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Case Report

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Bilateral Traumatic Carotid-Cavernous Fistula in a Child: Corkscrew Eyelid Vessels as an Indicator of Severe Congestion

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ABSTRACT

An 11-year-old girl presented with bilateral traumatic caroticocavernous fistula associated with corkscrew eyelid vessels, which were considered indicators of severe congestive disease in this case. Coil embolization was performed; similar to other congestive findings such as proptosis, orbital bruit, increased intraocular pressure, congested scleral and retinal vessels, engorged eyelid vessels resolved immediately after coil embolization. This pediatric case is unique given the caroticocavernous fistula was bilateral and was associated with prominent dilatation of the eyelid vessels, named for the first time as corkscrew eyelid vessels.

KEYWORDS: Pediatric trauma, Carotid-cavernous sinus fistula, Proptosis, Congestion, Coil embolization

INTRODUCTION

A caroticocavernous sinus fistula (CCF) is an abnormal communication between the cavernous sinus and the carotid arterial system. Anatomically, it can be classified as direct or indirect (1). Direct fistulas are characterized by direct shunting of carotid arterial blood into the venous cavernous sinus, whereas indirect fistulas have additional vessels connecting the carotid circulation with the cavernous sinus. Connecting vessels may be meningeal branches of the internal carotid artery (ICA) or external carotid artery, or both. Direct CCFs are commonly associated with a history of head trauma, high flow, prominent eye, and requirement of an early treatment. Indirect CCFs on the other hand are usually spontaneous and are associated with low flow, subtle findings, and a benign course. Both direct and indirect CCFs are rare in the pediatric population (2,6). Thus, reports on the clinical course, management, and prognosis of CCFs in the pediatric population are lacking. Furthermore, most of the available pediatric literature discusses direct, non-traumatic fistulas.

We herein describe a rare case of a pediatric patient with bilateral traumatic CCFs and discuss the diagnosis, management, and outcomes.

CASE REPORT

An 11-year old girl presented to our ophthalmology department with several months of bilateral eye swelling and bruit. She denied pain but did report blurry vision and intermittent diplopia. Her history was notable for a car accident 3 months ago. Her medical record revealed the presence of multiple cranium bone fractures, left clavicula fracture, temporal bone fracture associated with otorrhagia, and otorrhea on the right. The patient was presented to the emergency department 3 hours after the accident; she was hospitalized and subsequently discharged home with general advice after medical care and neurological observation.

On physical examination, slight hearing loss in the right ear was noted. Neurological examination was within normal limits. Ophthalmic examination revealed that her uncorrected

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visual acuity was 20/40 in both eyes. Intraocular pressure (IOP) measurements were 23 mm Hg in the right eye, and 26 mm Hg in the left eye with a combination of dorzolamide + timolol maleat (Cosopt, MSD Pharm. Ind. Turkey). Pupillary examination was normal. Bilateral proptosis, eyelid swelling, and conjunctival hyperemia associated with tortuous vessels were observed (Figure 1A). Hertel measurements were not documented. When the eyes were closed, marked subcutaneous vessel dilatation was noted on the eyelids (Figure 1B). Ocular auscultation showed bilateral orbital and frontotemporal bruits. Extraocular movements were limited in all directions, and diplopia was present. Dilated fundus examination revealed tortuosity and venous congestion of the retinal vein in both eyes (Figure 1C, 1D).

Computerized Tomography (CT) examination was positive for dilated superior ophthalmic veins, with the dilation on the left being more prominent (Figure 1E). Based on the clinical and radiological findings (Figure 1E, 1F), bilateral CCF was suspected, and cerebral angiography was performed for a definitive diagnosis (Figure 2A, 2B). Cerebral angiography confirmed the diagnosis and endovascular treatment was started in the same session. Coil embolization was performed for the right side and the fistula was completely occluded with the preservation of distal flow to the right ICA. The left side had to be sacrificed using multiple balloons and trapping technique due to total transection of the left ICA. Although distal flow to the left ICA was not preserved with this technique, patient's Willis polygon enabled flow to left MCA owing to a patent anterior communicating artery as usually seen in the pediatric population. Final angiographic control showed complete cessation of the fistulous flow from the carotid system to the cavernous sinus and ophthalmic veins, with complete radiological cure (Figure 2C). Clinically, near-complete resolution of the congestive orbital findings was achieved in 2 days, and only minimal eyelid swelling remained (Figure 3A). Extraocular movements on both sides were full without any complaint of diplopia. Dilated fundus examination showed normal-caliber retinal veins in both eyes with a small retinal hemorrhage near the upper major arcade in the right eye (Figure 3B, 3C). Visual acuity was 20/20 in both eyes with 16 mm Hg of IOP measurement without any glaucoma treatment.

