# Acute abducens nerve palsy as a presenting feature in carotid-cavernous fistula in a 6-year-old girl

## Abstract

Carotid-cavernous fistulas (CCF) are abnormal communications between the internal carotid artery and the cavernous sinus. Traumatic carotid-cavernous fistulae are rare potential complications of craniofacial trauma. Typical findings of CCF are proptosis, chemosis, headache, oculomotor or abducens nerve palsy, trigeminal pain and pulsating bruit over the temporal skull and the bulb.
CCF are reported very rarely in childhood. This report describes the clinical and radiological findings of a pediatric patient presented with CCF.

**Keywords:** carotid-cavernous fistula, child

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## Case report

A 6-year-old girl in previously good health presented with acute onset of inturning of the right eye and binocular diplopia of 10 days duration. There was no associated headache and eye pain. Her mother gave history of blunt injury by umbrella over right eye 10 days back.
On examination, visual acuity without correction was 20/20 in each eye, right eye had mild conjuctival congestion and normal findings on pupillary examination. Sensorimotor examination revealed a complete abduction limitation on the right. She had an esotropia of 45 PD when examined using the modified Krimsky method. The findings were consistent with a right abducens nerve palsy. Intraocular pressures were 18 mmHg in the right eye and 17 mmHg in the left eye. Patient was advised for MRI Brain imaging but she did not report back to clinic.
She presented again after 3 weeks with features of proptosis, chemosis, engorged veins in right eye (Figure 1). Visual acuity was 20/20 in both eyes. Slit-lamp biomicroscopy of the right eye revealed engorged episcleral vessels in a corkscrew configuration and clear cornea. Orbital and cephalic bruits were appreciated. Fundus examination revealed normal-appearing opticdisc, maculae, and retinal periphery with the exception of mild retinal vascular tortuosity in the right eye.
Magnetic resonance imaging (MRI) of the brain and orbits with contrast revealed fistulous communication of right cavernous sinus with enlarged right superior ophthalmic vein (SOV), enlarged right cavernous sinus with multiple flow voids, which were suggestive of carotid-cavernous fistula (Figure 2 and Figure 3). She underwent four vessel Digital Subtraction Angiography and intervention under general anaesthesia. The angiograms showed a direct CCF on right with small communication in horizontal part of cavernous ICA. High flow shunting was seen with predominant venous drainage through dilated SOV (retrograde flow) anteriorly and petrosal sinuses posteriorly. She was diagnosed as right direct CCF TYPE A and underwent intravascular coiling by transvenous route and transarterial route with complete obliteration of fistula after consulting interventional radiologists and neurosurgeons.
Postoperatively she had no neurodeficits, proptosis and conjuctival congestion improved significantly. On follow up visit after two month she had complete resolution of esotropia, proptosis and chemosis (Figure 4).

## Discussion

Carotid-cavernous fistula (CCF) is an abnormal arteriovenous communication in the cavernous sinus. The universally adopted classification system in the CCF literature is the schema developed by Barrow and his colleagues in 1985 [1] based on angiographic studies. According to their system, there are four types of CCF [2]:

1. Type A (direct): shunt between the ICA and cavernous sinus; usually associated with trauma (TCCF) and produce early signs and symptoms.
2. Type B (indirect): shunt between the meningeal branches of the ICA and cavernous sinus.
3. Type C (indirect): shunt between the meningeal branches of the external carotid artery and cavernous sinus.
4. Type D (indirect): shunt between the meningeal branches of the ICA, external carotid artery, and cavernous sinus.

Direct CCF, classified as type A or TCCF, are high-flow shunts, which occur three times as often as the indirect types. It is this category of CCF with which surgeons

