Case Reports in Ophthalmology

Case Rep Ophthalmol 2023;14:363–368
DOI: 10.1159/000531568
Received: April 17, 2023
Accepted: June 12, 2023
Published online: August 14, 2023

Case Report

A Rare Case of Direct Spontaneous Carotid-Cavernous Fistula in a 6-Month-Old Infant and Review of the Literature

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Keywords
CCFs · Direct CCFs · Infants · Proptosis · Cavernous sinus · Superior ophthalmic vein

Abstract
Direct or type A CCFs are the direct connection between the cavernous segment of the internal carotid artery and the cavernous sinus. While most direct CCFs are caused by trauma, spontaneous direct CCFs are extremely rare in infants. In this report, we describe a 6-month-old child with bulges in the right eye that had been present since 20 days after birth. On examination, there was a right eye abduction limitation with no deviation associated with proptosis. Bruit was present during auscultation. CEMRI showed an enlarged right cavernous sinus with dilatation of the superior ophthalmic vein, suggesting a carotid-cavernous fistula. The patient was referred to an advanced center where he was advised patch therapy to prevent amblyopia. He was kept under observation by a neurosurgeon until 3 years, after which he was scheduled to undergo transarterial coiling.

Introduction

A carotid-cavernous fistula is an abnormal vascular connection between the internal or external carotid artery and the cavernous sinus. Direct or type A is a high-flow variant of CCFs with retrograde blood flow from the cavernous sinus into a superior ophthalmic vein (SOV)

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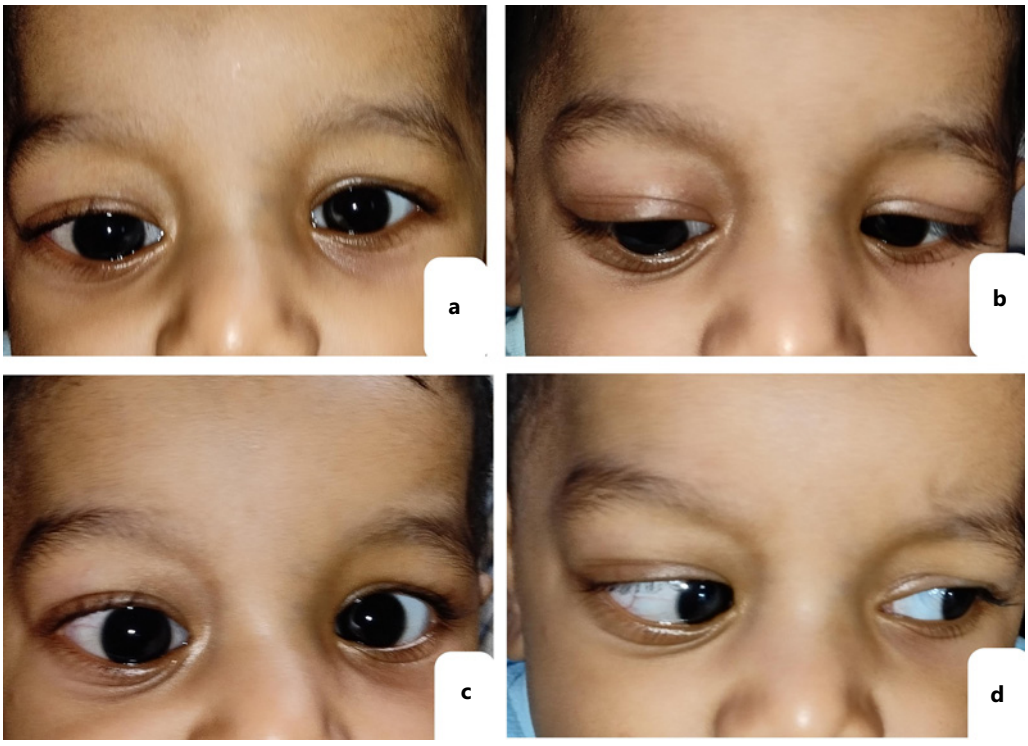

Fig. 1. **a** Primary gaze with inferior dystopia. **b** Down gaze showing right eye proptosis and eyelid fullness. **c** Right abduction limitation. **d** Levoversion.

leading to its dilatation. The majority of direct or type A CCFs are trauma related. Spontaneous direct CCFs may develop with the rupture of an intracavernous aneurysm or in patients with predisposing muscular and collagen disorders [1]. Direct CCF often presents with pulsatile proptosis, orbital bruit, and chemosis as well as oculomotor or abducens cranial nerve palsy [1]. Spontaneous direct CCFs in infants are very uncommonly reported in the literature with the youngest child being 9 months old. Our report presents a rare case of spontaneous direct CCFs in a 6-month-old infant having a history since 20 days after birth, without any sign of muscular or collagen disorders, presenting in our OPD as right proptosis with right abduction limitation.

Permission by the parents to print identifiable photographs was obtained and archived. The case described in this report is compliant with the Helsinki Declaration and health insurance portability and accountability act regulations. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000531568).

