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Case Series

Ophthalmology Section

Unmasking Hidden Vascular Threats through the Eye: A Case Series of Carotico-Cavernous Fistulas and Orbital Arteriovenous Malformations

SHIVANAND C BUBANALE[1], SHUBHRA BHARGAVA[2], NAVIN MULIMANI[3], BHAGYAJYOTHI KHANAGAVI[4], NIVEDITA S NANDA[5]

ABSTRACT

Carotico-Cavernous Fistula (CCF) and orbital Arteriovenous Malformation (AVM) are rare but vision-threatening vascular disorders that often present initially with ocular signs. Early recognition by ophthalmologists is critical to prevent irreversible visual morbidity. This case series describes the clinical spectrum, imaging features, and outcomes of four patients with orbital vascular anomalies presenting primarily with ophthalmic manifestations. All patients presented with varying combinations of proptosis, conjunctival congestion, chemosis, diplopia, headache, and a history of trauma. Comprehensive ophthalmic evaluation was supplemented with contrast-enhanced Computed Tomography (CT) and Magnetic Resonance Imaging (MRI), followed by Digital Subtraction Angiography (DSA) for definitive diagnosis and classification. Three patients were diagnosed with traumatic CCFs, including one direct high-flow and two indirect low-flow variants, while one patient had a rare high-flow infraorbital AVM with orbital extension. DSA played a pivotal role in delineating angioarchitecture and guiding management. All CCF cases were successfully treated with endovascular embolisation, resulting in rapid improvement of proptosis, resolution of conjunctival congestion, and symptomatic relief. The AVM case required a combined approach of embolisation followed by surgical excision, achieving complete resolution without recurrence. This case series uniquely highlights how subtle ophthalmic signs can be the first indicators of serious intracranial vascular pathology. Early ophthalmic suspicion, timely imaging, and multidisciplinary intervention are essential for optimal visual and anatomical outcomes.

Keywords: Digital subtraction angiography, Embolisation, Ophthalmic signs, Orbital diseases, Proptosis, Radiological correlation, Therapeutic

INTRODUCTION

The CCF is a rare but serious condition involving an abnormal connection between the carotid artery and the cavernous sinus located behind the eye [1]. This vascular anomaly can have profound ophthalmological and neurological symptoms due to altered haemodynamics, leading to venous congestion, raised Intraocular Pressure (IOP) and ocular ischaemia that lead to misdiagnosis and delayed treatment [2]. Direct (high-flow) CCFs are usually post-traumatic, occur predominantly in younger males, and present acutely with pulsatile proptosis, marked chemosis, bruit, raised IOP, and rapid visual compromise. Indirect (low-flow) CCFs are typically spontaneous, more common in elderly patients with a female predominance, and manifest insidiously with chronic red eye, mild proptosis, diplopia, or ocular discomfort, often leading to delayed diagnosis [3].

The AVMs are rare vascular lesions characterised by direct connections between arteries and veins, bypassing capillaries. While most AVMs occur within the skull, extracranial AVMs are less common. These malformations tend to persist or worsen over time and can be triggered by hormonal changes or trauma [4].

Diagnosing and managing CCF and AVMs requires a multidisciplinary approach that combines ophthalmological examination and radiological imaging. This article explores the correlation between ophthalmological findings and radiological investigations in CCF and AVM, emphasising their significance for early detection, classification, and treatment planning [5].

