Undiagnosed malignant hypertension presenting as a direct spontaneous carotid-cavernous fistula with complete loss of vision and hyphaema

Vinita Gupta 1, Saurabh Luthra,2 Athul Suresh Puthalath 3,1 Udit Chauhan1

1Department of Ophthalmology, AIIMS Rishikesh, Rishikesh, India
2Department of Ophthalmology, Drishti Eye Institute, Dehradun, India
3Department of Ophthalmology, All India Institute of Medical Sciences, New Delhi, India

Correspondence to
Dr Vinita Gupta;
drvinitagupta@hotmail.com

Accepted 7 November 2021

DESCRIPTION

A 60-year-old woman presented with complete loss of vision in the right eye (RE) with progressive protrusion and complete drooping of the right eyelid since 1 week (figure 1A,B). There was no history of trauma and patient was not on any systemic medications. Ocular examination revealed tense swelling of the eyelid with 6 mm axial non-pulsatile proptosis with an audible bruit in the RE along with total external ophthalmoplegia and severe chemosis. There was no perception of light in the RE. There was total hyphaema precluding view of intraocular details. Intraocular pressure (IOP) was elevated to 38 mm Hg. The left eye had best corrected visual acuity of 6/6 with a normal anterior segment. However, malignant hypertensive retinopathy changes were seen on posterior segment examination in the left eye (figure 1C–E). On systemic examination, she had elevated blood pressure of 220/130 mm Hg; however, she was not a diagnosed case of hypertension.

Ocular hypotensive treatment was initiated to control raised IOP and patient was referred for emergency medical management of her elevated blood pressure. On investigative work-up of the patient, no abnormality was detected in haemogram, peripheral smear examination, blood glucose and glycosylated haemoglobin levels, thyroid profile, liver function tests and lipid profile. Evaluation of her renal functions revealed normal serum creatinine, normal blood urea nitrogen, no abnormality on urinalysis and an unremarkable abdominal sonography. Her chest radiography was also normal. Her electrocardiography showed mild left ventricular hypertrophy. Patient was diagnosed to be in malignant phase hypertension and medical management for the same was initiated. Subsequent contrast enhanced CT (CECT) of brain and orbits revealed an enlarged right cavernous sinus with convexity of the lateral wall (figure 2A,B). Superior ophthalmic vein on right side was tortuous and dilated with early opacification on contrast along with thickened and bulky extraocular muscles (figure 2C,D). Based on these CECT findings corroborative of right direct carotid-cavernous fistula (CCF), patient was referred to neurosurgery for further management.

Direct CCFs are most commonly seen in young men and are due to trauma. However, they may also be caused by rupture of an internal carotid artery (ICA) aneurysm inside the cavernous sinus, Ehlers-Danlos type 4 or iatrogenic intervention.1 2 Indirect CCFs are usually seen in elderly hypertensive atherosclerotic patients.1 Rarely, a direct CCF can also be caused by rupture of an undiagnosed ICA aneurysm during hypertensive spikes in the elderly post menopausal women.3 Aneurysmal ruptures in these cases occur because of weakening of the aneurysmal wall by direct...
loss of vision with total hyphaema as the presenting feature. Oono et al have reported massive hyphaema in a case of spontaneous Barrow type C CCF.11 However, hyphaema in their case occurred following stereotactic radiosurgery which was necessitated 5 months after intervention with transcatheter arterial embolisation. Hyphaema in our case could be explained by the probable development of neovascularisation of the iris which can occur as early as 1 week following CRAO and Ophthalmic artery occlusions, the incidence and the rapidity of the onset of which is related to the level of overall ocular and retinal ischaemia.12

Learning points

► Spontaneous direct carotid-cavernous fistulas (CCFs) are extremely rare but potentially devastating cause of ocular symptoms, visual loss and periocular morbidity. They are usually caused by rupture of the intracavernous internal carotid artery aneurysms, which may occur during hypertensive spikes in elderly atherosclerotic women.

► A high index of suspicion of a direct spontaneous CCF should be kept in mind when a patient presents with rapidly progressive proptosis with severe visual loss even in the absence of trauma, connective tissue disorder or any pre-existing systemic disease.

► CCF patients may initially present to an ophthalmologist who should make a presumptive diagnosis, advice appropriate imaging studies and refer for timely neurosurgical intervention.

Figure 2  Contrast enhanced CT of orbit and brain findings consistent with right carotid-cavernous fistula (A) coronal section base of skull and (B) axial section base of skull-showing dilated right cavernous sinus with intense enhancement with convexity of the lateral wall (red arrow). (C) Axial section orbit showing dilated right superior ophthalmic vein (red arrow). (D) Coronal section orbit showing dilated right superior ophthalmic vein (red arrow) and bulky and oedematous extra ocular muscles on right side (yellow arrows).

increase in the mechanical stress. In addition, systemic hypertension can cause activation of local renin-angiotensin system leading to vascular inflammation and remodelling, and indirectly contributing to the rupture.4

Ophthalmic findings in CCF include proptosis, ocular bruit, eyelid swelling, proptosis, chemosis, arteriovenous conjunctival vessels, motility disturbances, decreased visual acuity, glaucoma, stasis retinopathy, central retinal vein occlusion, retinal ischemia, retinal artery occlusion and optic disc swelling.3 5 Visual loss may be secondary to corneal, retinal or optic nerve changes or may result from the accompanying glaucoma. Immediate visual loss in high flow direct CCF can be due to intraocular haemorrhage, central retinal artery occlusion (CRAO), traumatic optic neuropathy and posterior ischaemic optic neuropathy.6 8 Evaluation of a suspected CCF often involves non-invasive neuroimaging techniques which demonstrate a prominent SOV.3 4 However, digital subtraction angiography remains the gold standard if endovascular embolisation is contemplated.2 10

Our patient presented with total hyphaema with complete loss of vision as the presenting feature and was diagnosed as a case of direct spontaneous CCF with malignant phase hypertension based on clinical features and imaging. To the best of our knowledge ours is a first case of direct spontaneous CCF presenting with complete