

Pulsating exophthalmos of eye in carotido-cavernous fistula

Sir,

Carotido-cavernous fistula (CCF) is an entity which is commonly seen in traumatic injuries of the brain where the patient sustains an injury to the skull base. CCF may also develop spontaneously or rupture of intracavernous internal carotid artery. Most commonly the patient presents with sudden onset pain in the eyes, cranial nerve palsies, exophthalmos which can be pulsatile and varying degree of neurological deficits. Pulsatile exophthalmos is a classical sign seen in CCF which is generally associated with a bruit over the eyeball.

A 71-years-old female presented to our neurosurgical department with complaints of sudden onset, throbbing, and continuous pain in the right eye of 15 days duration associated with the pulsating movement of the right eyeball. The patient did not give any history of trauma to the eye or head injury. The patient did not have co-morbidities like hypertension or diabetes.

On clinical examination, the patient had pulsating mild exophthalmos of the right eye (video 1) associated with the diminished sensation of touch and pain over the right half of the face in the trigeminal nerve distribution. The patient also had bruit over the right eye, however, did not have any chemosis. Pupils were bilaterally 4 mm with normal reaction to light. A clinical diagnosis of CCF was entertained and she underwent diagnostic 4 vessels cerebral digital subtraction angiography (DSA). DSA revealed Barrow's Type ACCF (direct connection between the intracavernous internal carotid artery (ICA) and cavernous sinus).

Since this was a high flow direct fistula which has a significant risk of intracranial hemorrhage and neurological deterioration, she underwent endovascular intervention in the form of stent-assisted coiling for the closure of the fistula between the intracavernous ICA and the cavernous sinus. The patient was reviewed after three months and the pulsating exophthalmos of the right eye has resolved, however, she continued to have diminished sensation of the face in the trigeminal nerve distribution.

Carotido-cavernous fistula is an uncommon entity. The etiologies reported in literature are spontaneous (30 % of all CCFs) [1], traumatic (75% of all CCFs) [2] and secondary to rupture of aneurysms of cavernous segment of internal carotid artery (ICA) (3 to 24 % of patients with cavernous carotid aneurysms) [3]. These patients generally present with sudden onset headache, visual disturbances in 18% and palsies of cranial nerves coursing through cavernous signs [4]. The signs most commonly seen on clinical evaluations are in orbital bruit in 80%, complete ophthalmoplegia in 24%, proptosis in 72%, chemosis in 55%, cranial nerve VI palsy in 49%, and pulsating exophthalmos [4].

Pulsating exophthalmos is a pathognomic sign seen in CCF. Barrow has classified the CCF in four types [5] and it is most commonly seen in type A according to this classification. The pathophysiological basis for this sign is the transmitted pulsation of the ICA/ECA which is transmitted through the communication between the artery and the cavernous sinus and subsequently to the eyeball through the ophthalmic veins. The present recommendation for the management of CCF is mainly endovascular intervention [6] and closure of the communication between the artery and the vein using various materials like metallic coils and/or liquid embolic agents [6].

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