

## Permanent unilateral blindness associated with peripartum cardiomyopathy

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### Abstract

Peripartum Cardiomyopathy (PPCM) is a rare and ominous disease manifested in the peripartum period of women with complications concerning different organic systems. We describe a case of peripartum cardiomyopathy which was complicated with acute permanent unilateral blindness, further documented embolic retinal artery occlusion as a consequence of PPCM. This is a quite unusual event with PPCM, since to our knowledge there is no previously reported case with PPCM as the sole associated factor. The purpose of our paper is to emphasize that thromboembolism of the central retinal artery, seems to be compatible with peripartum cardiomyopathy and that, peripartum blindness, may be attributed to the cardiomyopathy diagnosis. Hippokratia 2009; 13 (1): 58-60

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Peripartum cardiomyopathy (PPCM) is mainly a dilated disease of the cardiac muscle, with failure of left ventricular function. In general, central retinal artery occlusion with loss of vision in one eye is a disease of the old age associated with atherosclerotic plaques. In younger patients though, valvular heart disease could be a common precursor. Visual disturbances in women, have been documented infrequently and always in the simultaneous presence of one or more of its known associated medical factors or toxemia during pregnancy. After thorough research in the available English-language medical literature, a case report like the present one has not been noted. We believe that this is the first reported case of permanent blindness in one eye, abreast with PPCM.

### Case Report

In February 2005, a 35 years old primigravida woman, with twin pregnancy after In Vitro Fertilization (IVF), delivered by an elective cesarean section in Iaso Maternity (Athens). The prenatal course was uneventful and all routine laboratory investigations were within normal limits. The blood pressure during pregnancy ranged from 115/70 mmHg to 135/80 mmHg and the weight gained was 15 kg. No signs or symptoms of headache migraines or thrombophlebitis had been recognized and only mild edema in lower extremities was present to a certain extent. No signs of systemic disease or hypercoagulable problems were noticed. The patient's past medical history was

insignificant as was her family's. The electrocardiogram showed a sinoatrial rhythm. The mother completed a successful postoperative course and left the maternity ward on the fifth day.

On 10<sup>th</sup> postoperative day, the patient complained about progressing dyspnea, weakness, and tendency for vomiting. Three days later, the symptoms deteriorated and the cardiologist who saw the patient considered the diagnosis of peripartum cardiomyopathy confirmed by echocardiograph. It demonstrated a left ventricular ejection fraction (LVEF) of 34% (normal limits >45%) with end-diastolic diameter of 63.3 cm (normal limits: 35-57 cm).

Immediate hospitalization was advised. On the same evening, she suddenly developed blurring of vision in the right eye rapidly progressing to visual loss without prodromal symptoms. Immediate funduscopic examination revealed confused perception of light reflexes, fixed pupil in mid-dilatation and a cherry-red staining at the site of the fovea. The retina was pallid and edematous with no abnormality, hemorrhage or detachment and its artery was estimated as thin red and faded line. The optic disc was not well outlined. The left eye was normal. Patient's blood pressure was 135/80 mmHg. He suggested that this instance of blindness was a complication of the peripartum cardiomyopathy, manifested postpartum. Shortly after, the patient was admitted to Laiko General Hospital of Athens for observation. The examinations performed by the cardiologist and ophthalmologist confirmed to the first diagno-

sis. No further testing for blindness took place. Treatment started, and was focused on peripartum cardiomyopathy which is similar to medical therapy for other forms of dilated cardiomyopathy.

Patient's laboratory results were as follows: A full blood count and serum creatinine and liver function tests were normal, as was the Wasserman test. Uric acid, cholesterol,  $\gamma$ -GT and LDH were mildly increased. Rheumatological and blood coagulation reports were within normal limits; thus excluded as predisposing factors of mural thrombus and emboli. A chest x ray showed mild cardiomegaly.

The patient had a good recovery, with symptoms of cardiac dysfunction almost eliminated. However blindness has remained with slightly improved vision by turning the eyeball of the affected eye to the right corner.

One year later vision loss is still sustained, as it was when the patient left the hospital, without any further sequel. In the meantime the symptoms of peripartum cardiomyopathy have disappeared (LVEF=55%). We believe that the prognosis for returning vision is very poor, since it has for so long persisted.

### Discussion

In the past, retinal abnormalities such as edema, vascular lesions and detachment were to be considered the origin of blindness in most cases. Later it was emphasized that toxemia associated with cortical disturbances is perhaps the rule. In 1995, Cunningham et al<sup>1</sup> experienced blindness in almost 15% of eclamptic women. He presented 15 toxemic patients with blindness; 13 had pure cortical blindness and in two instances detachment of the retina was observed. In each case, vision came back in four hours to eight days. Studies in radiology techniques enhance the reversibility of the neuroimaging figures<sup>2</sup>. They support that blindness returns to normal in almost all patients, although some cases of blindness lasting more than two months have been described.

