



Challenging Management of an Acute Traumatic Carotid-Cavernous Fistula in a 2-Year-Old Child with Literature Review

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Abstract

Traumatic carotid-cavernous fistula (TCCF) is a rare occurrence in the pediatric population. However, the neurological sequelae of TCCF are associated with higher morbidity and mortality in pediatric patients. We report the case of a 2-year-old child with TCCF treated at a public hospital in Peru. The etiology of the injury was due to a fall of approximately 5 meters. The diagnosis was made based on the clinical picture and neuroimaging findings. The initial proposed treatment was performed with the hope of preserving the parent artery; however, due to persistence of the TCCF, embolization of the parent artery with coils and embolizing substance was performed. A literature review of similar cases was performed and identified eight cases in children under 10 years of age. Endovascular management of an acute TCCF is a challenge due to the high morbidity and mortality during the acute phase and can be complicated when other traumatic injuries are present. Maintaining the parent artery is important; however, when this is not possible, trapping the parent artery may provide an alternate option when appropriate collaterals exist.

Keywords

- traumatic carotid-cavernous fistula
- type A fistulas
- embolization

Introduction

Carotid-cavernous fistulas (CCFs) are an uncommon pathology in the pediatric population with an incidence of 4.6% per 100,000,¹ and are a vascular malformation resulting in abnormal connection between the internal (ICA) and/or external (ECA) carotid artery and the cavernous plexus. Barrow et al² classified CCFs into four types, type A being the most common. The primary etiology of type A fistulas is head trauma, representing approximately 70% of all CCF, the majority occurring in young adults.³ Type B and C result from

dural branches of the ICA and ECA, respectively. Type D results from both dural branches from ICA and ECA.⁴

Traumatic carotid-cavernous fistula (TCCF) is a very rare occurrence in the pediatric population, with a frequency of 0.2 to 0.3% after craniofacial trauma, and increasing up to 4% following skull base fractures.^{3,5,6} Despite its rare occurrence, TCCF reported in children can be accompanied by drastic consequences such as intracerebral hemorrhage, de novo aneurysm formation, and irreversible vision loss or ophthalmoplegia.^{6,7} Due to the high vulnerability of pediatric patients, the TCCF's neurological sequelae are associated with

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higher burden of visual disability and potential mortality in this population.

We report a rare case of a traumatic cavernous carotid fistula in a 2-year-old child treated with endovascular surgery at the National Institute of Child Health—San Borja in Lima, Peru. The local institutional ethics committee approved the report. A literature review in PubMed and Google scholar databases was performed, in which eight cases of TCCF were identified in children under 10 (►Table 1).

Illustrative Case

A 2-year-old child with no significant prenatal history sustained a fall of approximately 5 meters 1 hour prior to admission. On admission, the patient had a poor general condition, with multiple abrasions of the skull and mild right ocular edema. Due to severe intracranial hypertension, effacement of the basal cisterns, and fracture of the skull base, the patient underwent emergent decompressive craniectomy. Two hours following surgery, patient had neurological worsening and new brain tomography without contrast was obtained, which demonstrate diffuse cerebral edema and hydrocephalus (►Fig. 1A). She, therefore, underwent a second surgery for placement of an external ventricular drain. The patient was transferred to the intensive care unit, where exophthalmos, ocular edema, chemosis, and venous engorgement of the right eye became more evident (►Fig. 1B). Postoperative brain computed tomography (CT) scan at 6 hours showed hyperdensity at the right parasellar level (►Fig. 1C) Due to suspicion of a posttraumatic cavernous carotid fistula, cerebral CT angiography was performed, demonstrating a direct communication with the right ICA, with increased flow in the superior ophthalmic, the angular, and the facial vein (►Fig. 1D).

Based on the cerebral CT angiography results, we decided to perform endovascular procedure to close the CCF while preserving the parent artery. The approach was performed under the Seldinger technique, using a 4 Fr femoral introducer and a 4 Fr guide catheter. The initial angiography showed a high-flow CCF, Barrow classification type A, and dilatation of the superior ophthalmic vein (►Fig. 1E). The injection of the contrast substance through the left vertebral artery showed adequate flow to the right carotid territory through the right posterior communicating artery. Selective angiography using a 1.3 Fr microcatheter approached the fistula (►Fig. 1F) to release eight coils. The control injection demonstrated persistent flow from the ICA into the cavernous sinus (►Fig. 1G–H) so we decided to deploy the embolizing agent Onyx (Medtronic, Minneapolis, Minnesota, United States) to aid in plug formation at the CCF point, to obtain controlled embolization, and prevent distal Onyx migration. For this purpose, the microcatheter was flushed with 3 mL of sodium chloride, then 0.25 mL of dimethyl sulfoxide was flushed slowly over 40 seconds, and finally Onyx-18 was slowly injected under roadmap guidance (two-dimensional

image).¹² However, a control injection obtained after this procedure demonstrated persistent CCF inflow. We, therefore, decided to trap the right internal artery at the cavernous segment to permanently close the CCF; a total of 4.5 mL of Onyx was injected. Trapping of the parent artery was performed due to the persistent CCF inflow despite placement of coils and Onyx, and after demonstration of adequate collaterals to maintain right carotid circulation. The final result was closure of the fistula with adequate collateral flow maintained in carotid circulation on the right side from the right posterior communicating artery (►Fig. 1I–J).