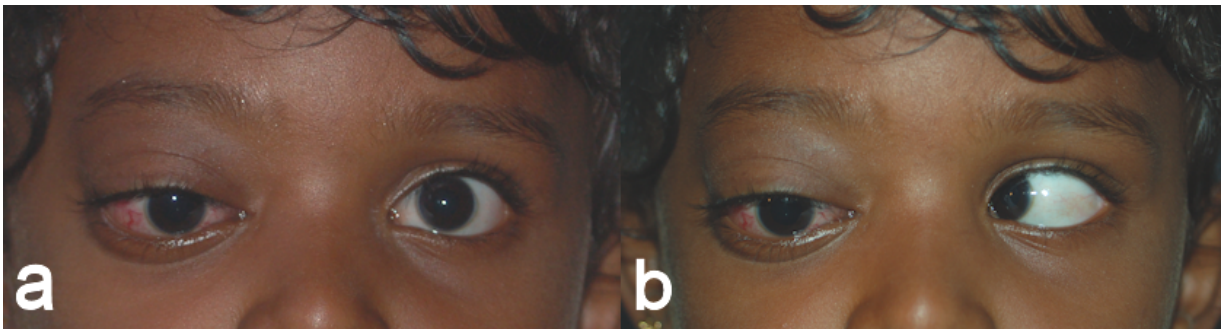


**Figure 1:** (a) showing dilated episcleral vessels and proptosis of the right eye and (b) showing abduction limitation of the right eye in right gaze.

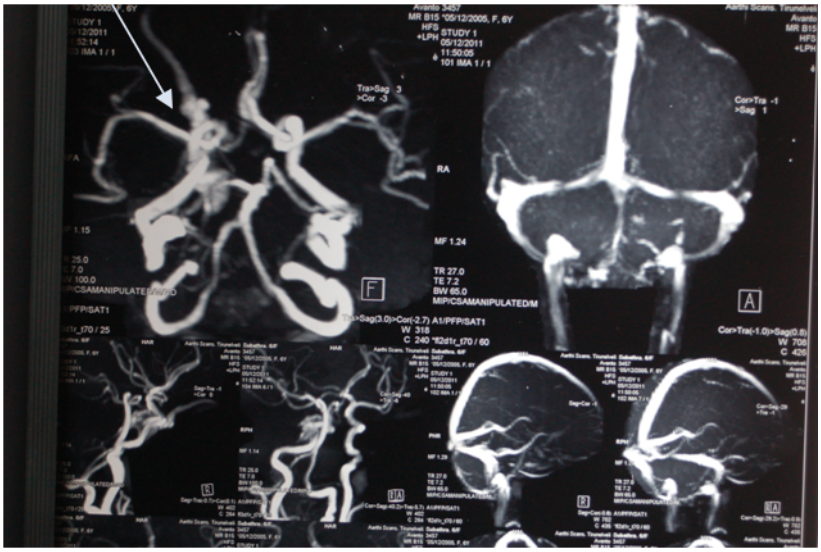

**Figure 2:** MRI showing carotid-cavernous fistula (arrow) and dilatation of right superior ophthalmic vein, with anterior venous drainage.