Case Report

A 6-month-old male, who had an uneventful delivery, was brought into the OPD by his parents for right eye bulging. It began 20 days after birth, but the parents delayed visiting since the symptoms were non-progressive and did not worsen with crying. There was no history of trauma or surgery. There was no sign of any other congenital anomaly.

Systemic examination was clinically unremarkable, ruling out any muscular and collagen disorder. Fixation was central, steady, and maintained with no resistance to left eye occlusion.

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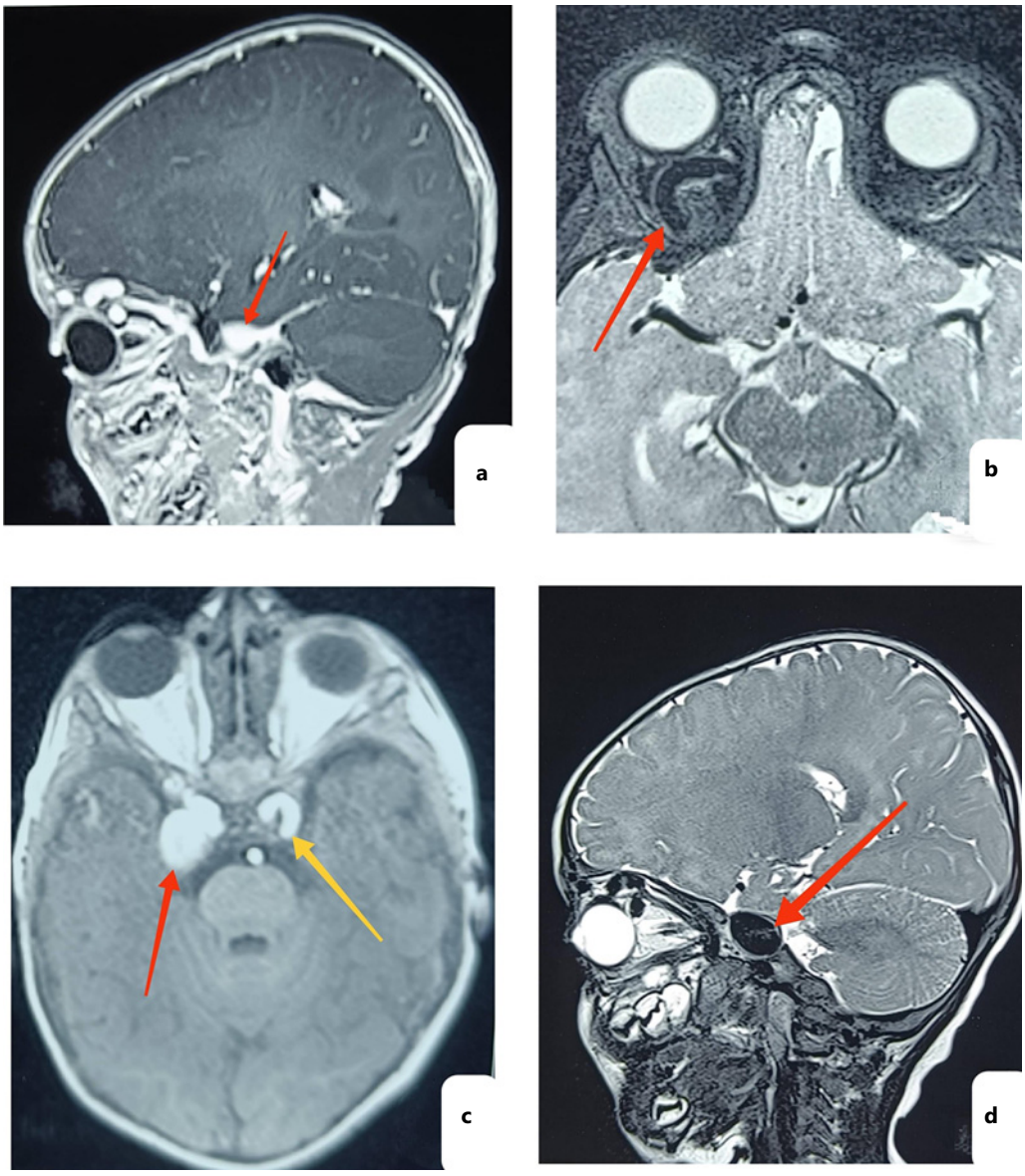

Fig. 2. a CEMRI T1W sagittal section showing hyper-intense dilated cavernous ICA/cavernous sinus (red arrow) with retrograde flow toward the SOV (yellow arrow). **b** T2W axial section showing the right dilated SOV (red arrow). **c** CEMRI T1W axial showing enlarged right cavernous sinus/cavernous ICA with lateral bulging (red arrow) and normal left ICA cavernous segment (yellow arrow). **d** T2W sagittal section showing an enlarged cavernous sinus/cavernous ICA (red arrow).