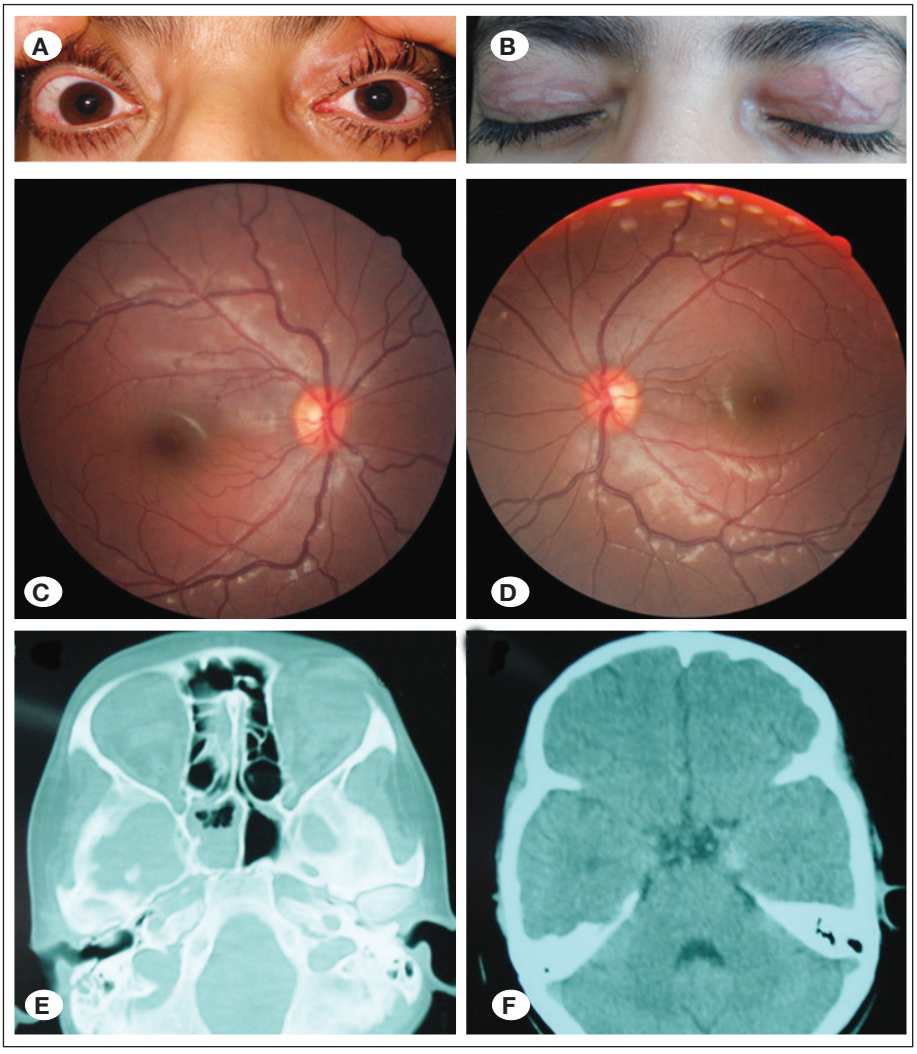

Figure 1: External ocular examination revealed bilateral proptosis, eyelid swelling, conjunctival hyperemia **(A)**, and marked subcutaneous vessel dilatation on the eyelids **(B)**. Fundoscopy revealed tortuosity and venous congestion of the retinal veins in both eyes **(C, D)**. An axial image from the head computerized tomography examination showed bilaterally dilated superior ophthalmic veins **(E)**, and engorgement of cavernous sinuses **(F)**, as an indicator of at least one direct caroticocavernous fistula (CCF).

DISCUSSION

Traumatic CCF is an extremely rare entity seen after a craniofacial trauma. However, incidence increases particularly in patients with skull base fractures. Therefore, its relationship with high-energy trauma and skull base fractures is well established (1). However, most reports have presented adult cases. With only few reports on pediatric cases of traumatic CCF, its actual incidence in this population is unknown. All described cases with traumatic CCF were unilateral lesions (5,8). Table I lists all pediatric cases of traumatic CCFs.

