Multigravida 37 Weeks Pregnant Not in Labour with Carotid Cavernous Fistula Life Single Fetus Head Presentation: Case Report

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Abstract

Background: Carotid cavernous fistula (CCF) is an abnormal shunt from the carotid artery to the cavernous sinus. The management of pregnant patients with CCF is individualized. The aims of this case report are to document a rare presentation of a multigravida at 37 weeks of gestation with a carotid cavernous fistula, describe clinical symptoms and management, report outcomes, and contribute insights to the medical literature.

Case Report: The referred patient, G2P1A0, who was 37 weeks pregnant with a live single fetus in cephalic presentation, presented with left eye swelling persisting since the first pregnancy at 6 months gestation, associated with headaches. The patient underwent neurosurgical intervention at Mohammad Hoesin Hospital, including digital subtraction angiography (DSA). Currently, experiencing preterm labor symptoms, the management includes inpatient care, blood transfusion (Hb > 10 g/dL), and termination via the perabdominal approach.

Discussion: A multigravida at 37 weeks pregnant in labor with carotid cavernous fistula and a live single fetus in head presentation, as existing literature suggests, has no clear link between maternal carotid cavernous fistula history and fetal outcomes. Despite concerns about potential fetal abnormalities and cancer risk from endovascular embolization therapy during pregnancy, postpartum follow-up with advanced digital subtraction angiography (DSA) is planned.

Conclusion: The complexity of managing a multigravida at 37 weeks pregnant in labor with carotid cavernous fistula and a live single fetus in head presentation emphasizes the importance of a multidisciplinary approach for optimal maternal and fetal outcomes.

Key words: Carotid Cavernous Fistula, Digital Subtraction Angiography

Multigravida Hamil 37 Minggu Belum Inpartu dengan Fistula Cavernosa Karotis Janin Tunggal Hidup Presentasi Kepala: Laporan Kasus

Abstrak

Latar belakang: Fistula kavernosus karotis (CCF) adalah celah/ lubang abnormal dari arteri karotis ke sinus kavernosus. Penatalaksanaan pasien hamil dengan CCF bersifat individual. Tujuan dari laporan kasus ini adalah untuk mendokumentasikan presentasi langka seorang multigravida pada usia kehamilan 37 minggu dengan fistula kavernosus karotis, mendeskripsikan gejala klinis dan penatalaksanaannya, melaporkan hasil, dan menyumbangkan wawasan untuk literatur medis.

Laporan Kasus: Pasien yang dirujuk, G2P1A0, pada usia kehamilan 37 minggu dengan janin tunggal hidup dengan presentasi kepala, datang dengan pembengkakan mata kiri yang berlangsung sejak kehamilan pertama pada usia kehamilan 6 bulan, yang berhubungan dengan sakit kepala. Pasien menjalani intervensi bedah saraf di Rumah Sakit Mohammad Hoesin, termasuk Digital Subtraction Angiography (DSA). Saat ini, mengalami gejala persalinan prematur, penatalaksanaan yang dilakukan meliputi rawat inap, transfusi darah (Hb > 10 g/dL), dan terminasi melalui pendekatan perabdominal.

Diskusi: Seorang multigravida dengan usia kehamilan 37 minggu yang melahirkan dengan fistula kavernosa karotis dan janin tunggal hidup dengan presentasi kepala, merupakan kasus yang jarang terjadi, karena literatur yang ada menunjukkan tidak ada hubungan yang jelas antara riwayat fistula kavernosa karotis ibu dan hasil janin. Meskipun ada kekhawatiran mengenai potensi kelainan janin dan risiko kanker dari terapi embolisasi endovaskular selama kehamilan, tindak lanjut pascapersalinan dengan angiografi pengurangan digital (DSA) lanjutan direncanakan.

Simpulan: Kompleksitas pengelolaan multigravida dengan usia kehamilan 37 minggu dalam persalinan dengan fistula kavernosa karotis dan janin tunggal hidup dengan presentasi kepala, menekankan pentingnya pendekatan multidisiplin untuk luaran ibu yang optimal. Perlunya pendektanan multidispilin keilmuan memeberiksan hasil yanga baik pada ibu dan bayinya.