After a satisfactory recovery, patient was discharged to home. At 2 months follow-up, ocular symptoms were markedly improved with preservation of vision in the right eye (►Fig. 1K). At 1-year, the patient has remained asymptomatic; however, no long-term imaging follow-up is available due to a change in the patient's health insurance coverage.

Discussion

Observations

Traumatic and spontaneous fistulas can occur in pediatric patients. Spontaneous fistulas are infrequent and often occur in the setting of other pathologies, such as Ehlers-Danlos syndrome.^{3,13} Traumatic fistulas, such as type A (direct) fistula, are most often due to head trauma associated with craniofacial trauma and skull base fracture.^{6,7}

Our patient, a 2-year-old girl, was younger than all previously reported cases (►Table 1). She presented with chemosis, proptosis caused by venous engorgement and palsy of the third cranial nerve, and orbital murmur caused by the arterial flow in the superior ophthalmic vein.³ Her traumatic high flow fistula resulted from a direct carotid lesion over the cavernous sinus following a basilar skull fracture.

The diagnosis was made based on the clinical picture and neuroimaging findings within 12 hours of admission. Given the acute of symptoms after presentation, the endovascular treatment was performed before major ophthalmological complications developed. The initial approach attempted to preserve the patent right ICA; however, as there was incomplete closure of the fistula and imaging demonstrated collateral flow, the right ICA was occluded; unlike other reported cases, we used embolic substance that had not been reported previously for treatment of TCCF in this age group (►Table 1).

Lessons

After embolization management with coiling and injection of embolizing substance, the patient's vision was preserved, and her symptoms improved. Endovascular management of an acute TCCF is a challenge due to the high morbidity and mortality during the acute phase and can be complicated when other traumatic injuries are present. Maintaining the parent artery is important; however, when this is not

Table 1 Reported cases of TCCF in the pediatric population under 10 years of age

Author/year	Number cases	Sex	Age	Side	Time to diagnosis	Treatment	ICA preservation	Evolution	Complete occlusion
Debrun et al, 1981 ⁸	1	F	5 y	ND	ND	Arterial access, removable balloons	No	ND	Yes
Barrow et al, 1982 ⁹	1	F	10 y	R	< 24 hours	Endovascular: 3 balloons	Yes	Yes, Recurrence of right oculomotor compromise after treatment, which remitted with high-dose steroids at 6-month follow-up	Yes
Paiva et al, 2013 ⁶	1	M	10 y	R	47 days	Venous approach, coils	Yes	Clinical improvement with amaurosis of the right eye	Yes
Pawar et al, 2013 ⁷	1	F	6 y	ND	10 days	Arterial and venous approach, coils	No	Complete resolution of symptoms at 2 months	Yes
Yang et al, 2015 ¹⁰	1	M	6 y	L	2 months	Venous coils	Yes	Mild proptosis of the left eye at 2 months	Yes
Wajima et al, 2017 ¹¹	1	M	6 y	L	2 years	Arterial coils	No	Complete resolution of symptoms	Yes
Morais et al, 2018 ³	1	M	8 y	L	7 days	Removable balloons	Yes	Slight improvement in ocular motility after 2 days of treatment	Yes
Tsuda et al, 2022 ⁵	1	M	10 y	R	28 days	1st intention: venous access, coils 2nd intention (21 days posttreatment 1): arterial access, flow diverter 3rd intention (2 weeks posttreatment 2): venous access, coils	Yes	Palsy of the cranial nerve on the third postoperative day. Decreased visual acuity and temporal hemianopsia	Yes
Our case	1	F	2 y	R	<12 hours	Arterial access coils and embolizing substance	No	Improvement of symptoms at 2 months of follow-up	Yes

Abbreviations: ICA, internal carotid artery; ND, not described; y, years old; L, left; R, right; TCCF, traumatic carotid-cavernous fistula.