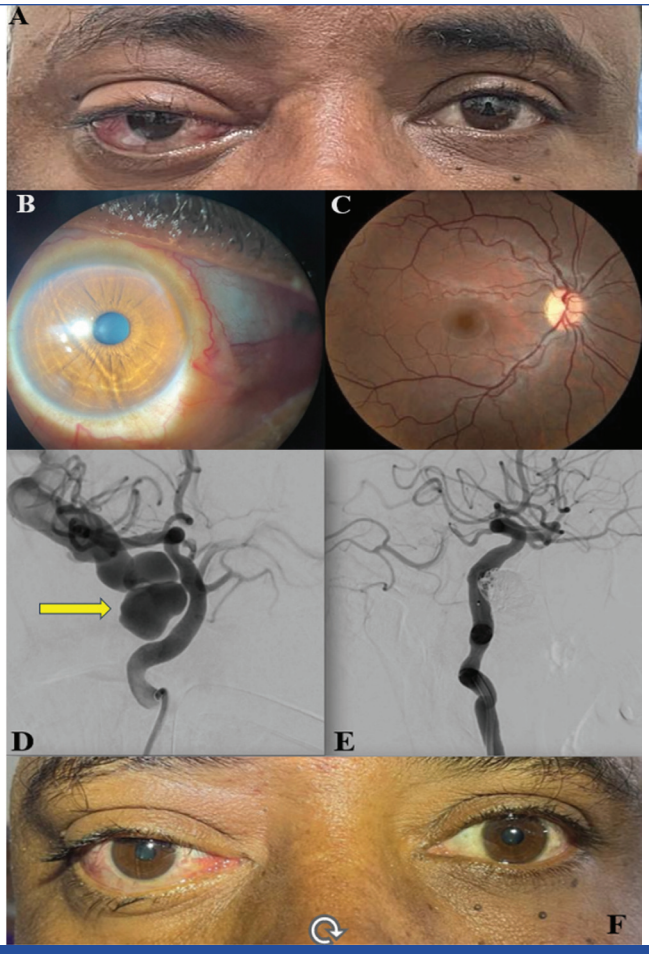
CASE SERIES

Case 1

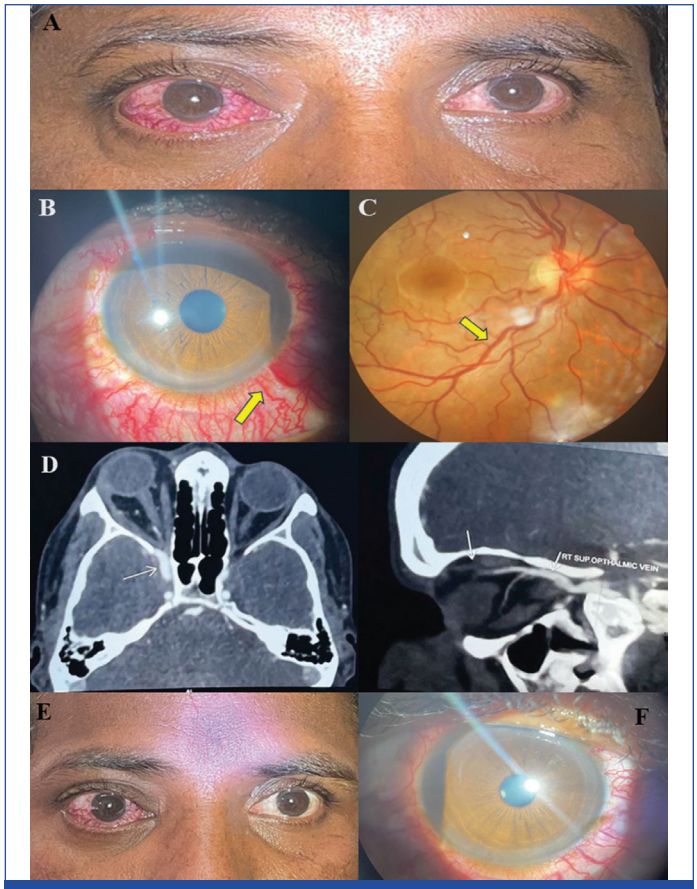
A 41-year-old male presented with a history of progressive right eye redness, watering, dull aching, persistent pain and proptosis for one and a half years following a road traffic accident two years ago, with no previous medical history. The accident caused fractures of the right temporal bone, nasal bone, and sphenoid sinus lateral wall. The Best Corrected Visual Acuity (BCVA) was 6/18 (OD) and 6/6 (OS). OD showed conjunctival congestion, chemosis, proptosis (OD: 22 mm, OS: 10 mm), increased vertical palpebral fissure height (OD: 17 mm, OS:12 mm) and elevated IOP (OD-21 mmHg, OS: 17.9 mmHg), as shown in [Table/Fig-1a,b]. Fundus examination revealed dilated, tortuous retinal vessels bilaterally, and a bruit was audible over the right superior orbital margin [Table/Fig-1c]. Cerebral angiography confirmed a Type A direct high-flow Carotid-Cavernous Fistula (CCF) from the right cavernous ICA-posterior genu [Table/Fig-1d,e]. Following endovascular embolisation, symptoms improved, BCVA improved to 6/9 (p) (OD) and 6/6 (OS). Post-treatment IOP was 18.6 mmHg (OD) and 19 mmHg (OS), with a reduction in proptosis to 19 mm (OD). No complications were noted. The patient did not return for long-term ophthalmic follow-up [Table/Fig-1f].