Kata kunci: Fistula Kavernosa Karotis, Angiografi Pengurangan Digital

Introduction

A carotid cavernous fistula (CCF) is an abnormal shunt from the carotid artery into the cavernous sinus. Symptoms of CCF depend on the involvement of important neural and vascular structures within the cavernous sinus. These structures include cranial nerves III (oculomotor nerve), IV (trochlear nerve), V1 (ophthalmic nerve), V2 (maxillary nerve), and VI (abducens nerve). CCF can be classified based on haemodynamic propertiesetiologi; aetiology, or shunt anatomy.[1] Haemodynamically, CCF is classified into high flow and low flow. Etiologically, it is divided into those caused by trauma and spontaneous lesions.

Spontaneous CCFs represent 30% of all CCFs and result from aneurysm rupture or genetic conditions that make patients susceptible to vascular injury such as Ehlers-Danl syndrome or fibromuscular dysplasia, which is most commonly seen in older adults women and low-flow, indirected type D fistulas.[2]

One important pathophysiology is increased intracavernous sinus pressure, resulting in damage to the fine vessels of the cavernous sinus, in the presence of certain underlying conditions. Clinical presentations include proptosis (81%), diplopia (68%), cranial bruit (49%), retro-orbital headache (34%), chemosis (87%), ocular motor paralysis, vision loss, and increased intraocular pressure (34%).[2,3]

Timing regarding the management of pregnant patients with CCF is individualized; definitive treatment before delivery should probably be considered if the complication occurs in the mid or early third trimester, although it may even lead to preterm labour.[3]

Case Report

A multigravida woman with a diagnosis of G2P1A0 and a 32-week-pregnant single fetus presented with swelling of the left eye. There was no complaint of abdominal pain radiating to the waist, no mucus blood discharge, and no water discharge. The patient had a history of swollen left eye since she was pregnant with her first child at the age of 6 months. Initial complaints of swollen eyes accompanied by headaches. Furthermore, the patient was advised to be controlled after giving birth. After giving birth to the first child, 1 month later the patient was examined by an ophthalmologist and was advised to perform DSA and embolization three times.

The patient said that he had undergone surgery with a neurosurgeon for DSA and embolization in August 2021 and April 2022, and DSA action plan in June 2022. The patient was then advised to control again in 3 months. While waiting, the patient became pregnant in May 2022.

History of marriage 1 time for 3 years. Menarche age 16 years, irregular, length 7 days, last menstrual period 10 April 2022. The patient has been giving birth once in 2021, 3000 grams assisted by gynecological obstetric specialists. Patients have been diagnosed with carotid cavernous fistula disease in August 26, 2020.

Physical examination revealed BP: 120/100 mmHg. Exophthalmus in the left eye. Ophthalmological physical examination the eye clinic on 12/12/2022 found superior palpebra: edema (+), conjunctiva: hyperemic (+), OS perimetry: OS: 21 mm.

Fundus uteri height 3 fingers below xypFundus uteri height 3 fingers below xyphoid processes (28 cm), elongated, left dorsal, under the head, U 5/5, his absent, FHR 146 /min, EFW 2635 g. Vaginal Toucher: Portio rubbery, posterior, OUE 1 multi, head, HI, amniotic fluid, and pointer could not be assessed. Laboratory examination on 12/12/22 found Hb 8.6 g/dL, erythrocytes 3.80 103/mm3, hematocrit 29%, MCV 75.3 fL, MCH 23 pg, and MCHC 30 g/dL. Urinalysis obtained 5-6 leukocytes. Laboratory examination

found Hb 8.7 g/dl, erythrocytes 3.93 103/mm3, hematocrit 29, MCV 73.8 fL, MCH 22 pg, MCHC 30 g/dl, fibrinogen 495.8, ferritin 19, and TIBC 11. Ultrasound examination, as shown in Figures 2 and 3, shows the patient's DSA.