The medical history of our patient clearly supported the diagnosis of skull base fracture. Although the risk of CCF increases with the presence of high-energy trauma and skull base fracture, it can occur after any (mild, moderate or severe) head injury in children (2,6). The typical clinical trial is proptosis, chemosis, and orbital bruit. However, the spectrum is rather large and any of the following symptoms manifest: ophthalmoplegia, headache, visual disturbances, conjunctival injection, elevated IOP, secondary glaucoma, and optic neuropathy (5). Although less frequently, it may be

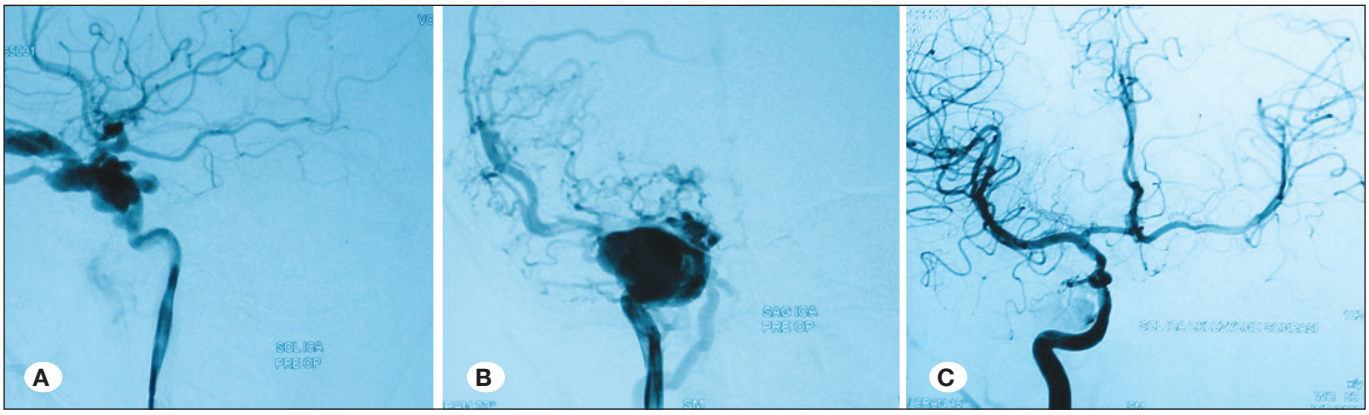

Figure 2: A lateral projection catheter digital subtraction angiogram (DSA) before treatment and an anteroposterior projection catheter DSA after endovascular treatment of bilateral fistulae. **A)** Selective contrast injection in the left carotid system through a left-sided catheter shows transection of the left internal carotid artery (ICA). The entire left ICA flow is diverted to the left cavernous sinus and left superior ophthalmic vein. **B)** Similar DSA results were seen for the right side except for shared blood flow volume between the extravasation toward the right cavernous sinus and normal intravascular flow toward the right cerebral hemisphere (not shown). **C)** Selective contrast injection through a catheter in the right carotid system shows blood flow confinement to the intraarterial compartments with complete cure of CCF sites. The left cerebral hemisphere is now being supplied by the right carotid system through the anterior communicating artery after endovascular occlusion (sacrifice treatment) of the left carotid system via trapping with detachable balloons.

