

A rare case of an ophthalmologic emergency: idiopathic central retinal artery occlusion

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Abstract:

Central retinal artery occlusion is a form of an acute stroke in the eye and is regarded as an ophthalmic emergency.^[1,2] This paper presents a case of Central retinal artery occlusion in an 18-year-old pregnant female who presented with sudden loss of vision in her right eye and was diagnosed with central retinal artery occlusion. The aim of this paper is to illustrate the clinical presentation, diagnosis and treatment but mainly focusing on the underlying etiologies.

Recent literature shows no reported cases explaining the possibility of retinal artery occlusion occurring in a young pregnant female who is otherwise free of any medical conditions.^[3] Due to the serious consequence, limited knowledge and treatment in the field of unexplained retinal occlusion in pregnancy; it's encouraged that more efforts are invested in research and reporting cases for a better understanding of the possible and future management options and preventative strategies of central retinal occlusions.

Keywords: central retinal artery occlusion, sudden painless loss of vision, eye stroke, ophthalmic emergency, retina, pregnancy. (Word count 2,540)

Introduction

The purpose of this case report is to discuss the rare occurrence of central retinal artery occlusion in a young female who suddenly experienced a painless loss of vision in her right eye.^[1]

Loss of vision may occur in one or both eyes either involving the loss of all or part of the visual fields. Loss is described as sudden when it develops within a few minutes to a couple of days. In general; there are three main causes of sudden loss of vision and those causes include Clouding of normally transparent eye structures, abnormalities of the retina or abnormalities of the nerves that carry the visual signals from the eye to the brain. In this case, the patient is found to have abnormalities with the retina and is no longer able to sense the light rays normally due to blockage of the central retinal artery. A diagnosis of central retinal artery occlusion was made in her case.^[1]

Central retinal artery occlusion (CRAO) is equivalent to an acute stroke of the eye and patients present with acute, painless loss of monocular vision. It's the result of a blockage in the main artery, which supplies the retina of the eyes. The central retinal artery arises from the ophthalmic artery itself a branch of the internal carotid artery. The estimated incidence of CRAO is 1 in 100000 people and accounts for 1 in 10000 ophthalmological outpatient visits and is more common in males than it is in females. This abnormality is most frequently encountered in older adults in their early sixties. Less commonly seen in individuals younger than 30. This medical condition is regarded as an

ophthalmic emergency because of its serious complications which include Glaucoma - Increased intraocular pressure of the eye-, partial or complete loss of vision in the affected eye and Stroke.^{[2][3]}

Blocked retinal arteries may be the result of a blood clot or fat deposits that get stuck in the arteries, which are more likely if there is hardening of the arteries -atherosclerosis- in the eye. Moreover, clots may dislodge from other parts of the body commonly the heart and the carotid artery of neck; and travel to the retina blocking the retinal artery or other branches.

Eventually; the retina will not receive enough blood and oxygen leading to ischemia of the retina and subsequently loss of part of vision.^[4]

There is no apparent tendency for one eye to be affected compared to the other but if bilateral involvement is present (1-2% of the cases); heart valve disease, giant cell arteritis (temporal arteritis), and other vascular inflammations should be strongly considered.^[5]

In a prospective study which aimed to investigate whether unilateral vision loss reduced any aspects of quality of life in comparison with normal vision; results showed that 260 eyes with CRAO suffer profound monocular visual loss, with 80% of patients having a visual acuity (VA) of 20/400 or worse.^[6] Clinical approach and management is similar to that of a stroke and investigations are done to prevent the occurrence of further vascular events. However; No definitive helpful treatment is available and the outlook for

restoring the vision in the affected eye is poor when treatment is delayed.^[7]

Case Report

This is the case of an 18-year-old Bahraini pregnant female (F.H.M) who presented to the Accident and Emergency department (A&E) in Qatar (while she was abroad for a visit) after experiencing sudden painless loss of vision in her right eye while praying one afternoon. She was diagnosed with a stroke, which happened a few hours-days prior her visit to the A&E. A week after the incident; she returned to the country and was in the A&E department in Bahrain Defense Force Hospital (BDF) on January 7th 2014 – two weeks after the sudden loss of vision - complaining of a severe headache as she waited for her ophthalmology appointment in the Ophthalmology and Laser clinic in BDF.