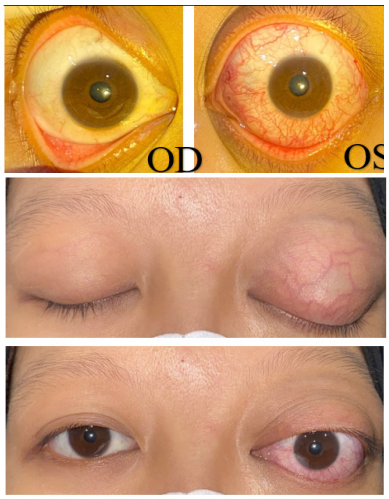

Figure 1 Proptosis of the Left Eye.

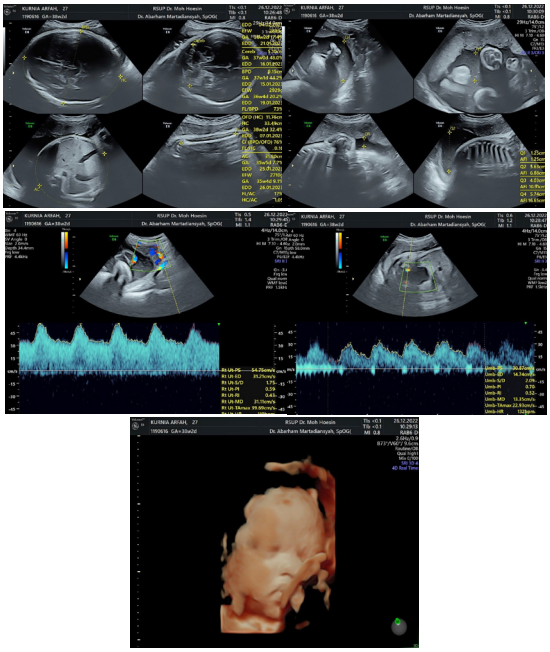

Figure 2 Ultrasound at 37 Weeks of Pregnancy Single Fetus Head Presentation

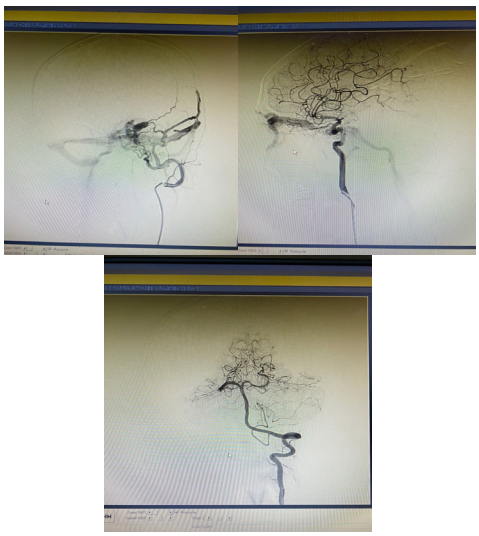

Figure 3 DSA and Embolization by Neurosurgery with Diagnosis of Carotid Cavernous Fistula

The patient was diagnosed with G2P1A0 pregnant 37 weeks with proptosis ec carotid cavernous fistula OS and moderate anemia iron deficiency single life fetus cephalic presentation. Prognosis of the mother dubia and fetus ad bona. Hospitalization management, PRC transfusion Hb >10 g/dl, preabdominal termination plan on December 29, 2022.

Discussion

During pregnancy, venous pressure increases and the effects of estrogen on blood vessels reach maximum levels in the third trimester. These changes increase the risk of the formation of new aneurysms or may worsen pre-existing aneurysms. Most arterial aneurysms that rupture in women under the age of 40 years are pregnancy-related. After aneurysm rupture, CCF occurs later and symptoms such as bruit, chemosis, and proptosis accompany ophthalmoplegia, which is the pressure effect of CCF.[4]

Clinical presentations include proptosis (81%), diplopia (68%), cranial bruit (49%), retroorbital headache (34%),

chemosis (87%), ocular motor paralysis, vision loss, and increased intraocular pressure (34%). The patient had proptosis, diplopia, retroorbital headache, increased intraocular pressure, and a pulsating mass. These findings described the incidence of CCF which was supported by the orbital CT scan. Headache is a common complaint among pregnant women and there were no associated neuro-ophthalmic symptoms or history of head injury. Thus, conditions such as CCF are often not suspected. Second, even if the diagnosis is made at 30 week of gestationg, management will remain the same regardless of closer monitoring of symptoms. In theory, symptoms may be intensified during pregnancy because of the associated haemodynamic and hormonal changes, resulting in vascular engorgement. However, in this patient with no history of head injury and who was previous asymptomatic, the onset of CCF was most likely de novo.[3]

