Ophthalmology Clinics & Research
OCR, 2(2): 73-77
www.scitcentral.com

ISSN: 2638-115X

Case Report: Open Access

# Rare Presentation of Spontaneous, Direct, Carotid Cavernous Fistula in Late Pregnancy: A Case Report

**Nisha Agrawal[1*], Lalit Agarwal[2], Abhishek Anand[3] and Archana Kumari[4]**

*[*1]Taparia Eye Care Pvt. Ltd., Banjara Chowk, Biratnagar, Nepal*

*[2]Vitreo Retina Department, NAMS, Biratnagar Eye Hospital, Biratnagar, Nepal*

*[3]Raj Retina Eye Centre, Patna, India*

*[4]Biratnagar Eye Hospital, Atithimarg, Biratnagar-17, Nepal.*

*Received May 25, 2019; Accepted May 31, 2019; Published August 06, 2019*

## ABSTRACT

**Purpose:** To report a rare case of spontaneous, direct, Type A Carotid cavernous fistula in late pregnancy.

**Observation:** A 30 year old woman presented to us 1month postpartum, with right-sided headache, progressive axial proptosis, total external ophthalmoplegia and diminution of vision which started one day prior to term normal delivery. Magnetic Resonance Imaging (MRI) and Digital Subtraction Angiography (DSA) confirmed direct anomalous communication between the cavernous sinus and carotid artery system.

**Conclusion:** Carotid Cavernous Fistula (CCF) may be associated with life and vision threatening implications. Meticulous clinical and radiological evaluations are critical for accurate diagnosis of this challenging condition. Not all CCF undergo spontaneous resolution. Treatment modalities should be individualized depending upon the rate of flow, type of shunt and stage of pregnancy. A high index of suspicion is deemed necessary to prevent delayed and missed diagnosis.

**Keywords:** Carotid cavernous fistula, Pregnancy, Direct, Proptosis, Diminution of vision, Chemosis

**Abbreviations:** MRI: Magnetic Resonance Imaging; DSA: Digital Subtraction Angiography; CCF: Carotid Cavernous Fistula; ICA: Internal Carotid Artery; ECA: External Carotid Artery; RE: Right Eye; LE: Left Eye

## INTRODUCTION

Anomalous communication between the carotid artery and cavernous sinus is known as carotico-cavernous fistula (CCF) [1]. Angiographically, they can be classified as direct; if the shunt is directly with the internal carotid artery (ICA) or indirect (dural) if the communication is with meningeal branches of ICA or external carotid artery (ECA) [2]. Pregnancy is a known precipitator of spontaneous CCF [3]. Haemodynamic and hormonal changes in pregnancy can lead to enlargement of aneurysm or devastating complications like cerebral hemorrhage [3-5]. Indirect CCF is a documented but rare condition in pregnancy comprising of limited number of reports in the literature [3,6,7]. A spontaneous Direct Type A Carotid Cavernous fistula is even rarer. We report such a case of angiographically confirmed direct CCF in a young female presenting with rapid onset proptosis in the peripartum period.

## CASE PRESENTATION

A 30 years old woman presented to us with history of Right sided headache and outward protrusion of Right eye (RE) since 1 month. Her symptoms started a day prior to full term normal vaginal delivery at home. Thereafter, it progressed gradually along with onset of proptosis, dizziness and vomiting.

There was no anteceding history of trauma, convulsion or limb weakness. Baby was born healthy and growing well.

Prior to presenting to us, she was treated with intravenous antibiotics with the suspicion of Orbital cellulitis without any improvement.

**Corresponding author**: Dr. Nisha Agrawal, Taparia Eye Care Pvt. Ltd., Banjara Chowk, Biratnagar, Nepal, Tel: 009779852049870; E-mail: drnisha_agr@hotmail.com

**Citation:** Agrawal N, Agarwal L, Anand A & Kumari A. (2019) Rare Presentation of Spontaneous, Direct, Carotid Cavernous Fistula in Late Pregnancy: A Case Report. Ophthalmol Clin Res, 2(2): 73-77.

Blood pressure was recorded as 110/80 mm Hg. Pallor was present. Systemic examination was unremarkable.

Ocular examination revealed best corrected visual acuity of 20/200 in the RE and 20/20 in left eye (LE). There was presence of RE proptosis measuring 25 mm on Hertels exophthalmometer. Proptosis was axial, non-tender, non-compressible, non-pulsatile and not associated with bruit. There was complete limitation of extra ocular movements along with lagophthalmos. Inferior subconjuctival space was severely engorged with bluish hue **(Figure 1)**. Anterior segment was unremarkable. Fundus examination revealed hyperaemic disc with well-defined margins. Peripapillary choreo-retinal striae were noted. Goldmann applanation tension was 17 mm Hg in both eyes.

