup>8</sup> Bruit was absent in our case. Diagnosis of CCF is mostly based on clinical symptoms and history. Definite diagnosis is based on angiographic study, which aids in classification of case and treatment protocol.<sup>9</sup> Small, spontaneous and asymptomatic fistula can be treated conservatively. However, treatment of choice for direct symptomatic fistula is trans-arterial embolization with detachable balloon. Balloons have also been substituted with coils made of platinum, used alone or in conjunction with balloon.<sup>10</sup>

## CONCLUSION

Early diagnosis and prompt treatment can save a patient from vision threatening complications of CCF as is depicted from our case. Advancement of interventional procedures like trans-arterial stents have

shown tremendous improvement in redirecting blood flow to normal anatomical pathway.

**Conflict of Interest:** Authors declared no conflict of interest.

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### **Authors' Designation and Contribution**

Fizzah Farooq; Medical Officer: *Concepts, Design, Literature search, Data acquisition, Manuscript preparation, Manuscript editing.*

Mehvash Hussain; Assistant Professor: *Literature search, Manuscript preparation, Manuscript editing, Manuscript review.*

Muhammad Munir Quraishy; Professor: *Manuscript editing, Manuscript review.*