Case 2

A 32-year-old male who presented with a six-month history of headache and redness in the right eye, with a history of a road traffic accident two years earlier, causing limb injuries and head trauma. He was a known case of hypertension for two months. He went to the primary health care centre for the above complaints and was started on topical medications that were ineffective. Ocular examination showed visual acuity of 6/6 in both eyes, IOPs of 11 mmHg (OD) and 10.5 mmHg (OS) and proptosis of 24 mm (OD) and 20 mm (OS). OD had dilated superficial and deep corkscrew vessels on anterior segment examination and fundus examination revealed dilated, tortuous vessels; however left eye was within normal limits, as depicted in [Table/Fig-2a-c]. Contrast-enhanced CT of the brain



[Table/Fig-1]: Ocular and radiological findings of case 1: a) Redness and chemosis in the right eye; b) Anterior segment of right eye depicting corkscrew vessel on slit lamp examination; c) Fundus diagram showing dilated and tortuous vessels in right eye; d) Cerebral angiogram shows rent at right cavernous ICA-posterior genu communicating directly with cavernous sinus causing moderate flow fistulous communication and reflux into superior ophthalmic vein S/o type a direct Carotico-Cavernous Fistula (CCF) on the right-side (marked with yellow arrow) e) Post-coiling picture of the CCF; f) Anterior segment picture post-coiling.



[Table/Fig-2]: Ocular and radiological findings of case 2: a) Redness in the right eye; b) Anterior segment of right eye depicting corkscrew vessel on slit lamp examination; c) Fundus diagram showing dilated and tortuous vessels in right eye; d) Contrast-enhanced CT showing bulging of the right cavernous sinus and hypodense defect seen in terminal portion of superior ophthalmic vein noted along with right-sided ocular proptosis seen with mild periorbital oedema S/o indirect Carotico-Cavernous Fistula (CCF) with superior ophthalmic vein thrombosis in its terminal portion on the right-side; e) Reduction in exophthalmometry noted post endovascular coiling; f) Anterior segment picture post endovascular coiling.

suggested a right-sided Type D Indirect low-flow CCF, which was subsequently confirmed on DSA [Table/Fig-2d]. Post-endovascular embolisation, BCVA remained 6/6 in both eyes, with IOP values of 14.9 mmHg (OD), 8.5 mmHg (OS) and mild reduction in proptosis (23 mm OD) as shown in [Table/Fig-2e,f]. No complications were encountered. The patient was lost to ophthalmic follow-up.