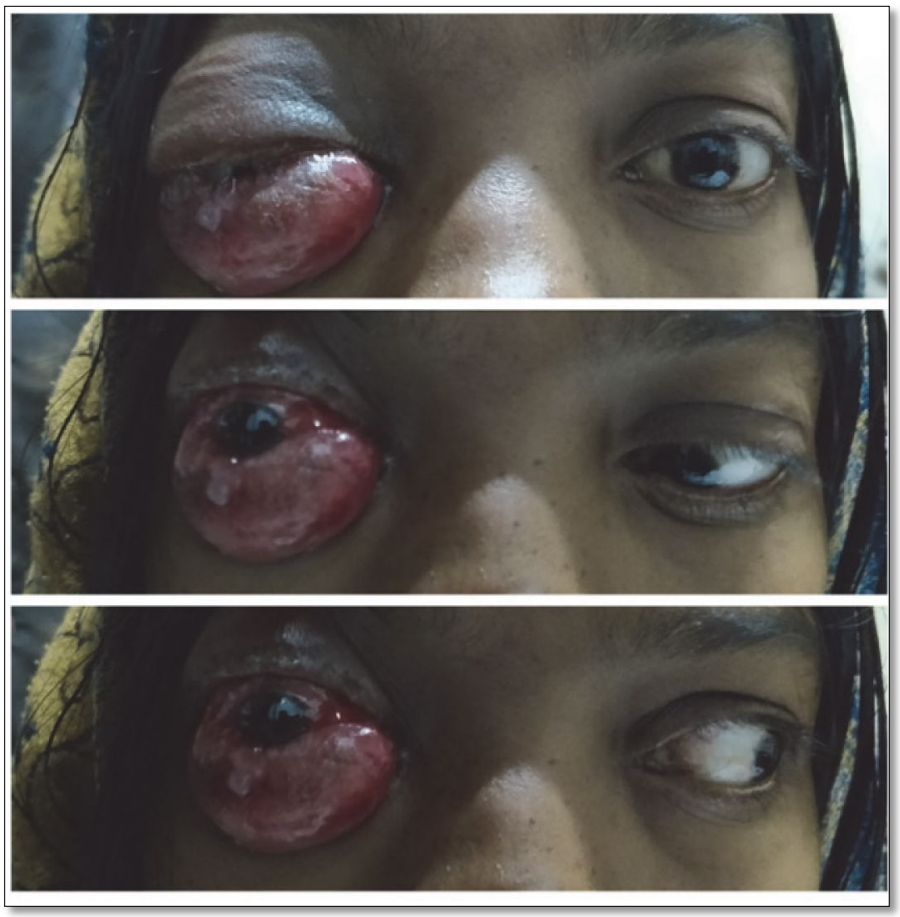

**Figure 1.** Clinical photograph of the patient. Inferior conjuctival space is markedly engorged with bluish hue. There is limitation of movement of right eye dextroversion and levoversion.

Contrast Enhanced Computed Tomography of orbit was ordered which revealed RE proptosis, dilated tortuous vessels in retrobulbar region, prominent Right cavernous sinus with convex lateral margin.

Further Magnetic Resonance (MRI) Imaging displayed Right CCF with dilated right superior ophthalmic vein (Hockey stick sign) [1] showing prominent flow voids with significant RE proptosis **(Figure 2)**.

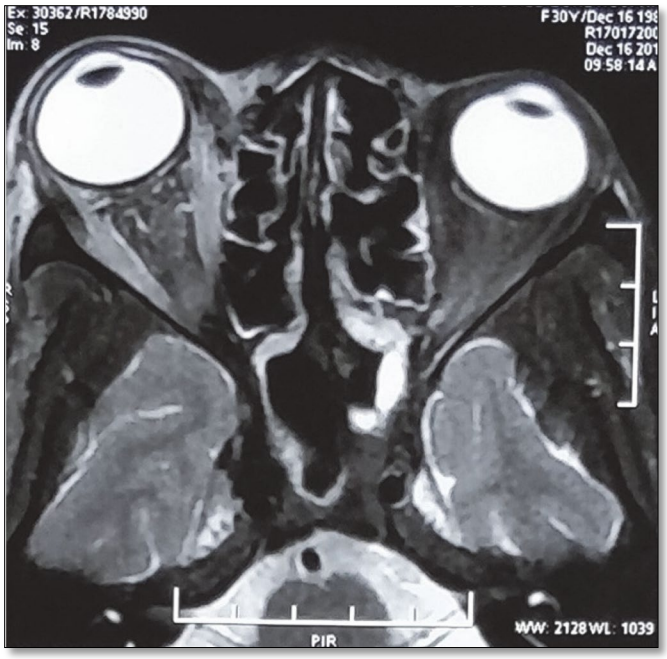


**Figure 2.** Magnetic resonance imaging of a 30 year female displaying proprtosis of right eye and right carotid cavernous fistula with dilated right superior ophthalmic vein (Hockey stick sign) and also showing prominent flow voids.