On examination, it was a non-reducible, non-compressible, and pulsatile proptosis with inferior dystopia (Fig. 1a, b). The intraocular pressure measured by the finger tension method and Perkins tonometry during sleep was within the normal range. The extraocular movement was limited to the right eye abduction with few dilated conjunctival vessels in the temporal region (Fig. 1c). Orbital bruit was present during auscultation. Examination of the anterior segment and the pupillary reaction was within normal limits, and the dilated fundus was clinically unremarkable. A provisional clinical diagnosis of vascular malformation was made. CEMRI showed an enlarged right cavernous sinus (Fig. 2a) with multiple tortuous and dilated vascular structures within, with asymmetrical dilatation of the right SOV (Fig. 2b), suggesting

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Table 1. Literature review

Subject No.	Age	Sex	Feature	Presenting symptoms	Treatment	Outcome	Author
1	5 yrs	M	Spontaneous	Progressive proptosis, hyperemia, abduction restriction	Balloon catheter technique	Resolution	Gossman MD, et al. [3] (1993)
2	11 mo	F	Spontaneous (h/o repeat lacrimal probing	Eyelid fullness, hyperemia	Transarterial embolization	Resolution	Ansaar TR, et al. [4] (2004)
3	3 yrs	M	Spontaneous	Progressive proptosis, hyperemia	Transvenous embolization	Resolution	Mercado GB, et al. [6] (2011)
4	9 mo	M	Spontaneous	Proptosis, hyperemia	Transarterial embolization	Resolution	Nikolaev AS, et al. [7] (2020)

right carotid-cavernous fistula (Type A). The largest intracavernous vasculature diameter is 12.5 mm, causing lateral bulging of the right cavernous sinus lateral wall (Fig. 2c). A right lateral rectus with 1.4 mm is thinner than a left lateral rectus with 3 mm. Carotid-cavernous fistulas require interventional radiologists and neurosurgeons; therefore, patient was referred to an advanced center for further management. After neurosurgeon's opinion, he was kept under observation until 3 years due to his stable clinical condition and young age, after which he was scheduled to undergo trans-arterial coiling. Additionally, patch therapy was recommended to prevent amblyopia. At the 5-month follow-up, a telephone interview revealed no worsening of symptoms.

Discussion

As described by Barrow et al. [1], CCFs are classified into 4 types based on the arterial system involved. Type A is direct fistulas, whereas types B, C, and D are indirect fistulas. CCFs are further classified on the basis of etiology, anatomical type, and hemodynamic type. Gupta et al. [4] identified 85 trauma-related causes out of 91 direct CCFs, while 6 cases were secondary to aneurysm rupture. Spontaneous direct CCFs are extremely rare in children, and few cases are documented in patients with muscular and collagen disorder.

Gossman et al. [3] first described spontaneous CCFs in 5-year-old children where the authors applied a combined surgical treatment strategy. Rai et al. [4] presented an 11-month-old infant with spontaneous direct CCFs with proptosis, dilated conjunctival vessels, and fullness of eyelids, after repeated probing and irrigation procedure. However, they concluded that it was unlikely to be a traumatic fistula because of its remote anatomic location and minimal tissue manipulation. Yakovlev et al. [5] analyzed 358 patients with direct CCFs where 317 had traumatic fistulas. In contrast, 38 had spontaneous fistulas, including 1 congenital fistula. Mercado et al. [6] documented a 3-year-old child with direct spontaneous CCFs presenting with progressive proptosis and hyperemia; they preferred transvenous embolization for management. Nikolaev et al. [7] described a 9-month-old child with direct spontaneous high-flow CCFs after nasolacrimal duct repair with left eye proptosis, conjunctival hyperemia, and vein tortuosity.

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During our literature review (Table 1), we found that this case of direct spontaneous CCFs presented at a very young age of 6 months, with a history that began 20 days after their birth. Imaging suggested a right carotid-cavernous fistula (Fig. 2), with thinning of right lateral rectus may be due to the sixth nerve involvement affecting lateral rectus function and leading to its wasting. Digital subtraction angiography is essential for confirming diagnosis and venous drainage patterns. Treatment is indicated for visual compromise or for non-resolving or progressive CCF. The goal of direct CCF treatment is to cease flow into the cavernous sinus and reconstruct cavernous ICA. Technique to achieve this include coil embolization (trans-arterial or transvenous), coil embolization with balloon remodeling of the ICA, and coil embolization with stent assistance.

Conclusion

Spontaneous direct CCFs presenting as right abduction limitation with pulsatile proptosis and dystopia in a 6-month-old infant is an extremely rare entity. This report describes the presentation of CCFs in a youngest age group with review of the literature. Therefore, early diagnosis and treatment are imperative in preventing vision-threatening complications.

Statement of Ethics

Ethical approval is not required for this case report in accordance with local and national guidelines. Written informed consent was obtained from the parents of the patient in question to publish their medical case and accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Funding Sources

The authors had no funding sources for the research and publication of this article.

Author Contributions

Sandeep Pal, Pradhnya A. Sen, Narendra Patidar, Harshdeep Singh Gabba, and Sangeeta Bhadra contributed to data collection, data analysis, manuscript writing, manuscript editing, and manuscript review.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material. Further inquiries can be directed to the corresponding author.

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