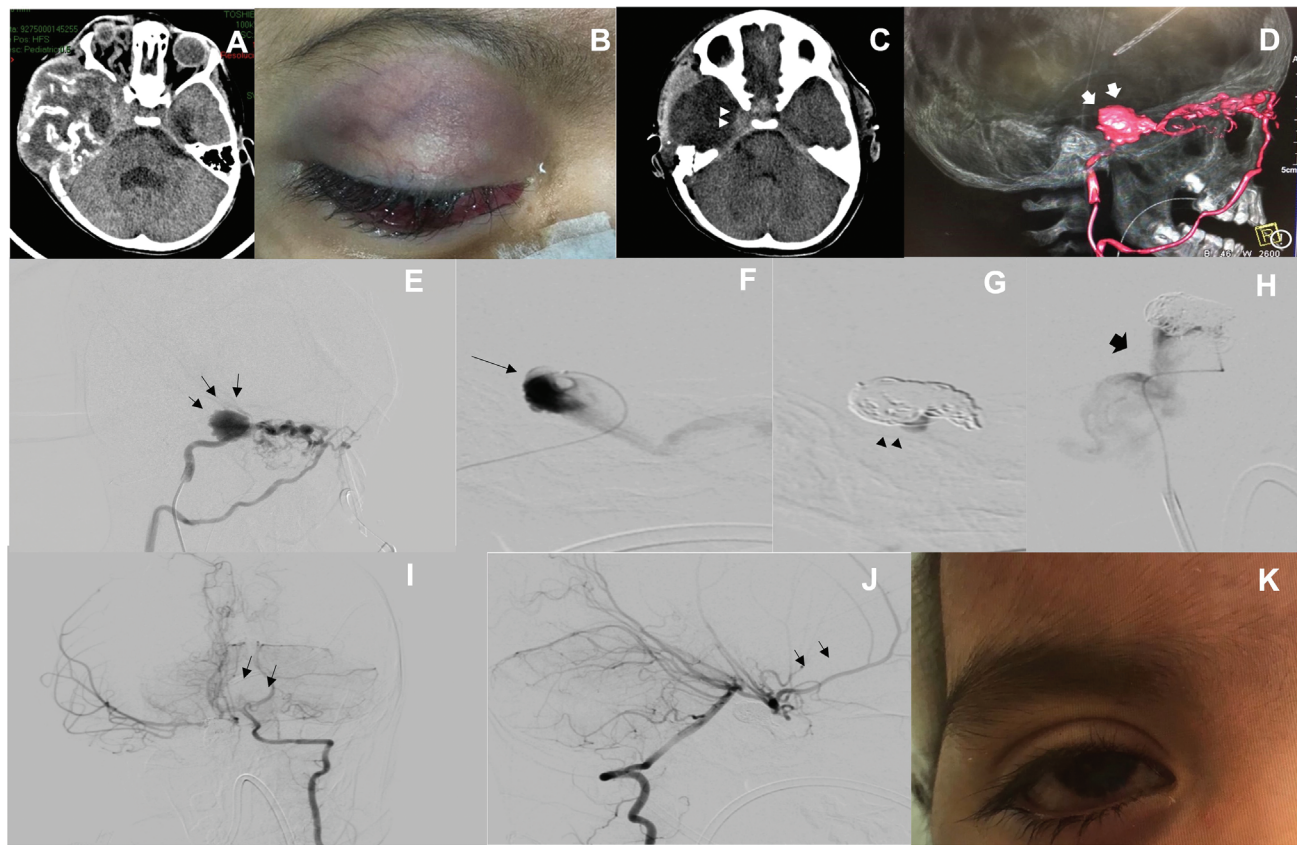


Fig. 1 (A) Two hours after admission a brain computed tomographic (CT) scan without contrast demonstrating right hemispheric injury and hydrocephalus. (B) Prior to treatment, showing right exophthalmos, ocular edema, and chemosis; (C) Six hours after admission, a brain CT scan without contrast demonstrating hyperdensity at the level of the right cavernous sinus (white head arrows); (D) Brain CT angiography showing the direct cavernous carotid fistula (white arrows); (E) Angiography in lateral view showing the cavernous carotid fistula with direct flow toward the superior ophthalmic vein, angular vein, and facial vein (black thin arrows); (F) Selective angiography showing the cavernous carotid fistula (black thin arrow); (G and H) After coil placement, partial flow persists toward the cavernous sinus through the fistula (black head arrows); (I and J) Final anteroposterior and lateral angiography showing adequate closure of the fistula, as well as adequate collateral flow that maintains flow in the right hemisphere through the left circulation; (K) Follow-up at 3 months after surgery demonstrating substantial ocular improvement.

possible, trapping the parent artery may provide an alternate option when appropriate collaterals exist.^{14,15}

Authors' Contribution

F.S., L.M., and M.T. were involved in data collection, conceptualization, and manuscript drafting. R.G. helped in data review and translation. R.E. contributed to data review, image preparation, technical and manuscript drafting.

Conflict of Interest

None declared.

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