Digital Substraction Angiography (DSA) confirmed direct CCF with venous engorgement of orbit. Venous drainage via Right superior ophthalmic vein anteriorly, right inferior petrosal sinus posteriorly, superficial middle cerebral vein superiorly and minimal reflux into ipsilateral pterygoid plexus. Poor arterial collaterals to right hemisphere via PCOM and ACOM were noted **(Figure 3)**.

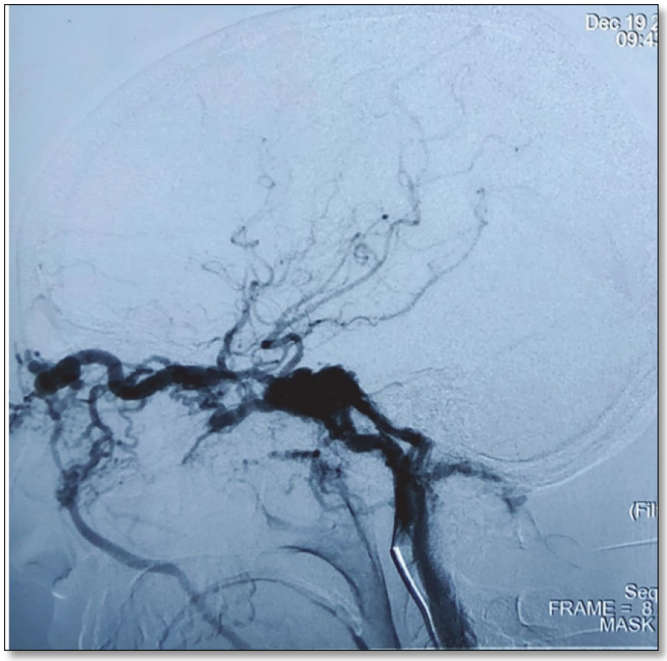


**Figure 3.** Digital substraction angiography of a 30 year female showing direct carotid cavernous fistula with venous engorgement of orbit. Venous drainage via Right superior ophthalmic vein anteriorly, right inferior petrosal sinus posteriorly, superficial middle cerebral vein superiorly and minimal reflux into ipsilateral pterygoid plexus. Poor arterial collaterals to right hemisphere via PCOM and ACOM were noted.

Patient was advised endovascular coiling of right CCF which she refused because of financial constraint and thereafter she failed to turn for follow up.

## DISCUSSION

CCF comprises one or more anomalous arteriovenous communication between the cavernous sinus (CS) and the carotid arterial system [1]. These lesions can be classified on basis of aetiology as traumatic and spontaneous, haemodynamically as high and low flow [2]. 25% of them are spontaneous fistulas [1]. Angiographically, it can be classified into four types. Type A is direct connection between ICA and CS. Type B, C and D are dural shunts in which there is anomalous connection of CS with meningeal branch of ICA, ECA and both, respectively [2]. Spontaneous low flow system is commonly associated with pregnancy [1]. However, our patient presented with spontaneous high flow fistula which has rarely been reported.

Pregnancy, physical straining, atherosclerosis and collagen vascular disease is well known precipitator of these anomalous arterio-venous shunts [3,7]. Pregnancy is characterized by hemodynamic and hormonal changes which make the vessel wall leaky leading to aneurysm formation, enlargement, rupture and fistula formation [8]. Blood pressure of our patient was constantly normal throughout pregnancy and postpartum.

Usually 25 to 30% of the pregnant woman present with this condition in the late third trimester or during delivery because of increased cardiac output [3,5,9]. Similarly, in our patient, symptom first appeared a day prior to delivery and continuously progressed thereafter.

Commonly, presenting clinical signs are proptosis (81%), diplopia (68%), cranial bruit (49%), headache (34%) and chemosis (87%) [9,10]. Our patient presented with proptosis, chemosis, oculomotor paresis and visual loss. This was not associated with any history of head injury. Such patients may be misdiagnosed with infectious and non-infectious conditions. Headache is a common complaint among pregnant women which should not be neglected without detailed evaluation if warranted.

There are reported instances of complications like rapid enlargement of aneurysm or rupture of aneurysm leading to subarachnoid haemorrhage in 1 in 10000 pregnancies [4,5,11]. Intracerebral haemorrhage can lead to fetal and maternal morbidity and mortality [12]. Our patient did not develop any hemorrhage.