Timing regarding the management of pregnant patients with CCF is individualized; definitive treatment before delivery should probably be considered if the complication occurs in the mid or early third trimester, although it may even lead to preterm delivery. Spontaneous CCF complicating pregnancy was successfully treated with craniotomy and surgical occlusion of the internal carotid artery during the third trimester, but the pregnant woman had to deliver prematurely.[3]

Endovascular embolization therapy for the treatment cerebral artery aneurysm CCF during pregnancy has not been reported. Concerns often expressed are the potential for fetal abnormalities and the development of childhood cancer due to radiation exposure. The management plan for the patient based on the literature recommended termination to deliver the patient preferably at 37 weeks gestation, as fetal prematurity was not a problem, and termination of pregnancy could facilitate maternal management without fetal irradiation during endovascular imaging and embolization. Regarding the mode of delivery, there are no previous reports or studies examining the safety of vaginal delivery. However, a cesarean section is recommended to avoid the Valsalva maneuver which may cause a further increase in intraocular pressure and may lead to vision loss. Labour pain may also increase arterial pressure and the risk of CCF rupture during vaginal delivery.[3]

Endovascular embolization was indicated because the patient had intolerable headaches and bruiting even weeks after delivery. Indications for the treatment of spontaneous CCF are as follows: (1) decreased vision; (2) obstructive diplopia associated with vascular swelling and enlargement of extraocular muscles or nerve compression within the cavernous sinus; (3) intolerable bruit or headache; and (4) malignant proptosis with untreatable corneal exposure. In general, the goals of endovascular embolization are to occlude the retrograde drainage channels to the ophthalmic veins that cause ocular symptoms; occlude the retrograde drainage channels to the superficial media cerebral veins that may cause hypertension and cerebral venous hemorrhage; and obliterate the dural CCF.[3]

Based on several journals, it is said that there is no relationship between the mother's history of carotid cavernous fistula and the fetus. However, endovascular embolization therapy to treat cerebral artery aneurysm CCF during pregnancy is of concern because of potential fetal abnormalities and development of childhood cancer due to radiation exposure. Thus, in this patient, follow-up DSA was also planned after delivery.

Serious complications such as visual impairment affect 60-90% of patients and may develop if left untreated. Total vision loss occurs in 25% of patients but may be reversible. Cerebral hemorrhage may occur in 30-40% of patients because of direct drainage into the cerebral veins. Spontaneous

intracranial hemorrhage from occult CCFs in pregnancy and puerperium has been reported, and cerebral angiographic examination should be performed to confirm the actual loss of CCFs.3

Carotid cavernous fistula has no adverse effect on pregnancy, but potentially serious maternal complications such as intracranial haemorrhage can lead to morbidity or even fetal and maternal death.3 Based on the orbital CT scan examination and after DSA and embolization of the sinistra ECA, it was found that there was no retrograde flow into the cerebral veins in the patient and the blood flow mainly refluxed back to the ophthalmic and facial venous systems, reasoning that the risk of intracranial haemorrhage was minimal.

Spontaneous resolution is known in 5-60% of cases. A case of marked regression of CCF 2-3 days after delivery and it is speculated that spontaneous improvement of CCF after pregnancy is due to thrombosis related to changes in blood coagulation that occur during pregnancy and delivery.3,5 However, based on our patient's findings, it did not resolve spontaneously and the symptoms worsened until the time of presentation of the second pregnancy.

Conclusion

Carotid cavernous fistula (CCF) is an abnormal shunt from the carotid artery into the cavernous sinus, with symptoms depending on neural and vascular involvement. The decision to terminate pregnancy is influenced by increasing gestational age and should be promptly executed while keeping the patient under observation. Ethical, legal, medical, and religious considerations should guide the evaluation of terminating pregnancies in mothers with a history of CCF. Postnatally, infants born to mothers with a history of CCF can be managed conservatively.

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