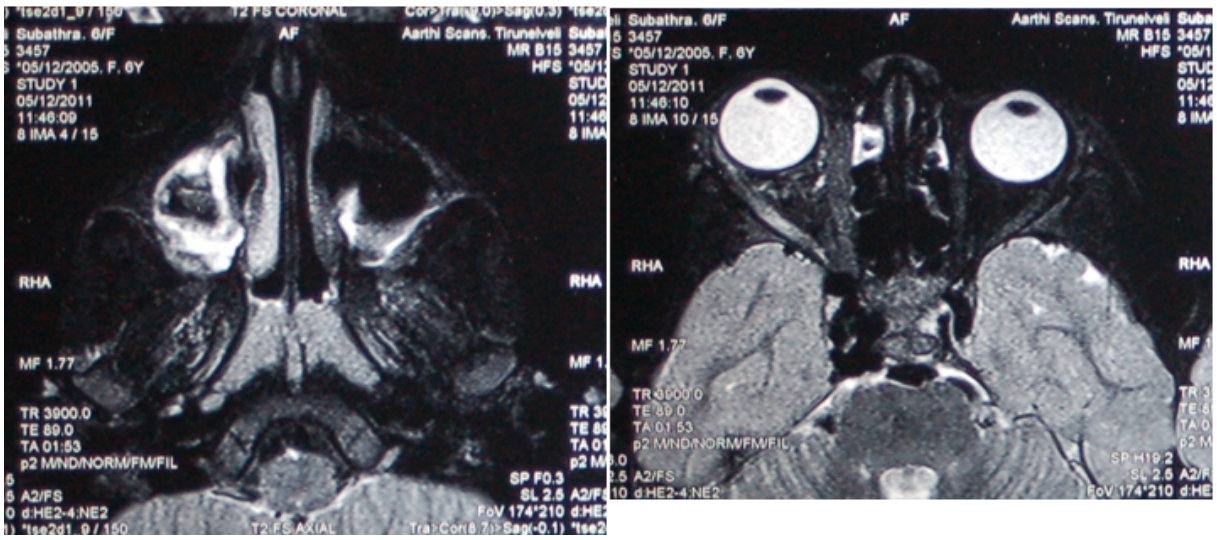


**Figure 3:** MRI showing exophthalmos in right eye and dilatation of superior ophthalmic vein.

managing craniofacial trauma should be most familiar. Direct CCF results from a tear in the intracavernous carotid artery. Typically, it has a high flow and usually presents with oculo-orbital venous congestive features such as pulsating exophthalmos, chemosis, orbital or cephalic bruit, impairment of vision due to hemodynamic disturbance in the optic nerve or the retina, and extra-ocular palsies with diplopia.Indirect CCF generally occurs spontaneously with subtle signs [3]. The specific diagnosis of traumatic CCF is usually not complicated, although the majority of indirect CCF are not diagnosed for weeks or even months after the onset of symptoms [2].

**Figure 4: showing pre- and post-treatment (intravascular coiling) with complete resolution of proptosis, and other symptoms of carotid-cavernous fistula after two months.**

**Table 1: Carotid-cavernous fistulas in the pediatric literature (0–10 year)**

| S. No. | Age | Sex | Associated features | Presenting symptom | Treatment | Outcome | Author |
|---|---|---|---|---|---|---|---|
| 1 | 7 weeks | M | Spontaneous | Proptosis | Gelfoam embolization | Resolution | Pang et al. [6] 1981 |
| 2 | 2 months | M | Spontaneous | Vision changes | Intravascular coiling | Resolution | Konishi et al. [7] 1990 |
| 3 | 5 years | M | Spontaneous | Proptosis | Direct ligation | Resolution | Gossman et al. [4] 1993 |
| 4 | 9 years | M | Spontaneous | Pain, conjunctival injection, vision change | Embolization | Resolution | Skolnick et al. [8] 2000 |
| 5 | 9 years | M | Spontaneous | Eye swelling, headache, neck pain | Conservative | Spontaneous resolution | Kurul et al. [5] 2001 |
| 6 | 8 months | F | Spontaneous | Proptosis | Coil embolization | Resolution | Rai et al. [9] 2004 |
| 7 | 3 months | M | Congenital | Proptosis, chemosis | Embolization | Resolution | Albayram et al. [11] 2004 |
| 8 | 6 years | F | CNS Choriocarcinoma | Proptosis, chemosis | Endovascularly embolized | Progressive disease, death | Lawton et al. [10] 2008 |

CCFs are rare in childhood and PubMed literature search reveals fewer than 10 cases in children under age 10 years (Table 1). Spontaneous resolution has been described in children and adults [4]. We are able to find 8 reported cases in English literature using the MEDLINE search under age of 10 (Table 1). Five of these patients required surgical intervention, such as embolization, and 1 underwent spontaneous closure [5], [6], [7], [8], [9], [10], [11]. Carotid-cavernous fistulas causing increase in intraocular pressure in infants and young children may lead to eyeball elongation and anisometropic amblyopia. Treatment is indicated for visual compromise, or for non-resolving or progressive cavernous sinus syndrome [11]. Therapeutic embolization is indicated if there is progressive decrease in vision, ophthalmoplegia, progressive proptosis, intractable glaucoma, pain or vascular steal syndrome. Transvenous embolization via the superior ophthalmic vein or superior orbital fissure, the double-balloon embolization technique, and coil embolization modalities have been used with various success rates [2], [11].

Our patient had history of only blunt trauma with no clinical evident signs of trauma and she presented with initial acute onset of esotropia and 3 weeks later features of CCF such as proptosis, engorged vessels appeared requiring surgical intervention. She underwent intravascular coiling and complete obliteration of fistula with resolution of proptosis, chemosis,dilated episcleral vessels and esotropia.

Cases presenting with motility impairment and history of trauma should be closely observed for subtle presentation of CCF. Close observation is necessary, and surgical intervention may be indicated when there are signs of impending ocular complications.

The authors urge close observation and monitoring of visual function and early treatment of symptomatic CCF to avoid serious complications, even though spontaneous regression of CCF has been reported in some cases.

# Notes

## Competing interests

The authors declare that they have no competing interests.

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