Case 3

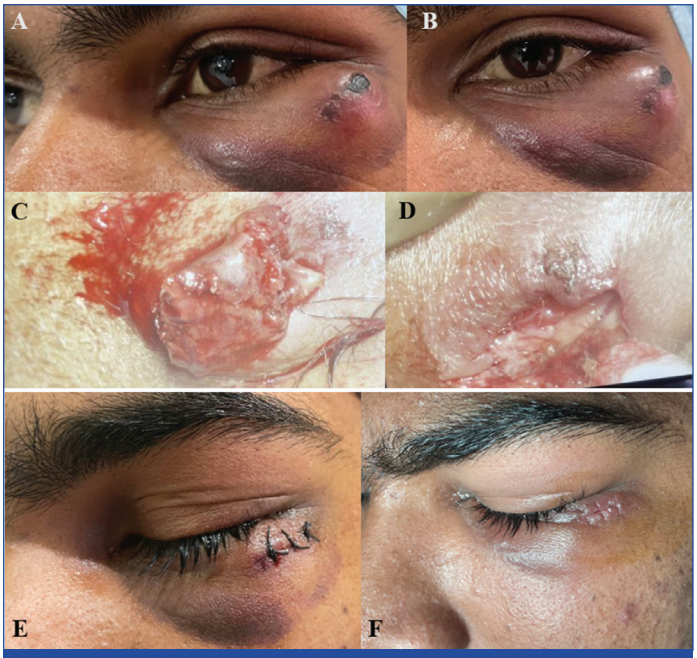
A 45-year-old female presented with six months of headache and redness in the left eye, with a history of head trauma from a road traffic accident five years earlier, with no previous medical history. Initially, she was treated as a case of allergic conjunctivitis for four months, but her symptoms did not improve. Ocular examination revealed BCVA of 6/9 (OD) and 6/12 (OS), IOPs of 13 mmHg (OD) and 12 mmHg (OS), and left eye proptosis of 25 mm and 20 mm in the right eye. The left eye had dilated corkscrew conjunctival vessels, venous engorgement, and mild disc hyperaemia. Contrast-enhanced CT and DSA confirmed a left-sided Type B indirect low-flow CCF. She underwent successful transvenous embolisation, with post-operative BCVA improvement to 6/6 (OD) and 6/9 (OS), with stable IOP values (OD: 10.5 mmHg, OS: 16 mmHg) and reduction in proptosis to 23 mm (OS). No complications were observed. Long-term follow-up was unavailable.

Case 4

A 23-year-old male presented with a one-month history of persistent, painless swelling in the left infraorbital region following blunt facial trauma from a fall, with no known medical comorbidities. There was no redness, discharge, or visual disturbance. Ocular examination was unremarkable, with 6/6 vision, IOP values of 15 mmHg (OD), 9 mmHg (OS) and normal anterior and posterior segments in both eyes. Contrast-enhanced MRI and colour Doppler USG revealed abnormal vascular channels, and DSA confirmed a high-flow left infraorbital AVM. The patient underwent combined endovascular and percutaneous embolisation, following which BCVA remained 6/6 in both eyes and IOP was 13 mmHg (OD) and 10 mmHg (OS). It resulted in incomplete regression of the swelling as shown in [Table/Fig-3a,b]. Over time, the swelling progressively enlarged into a lobulated mass, prompting the patient to seek ophthalmic evaluation. Surgical excision under local anaesthesia was subsequently performed, and the excised specimen was submitted for histopathological examination [Table/Fig-3c,d]. Histopathology indicated fibrovascular granulomatous tissue. Postoperatively, the swelling resolved completely, with no complication and no recurrence to date [Table/Fig-3e,f].

Case 1, Case 2, Case 3, and Case 4 are described similarly with respect to presenting complaints, ocular findings, imaging characteristics, and management outcomes. All patients underwent a complete neurological examination (cranial nerves, motor and sensory systems, cerebellar signs), which was normal except for ocular motor involvement related to the fistula. Ophthalmic evaluation included BCVA, slit-lamp examination, Goldmann applanation tonometry, fundus examination, and exophthalmometry measured using the two-ruler method.

[Table/Fig-4] summarises the pre- and post-intervention ophthalmic outcomes, enabling objective assessment of treatment response, while [Table/Fig-5] provides a comparative overview of key clinical and radiological features across cases, improving clarity and facilitating cross-case clinico-radiological correlation.



[Table/Fig-3]: Ocular findings of case 4: a,b) Left-sided infraorbital swelling after combined endovascular and percutaneous embolization; c,d) Intraoperative pictures of the mass suggestive of fibrovascular granulomatous tissue; e,f) Postoperative day 1 and day 10.

congestion, chemosis, corkscrew episcleral vessels, raised IOP, and headache. These observations highlight that ocular signs often precede neurological manifestations. Classically, CCFs present with pulsatile proptosis, chemosis, arterialised episcleral vessels, bruit, diplopia, elevated IOP, and visual impairment, frequently leading to misdiagnosis, particularly in indirect (dural) CCFs. [8].