The headache was localized to the right side of her head and it didn't radiate to the neck or elsewhere; it was sudden and sharp. With regards to her eye; she described her loss of vision as being painless and she has loss of function of her right eye. "I can only see a blank black image," she says. She had no similar headaches associated with the loss of her vision. It was completely painless and this was her first headache since her medical emergency. At the time of her visit; she was 28 weeks pregnant but had no known medical conditions otherwise and has no history of abortions or miscarriages, diabetes, hypertension, heart abnormalities, carotid artery disease, temporal arteritis or Intravenous drug abuse.^[4] She is not known to have any allergies to either medications or food.

She has a history of a previous Road Traffic Accident, a year and four months back. The RTA resulted in fractures of the Jaw and the femur requiring surgical intervention and fixation. She is not known to have any significant past medical history. Both her parents are alive and well and they have no history of diabetes mellitus (DM), high blood pressure, and atherosclerosis or any heart disease. No member of her family had experienced a similar condition or any form of stroke. Regarding her social history, she does not smoke nor consume alcohol. She is married, and has no children, as this is her first pregnancy. She is a housewife with a high school degree.

Upon examination, the patient was vitally stable and had Glasgow coma scale (GSC) of 15. Her Cranial nerves were intact with no cerebellar signs. She had normal reflexes except for the pupillary light reflex (PLR) of the right eye. Noticeably; the consensual light reflex of the right eye was present. Her visual acuity was also decreased.

Diagnostic assessments in the A&E were done to find the possible causes of both the sudden loss of vision and headache. Possible differential diagnosis included Central

retinal artery or vein occlusion, Vitreous hemorrhage, Giant cell (temporal) arteritis, Ischemic optic neuropathy, Macular hemorrhage, Ocular migraine, Radiation retinopathy, Detachment of the retina and Strokes or transient ischemic attack.^[1]

Urgent Investigations include Fasting blood sugar, echocardiogram ECHO (mainly to exclude patent foramen ovale or venous thrombus), Magnetic resonance imaging MRI, and ophthalmologic investigation including fundoscopy, fluorescein angiography and OCT. additionally; vasculitic screen and thrombophilia screen were ordered. The patient was given follow up appointments with the obstetrics and gynecologist, ophthalmologist, and neurologist.^[8] Ophthalmological investigations with exception to the fluorescein angiography which was unsafe for her pregnancy; confirmed central retinal artery occlusion (CRAO) and was immediately given Enoxaparin sodium 80 mg.^[9] Enoxaparin sodium is a drug used to treat clots and to prevent further clots from happening. Also; the patient was admitted to the obstetrics and gynecology ward for further assessment since she is suspected to be in a hypercoagulable state, or may have a case of vasculitis as the patient is younger than 50 years of age and has no vascular risk factors which may have caused CRAO. Initially; the patient was managed with steroids (Prednisolone 40 mg) and supplements (Calcium and vitamin D). In addition; multidisciplinary team follow up was arranged.

Discussion

Central retinal artery occlusion is an ophthalmic emergency thus it is essential to find the underlying cause to prevent further similar events from happening. Proper diagnostic investigations based on adequate knowledge are required for clinical suspicion and making a diagnosis.

Referring to the case of F.H.M; since the patient belongs to the younger category and has no identifiable risk factors; the investigations recommended in her case included: fluorescein angiography, vasculitis screen including anticardiolipin antibodies, antinuclear antibodies, anti-double stranded DNA antibodies, routine coagulation tests (prothrombin time PT, partial thromboplastin time PTT), specialized clotting factor and platelet activity studies (levels of protein S, protein C, antithrombin III, plasminogen activator, plasminogen activator inhibitor, fibrinogen and resistance to activated protein C) and homocysteine.^[8] However; fluorescein angiography was contraindicated in her case because as mentioned the patient was 7 months pregnant and the gynecologist questioned the safety of the test. Overseas results including Antineutrophil cytoplasmic antibodies ANCA, factor V Leiden mutations, and Anti-cardiolipin antibodies ACA tested to be negative which excludes thrombophilia, antiphospholipid syndrome

APS and vasculitis as possible etiologies of the CRAO. Thus so far; No evidence of any systemic disorder was revealed on history or in the laboratory investigations.