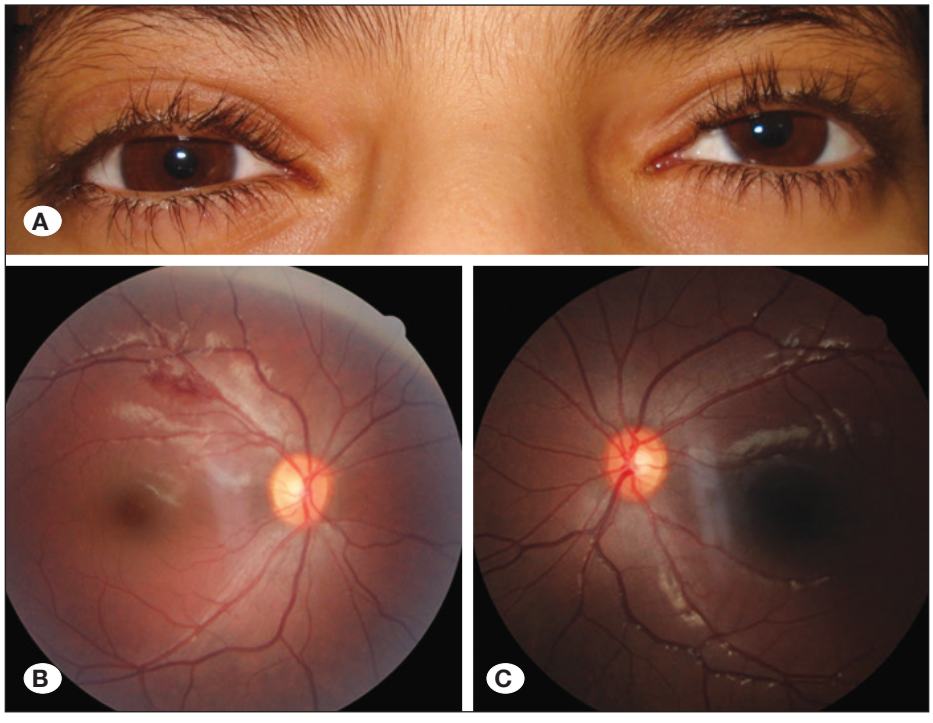

Figure 3: External ocular examination revealed near-complete resolution of congestive orbital findings with minimal eyelid swelling **(A)**. Normal-caliber retinal veins in the right eye with a small retinal hemorrhage near the upper major arcade **(B),** and normal-caliber retinal veins in the left eye **(C)** could be seen on fundoscopy.

Table I: Summary of Pediatric Age Carotico-cavernous Fistulas Reported in the Literature

	Age (years)	Etiology	Time*	Localization	Findings	Treatment	Year	Author
1	9	Falling Down From Height	5 months	Right	Proptosis	-	1984	Ciesielski et al., (3)
2	6	Falling Down From Height	20 days	Left	Exophthalmos Ophthalmoplegia	Coil Embolization	1996	Kanaan et al., (4)
3	7	Blunt Ocular Trauma	1 week	Right	6th nerve palsy	Coil Embolization	2004	Bekendam et al., (2)
4	11	Car Accident	On Time	Right	Proptosis	Endovascular Detachable Baloon	2013	Wyrick et al., (9)
5	6	Blunt Ocular Trauma	2 months	Left	Proptosis 6th nerve palsy	Coil Embolization	2015	Yang et al., (10)
6	10	Blunt Ocular Trauma	47 days	Right	Proptosis Ophthalmoplegia	Coil Embolization	2013	Paiva et al., (6)
7	6	Blunt Ocular Trauma	10 days	Right	6th nerve palsy	Coil Embolization	2013	Pawar et al., (7)
8	6	Car Accident	2 years	Left	6th nerve palsy	Coil Embolization	2016	Wajima et al., (8)
9	8	Penetrating Eyelid Injury	1 week	Left	Proptosis Ophthalmoplegia	Endovascular Detachable Baloon	2017	Morais et al., (5)

associated with epistaxis, otorrhagia, and intracerebral and subarachnoid hemorrhages which may lead to catastrophic morbidity or mortality outcomes (5). Although no evidence-based data correlating the severity of CCF and the severity of ophthalmologic findings exist, we hypothesize a correlation exists. Of interest, we observed marked dilatation of the eyelid vessels, which could be described as "corkscrew vessels." To our knowledge, such a finding has not been reported. Furthermore, prior reports of bilateral CCFs in a pediatric patient do not exist. Orbital volume increases linearly until the age of 15 years. Bilateral CCFs in a pediatric patient with developing orbits may be responsible for this unique finding. Further studies are needed to confirm this observation.

CONCLUSION

Endovascular embolization is the treatment of choice for CCF, and prompt reversal of ophthalmologic findings after treatment is well reported. Although the distal flow of the left ICA was not preserved in our case, resolution of symptoms was similar in both eyes. Barring minimal eyelid swelling and small retinal hemorrhage in the right eye, all congestive findings were resolved. An omission in this case was that we had not evaluated the effects of increased IOP on both eyes using any ancillary glaucoma tests. To the best of our knowledge, this is the first pediatric case of bilateral CCFs and corkscrew eyelid vessels, which may be an indicator of severe congestive disease.

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