Spontaneous resolution is known in 5 to 60% of the cases [5,6]. Hirata et al. [3] described a case of marked regression of a CCF 2 to 3 days after delivery and speculated that spontaneous improvement in CCF after pregnancy was due to thrombosis related to changes in blood coagulation that occur during pregnancy and delivery. Unfortunately our patient did not resolve spontaneously and her symptoms progressively deteriorated till the time of presentation.

Weather to deliver a child vaginally or by caesarean section is still a matter of debate. Some authors have suggested caesarean delivery to avoid valsalva maneuver induced raised intraocular pressure [7,13].

Barrow et al. [2] proposed the following as indications for treatment of a spontaneous CCF: (1) visual deterioration; (2) obstructive diplopia related to vascular engorgement and enlargement of the extra-ocular muscles or neural compression within the cavernous sinus; (3) intolerable bruit or headache; and (4) malignant proptosis with untreatable corneal exposure.

The timing of treatment should be decided according to the presentation. Cases presenting with complication in early or mid-third trimester may warrant need for immediate delivery, even preterm [12]. It has been suggested that pregnancy towards term should be dealt with delivery of child first followed by treatment as involves exposure of infant to irradiation. Timing of treatment following delivery may vary as early as 1 day post-partum to 7 weeks post-delivery [7,14]. Detachable balloon and Endovasular coiling is a documented successful treatment option [7,14]. There is a case report of successful treatment with craniotomy and occlusion of ICA during pregnancy but the patient delivered preterm [12].

## CONCLUSION

Spontaneous, direct CCF during pregnancy is a rare condition. It may be associated with life and vision threatening implications. Meticulous clinical and radiological evaluations are critical for accurate diagnosis of this challenging condition. Not all CCF undergo spontaneous resolution. Treatment modalities should be individualized depending upon the rate of flow, type of shunt and stage of pregnancy. A high index of suspicion is deemed necessary to prevent delayed and missed diagnosis.

## REFERENCES

1. Das JK, Medhi J, Bhattacharya P, Borah N, Bhattacharjee K, et al. (2007) Clinical spectrum of spontaneous carotid-cavernous fistula. Indian J Ophthalmol 55: 310-312.
2. Barrow DL, Spector RH, Braun IF, Landman JA, Tindall SC, et al. (1985) Classification and treatment of spontaneous carotid-cavernous sinus fistulas. J Neurosurg 62: 248-256.
3. Hirata Y, Matsukado Y, Takeshima H, Seto H (1988) Postpartum regression of a spontaneous carotid-cavernous fistula - Case report. Neurol Med Chir (Tokyo) 28: 673-676.

4. Weir BK, Drake CG (1991) Rapid growth of residual aneurismal neck during pregnancy. Case report. J Neurosurg 75: 780-782.

5. Lin TK, Chang CN, Wai YY (1992) Spontaneous intracerebral hematoma from occult carotid-cavernous fistula during pregnancy and puerperium. Case report. J Neurosurg 76: 714-717.

6. Toya S, Shiobara R, Izumi J, Shinomiya Y, Shiga H, et al. (1981) Spontaneous carotid-cavernous fistula during pregnancy or in the postpartum stage. J Neurosurg 54: 252-256.

7. Yeung SW, Suen SSH, Yu SCH, Lao TT, Leung TY, et al. (2013) Spontaneous carotid cavernous fistula complicating pregnancy. Hong Kong Med J 19: 258-261.

8. Marshman LA, Aspoas AR, Rai MS, Chawda SJ (2007) The implications of ISAT and ISUIA for the management of cerebral aneurysms during pregnancy. Neurosurg Rev 30: 17.

9. Walker AE, Allegre GE (1956) Carotid-cavernous fistulas. Surgery 39: 411-422.

10. Meyers PM, Halbach VV, Dowd CF, Lempert TE, Malek AM, et al. (2002) Dural carotid cavernous fistula: Definitive endovascular management and long-term follow-up. Am J Ophthalmol 134: 85-92.

11. Ortiz O, Voelker J, Eneorji F (1997) Transient enlargement of an intracranial aneurysm during pregnancy: Case report. Surg Neurol 47: 527-531.

12. Raskind R, Johnson N, Hance D (1977) Carotid cavernous fistula in pregnancy. Angiology 28: 671-676.

13. Lawrence MS (2003) Effects of bending, lifting and valsalva maneuver on intraocular pressure. Invest Ophthalmol Vis Sci 44: 1297.

14. Dogan S, Salman MC, Deren O, Geyik S (2012) Carotid-cavernous fistula in term pregnancy due to spontaneous rupture of carotid-cavernous aneurysm. J Obstet Gynecol 38: 427.