In the current series, three patients had post-traumatic CCFs with delayed onset of symptoms, ranging from months to years after injury. This delayed presentation is well documented in the literature, especially for indirect CCFs, which are known for their insidious course and subtle clinical signs. Direct CCFs are often post-traumatic and have an acute onset with prominent signs, whereas indirect CCFs (dural AVFs) develop insidiously, usually in older individuals with vascular risk factors [9]. Our series reflects these distinctions. Case 1 (direct CCF) demonstrated pronounced orbital congestion, raised IOP, and audible bruit, consistent with high-flow dynamics. Cases 2 and 3 (indirect CCFs) showed relatively milder but persistent symptoms with delayed diagnosis, a pattern well documented in dural fistulas. Recognition of these clinical contrasts is essential, as management strategies differ substantially.

Radiological correlation played a central role across all cases in our series. Initial evaluation includes non-invasive imaging such as CT and MRI [10]. MRI's high soft-tissue resolution enables detailed orbital assessment of proptosis, orbital oedema, dilated ophthalmic

Case No.	BCVA Pre (OD/OS)	BCVA Post (OD/OS)	IOP Pre (OD/OS) mmHg	IOP Post (OD/OS) mmHg	Proptosis Pre (mm)	Proptosis Post (mm)	Follow-up
1	6/18, 6/6	6/9 (p) 6/6	21/17.9	18.6/19	22/10	19/10	Lost
2	6/6, 6/6	6/6, 6/6	11/10.5	14.9/8.5	24/20	23/20	Lost
3	6/9, 6/12(p)	6/6, 6/9	13/12	10.5/16	20/25	20/23	Lost
4	6/6, 6/6	6/6, 6/6	15/9	13/10	Normal	Normal	Lost

[Table/Fig-4]: Ophthalmic outcomes before and after intervention.
*Normal exophthalmometry reference range (two-ruler method): 12-20 mm, inter-ocular; difference ≤2 mm [6].

Feature	Case 1	Case 2	Case 3	Case 4
Lesion type	Direct CCF	Indirect CCF	Indirect CCF	AVM
Flow type	High flow	Low flow	Low flow	
Barrow type	A	Type D	Type B	
Aetiology	Trauma	Trauma	Trauma	Trauma
Laterality	OD	OD	OS	OS
Treatment	Endovascular embolisation	Endovascular embolisation	Transvenous embolisation	Embolisation + excision
Complications	None	None	None	None

[Table/Fig-5]: Comparison of clinical and radiological features across cases.

DISCUSSION

The present case series highlights the diverse ophthalmic presentations of CCFs and orbital AVMs, underscoring the pivotal role of the ophthalmologist in early detection, while also reflecting how advances in imaging and interventional techniques have significantly transformed their diagnosis and management.

Barrow classified CCFs into direct (type A) and indirect (types B-D) based on angioarchitecture and arterial feeders [7]. Direct CCFs typically arise from a defect in the intracavernous internal carotid artery, producing a high-pressure arterial shunt into the cavernous sinus. This results in rapid venous hypertension, explaining the acute and dramatic clinical picture characterised by pulsatile proptosis, marked chemosis, bruit, elevated IOP, and early cranial nerve dysfunction. In contrast, indirect CCFs involve dural branches of the ICA/ECA, generating a low-flow shunt with gradual venous congestion. Consequently, patients often present insidiously with chronic red eye, mild proptosis, intermittent diplopia, or ocular discomfort. These haemodynamic differences account for the greater clinical severity and faster progression observed in direct CCFs compared with dural CCFs [1,3]. In our series, the predominant presenting features of CCFs were proptosis, conjunctival veins, and cavernous sinus enlargement and detection of vascular and tissue changes from venous congestion. Enlargement of the Superior Ophthalmic Vein (SOV) is a key sign on CT or MRI [11]. Orbital congestion, extraocular muscle swelling, and lateral cavernous sinus wall convexity may also be seen.