Based on the current literature; it's agreed that Retinal artery occlusions before the age of 30 are rare and patients usually have a detectable etiology.^[10] Patients often have abnormalities associated with their sudden loss of vision which may help determine the causative mechanism of the retinal occlusion. Such associations may include Systemic arterial hypertension HTN, diabetes mellitus DM, cardiac valvular disease and carotid atherosclerosis mainly in the form of stenosis or plaques.^[5] Major possible causes of this patient's CRAO have been ruled out. This leaves the doctors with the same question they started with: what might have caused the occlusion in the first place? For this reason along with her pregnancy; her medical case is considered both complex and distinct from other cases of retinal occlusions.

Retinal diseases may be associated with pregnancy; either uniquely occurring or generally worsening. Alterations in the body during pregnancy could be the result of hormonal, hematological, cardiovascular and immunological changes that occur during this period. A study revealed that the most common retinal change that occurs in pregnancy is diabetic retinopathy DR which is related to risk factors including uncontrolled blood sugar or blood pressure prior to pregnancy. On the other hand; the same study states that other retinal and chorioretinal disorders may arise including retinal vascular occlusion.^[5]

Based on a study by Rumelt & Brown (2003); it was suggested that the majority of retinal occlusions are either thrombotic or embolic in their nature. In 20% of the patients; an embolus is visible either with central or branch retinal artery occlusion. Retinal occlusions in the eye are almost always the result of a microembolism, which comes from a plaque(s) present with significant carotid artery stenosis. The carotid artery branches to give the ophthalmic artery, which in turn gives the retinal arteries supplying the retina. But; microembolisms from plaques are not always associated with carotid artery stenosis. Thus; the absence of the latter doesn't exclude the carotid artery as the source of the microembolism.^[11] Other differentials which could explain the case of F.H.M; undetected cardiac-valvular heart disease, arterial spasms, Purtscher's disease, Oral contraceptives or simply due to Pregnancy.

In March 2008, a case report done by Y. Chung et al was published by Korean Journal of Ophthalmology. They reported a case of retinal artery occlusion but it was a branch occlusion rather than central. Nevertheless; the main result was BRAO can occur in healthy patients without any systemic or ocular disorders, despite an extensive evaluation.^[12] The previous may suggest that retinal

occlusions may indeed occur in healthy young patients with no exact etiology determined.

Regarding the treatment of Central retinal artery occlusion, there is no specific treatment, which is effective, but some of the standard treatments used in the past include: Carbogen inhalation, acetazolamide infusion, ocular massage and paracentesis, as well as vasodilators like intravenous glyceryl trinitrate. Additionally; a few studies demonstrated the successful use of thrombolytic agents in treating CRAO which can either be delivered intravenously or intra-arterially by direct catheterization of the ophthalmic artery. However; thrombolytic therapy was not proven successful if treatment is delayed beyond 6 hours after the onset of the symptoms. Occlusion generally leads to permanent blindness after 105 minutes thus time is essential for management of CRAO and reperfusion must be attempted as soon as possible as well as early secondary prevention.^{[13][14]}

In our case of F.H.M; reperfusion was not attempted as her condition was identified hours-days after the event thus early secondary prevention remains the preferable treatment for her case to prevent further vascular events. Our patient is currently treated with steroids Prednisolone 40 mg, which is supported by several studies in CRAO and its effectiveness when administered as soon as possible.^[14] An angiographic study looking at the effect of high dose steroid on central retinal artery occlusion showed that steroids could be suitable for emergency treatment and doesn't interfere with other drugs that could be given.^[14] The safety of steroids is not questionable in our case but there is no documented evidence yet of the immediate effect of steroids on the vessel wall.^{[14][14]}

This case report illustrates the possibility of having central retinal artery occlusion CRAO in a very young female during pregnancy with no exact etiology yet determined subsequently the challenges that reside when attempting to treat or prevent CRAO in this case.

Conclusion

To conclude; this is a case of an 18 year female with sudden unilateral loss of vision. The case demonstrated the rare occurrence of the clinical scenario, and the challenges involved in identifying the underlying etiologies as no identifiable cause had been found.

Central retinal artery occlusion is a challenging ophthalmological emergency requiring a focused diagnostic workup to identify underlying etiology to institute appropriate secondary prevention and reduce risk of vascular events in the future. In cases of uncertain etiology, management with antiplatelet agents is recommended if not contraindicated. The need for further research in this field is

of no doubt required as well as the need for more case reporting if and when such cases are encountered.

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