The DSA remains the gold standard for diagnosis, which offers high-resolution vascular mapping of feeders, fistula sites, venous drainage patterns, and collateral vessels, facilitating haemodynamic assessment and treatment planning [12]. Chen CC et al., reported a 94.4% sensitivity of DSA for CCF detection [13]. Advanced cross-sectional imaging plays a crucial role in distinguishing CCFs from other conditions that may present with similar clinical features, including neoplastic lesions, inflammatory processes, or infectious pathologies, by providing precise anatomical and pathological details [1].

Treatment strategies are both conservative and interventional. Conservative management includes observation, IOP-lowering drugs and intermittent carotid or SOV compression. These measures are appropriate in low-risk, non-vision-threatening dural CCFs, which close spontaneously in up to 70% of cases due to local thrombosis [14]. Indications for intervention include uncontrolled IOP, persistent diplopia, severe proptosis with corneal

exposure, optic neuropathy, retinal ischaemia, pronounced bruit, or cortical venous drainage [15].

Endovascular embolisation is first-line therapy, via transarterial or transvenous routes. Embolisation employs detachable coils, acrylic glue, or Onyx, singly or in combination, depending on angio-architecture and flow [16]. Flow-diverting stents may be used alone or with coils, reconstructing the parent artery and promoting fistula thrombosis [9]. The transvenous route via the Inferior Petrosal Sinus (IPS) is preferred for its short, direct access for most dural CCFs. Advances in microcatheters and guidewires have increased procedural success [17,18].

The orbital AVM in this series adds clinical relevance, as extracranial and orbital AVMs are rare and often pose diagnostic and therapeutic challenges. According to the International Society for the Study of Vascular Anomalies, AVMs are classified as high-flow vascular anomalies characterised by direct arteriovenous shunting that bypasses the capillary bed [19], and orbital AVMs may present with proptosis, conjunctival hyperaemia, corkscrew vessels, bruit, and raised IOP when symptomatic [20]. The need for combined endovascular embolisation followed by surgical excision in our case supports existing evidence that a multimodal approach provides better lesion control and reduces recurrence compared to single-modality treatment.

This case series contributes to existing knowledge by systematically documenting objective ophthalmic outcomes across different orbital vascular pathologies and treatment modalities, enabling meaningful comparison and demonstrating rapid functional improvement with timely intervention. The inclusion of both CCFs and a rare orbital AVM highlights the spectrum of vascular anomalies that can mimic primary ocular disease, underscoring the need for standardised assessment and multidisciplinary collaboration to optimise visual outcomes.

CONCLUSION(S)

The CCFs and AVMs are rare, vision-threatening vascular lesions that require timely diagnosis through clinico-radiological correlation using MRI and DSA. Subtle and seemingly benign ophthalmic signs such as unilateral redness, proptosis, raised IOP, or orbital swelling may be the earliest indicators of serious underlying vascular anomalies such as CCFs and orbital AVMs. Endovascular embolisation, often supplemented by surgery, remains the cornerstone of management. Early identification of characteristic ophthalmic features, combined with the use of advanced imaging modalities and coordinated multidisciplinary management involving ophthalmologists, radiologists, and neurosurgeons, is crucial for optimising patient outcomes.

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PARTICULARS OF CONTRIBUTORS:

1. Vice Principal and Head, Department of Ophthalmology, KAHER, Jawaharlal Nehru Medical College, Belagavi, Karnataka, India.
2. Postgraduate Student, Department of Ophthalmology, KAHER, Jawaharlal Nehru Medical College, Belagavi, Karnataka, India.
3. Head, Department of Interventional Radiology, KAHER, Jawaharlal Nehru Medical College, Belagavi, Karnataka, India.
4. Associate Professor, Department of Ophthalmology, KAHER, Jawaharlal Nehru Medical College, Belagavi, Karnataka, India.
5. Postgraduate Student, Department of Ophthalmology, KAHER, Jawaharlal Nehru Medical College, Belagavi, Karnataka, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Shivanand C Bubanale,
Kedar Nivas, CCB 371, 3rd Main 2nd Cross, Sadashivnagar, Belagavi, Karnataka, India.
E-mail: docshivanandcb@yahoo.co.in

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