National Institute of Health Office of Rare Diseases defines peripartum cardiomyopathy as cardiac failure of unknown origin occurring within the final month of pregnancy or within 5 months of delivery, with no known heart disease before the final month of pregnancy. This is best confirmed by echocardiography showing left ventricular systolic dysfunction and frequently intracardial mural thrombi<sup>3,4</sup>. The latter accounts for systemic embolism, mainly in the lungs. Thus, peripartum cardiomyopathy could be recognized as a cause for obstruction of the retinal artery. Although it is considered as an idiopathic entity, myocarditis has been found in 8% to 29% of cases.

Occlusion of the retinal artery attributed to embolism is accompanied by sudden, painless, and deep blindness. If a vasospasm is the cause, this may be delayed for a short time. In such a case, premonitory signs of visual disturbances occur before vision loss, as in case of retinal vein thrombosis too.

The fundoscopic examination of the retina presents

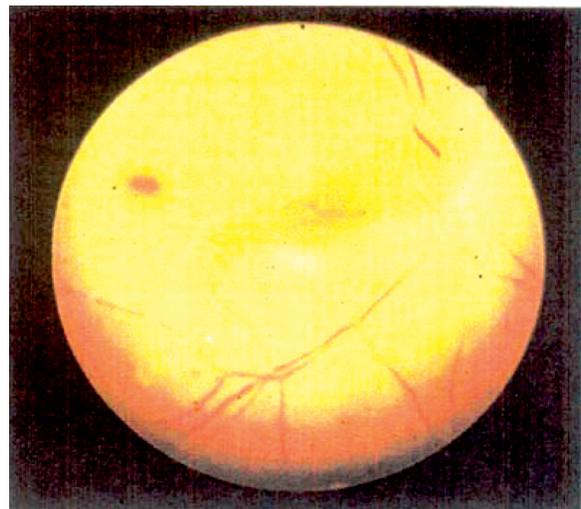
the characteristic cherry-red staining at the fovea, invested by a pale and edematous fundus (Figure 1). The report given by the two ophthalmologists has similarities with the picture of central retinal artery thrombosis-induced closure. The fovea remains red and feeding since it does not have the interior retinal layers, which are fed by the retinal artery. The exterior retinal layers are fed by the choroid arteries and thus are unharmed. The retinal arteries are revealed as thin red-faded lines. The embolus may be lodged in the central retinal artery, one or more of its branches or the cilioretinal artery and in some instances it is visible. The occlusion could be complete or incomplete. In the latter situation, vision alterations in the background will be restricted in the field nourished by the closed branches. Central vision will be retained, but there will be a sector-shaped deficit in the area.

An embolus making an obstruction is less common than a thrombus, the latter being the result of an underlying disorder such as an endarteritis in atherosclerosis.

With reference to visual prognosis, embolus dislocation and spasm's relief within one hour may reinstate all vision. However, the site and percentage of obstruction, the aetia and the lasting-time are significant.

Dahring<sup>5</sup>, based on histopathologic findings, said that in cases of retinal severe lesions within 20 minutes of occlusion, return of vision will not always be successful. When retinal artery obstruction happens the whole retina is affected; in cases where a cilioretinal branch from the choroid circulation is preserved, the retina between the macula and the optic disc will be functional.

Treatment methods of obstructed blindness have been described with varying results and include: digital massage of the eyeball in order to drive the plug into smaller branches, decompression with paracentesis of the anteri-



**Figure 1:** Occlusion of the central retinal artery. Look out the cherry-red staining at the fovea and the pale edematous fundus. (Theodosiadis G. Manual Ophthalmology. Med Editions, Litsas, Athens, 1996; with permission).

or chamber, retrobulbar injection of vasodilators and xylocaine, and inhalation of 95% O<sub>2</sub> and 5% CO<sub>2</sub>.<sup>6</sup>

To our knowledge, this case report may be the first one described with central retinal artery occlusion - induced permanent unilateral blindness associated with peripartum cardiomyopathy. So, in general, when visual impairment develops in pregnancy, searching for a coexisting condition predisposing to retinal artery obstruction is always useful.

Brown et al<sup>7</sup> presented two pregnant women, both with a history of migraine headaches. The one also had elevated factor VIII activity. These two patients suffered a retinal branch and cilioretinal artery occlusion, not of the central retinal artery alone, and were in the first trimester of pregnancy.

Brown et al<sup>8</sup>, even reported one pregnant woman of 18 years of age with cilioretinal artery occlusion and atherosclerotic cardiovascular lesions.

Concerning the primary mechanism of obstruction in our case, a small intracardial mural thrombus or fragment metastasised from the left ventricular chamber to the retinal artery by way of the arterial system. The presence of peripartum cardiomyopathy and acute blindness eliminate the possibility that a process within the retinal artery or the retinal vein is the cause and therefore reinforces that embolism from a mural thrombus is the etiology. It is noteworthy that embolus was not seen by two separate echocardiographers and the possibility of missing something like this is a question. The patient had no change in her mental status and the neurological exam was normal.

In case of an early vision recovery, the blocking has probably been induced by a spasm of the retinal artery wall. Also women with true cortical blindness, usually have normal fundusoscopic findings. Unfortunately blind-

ness in this case has insisted since the patient left the hospital. Scarcely improved vision of the affected eye may be attributed to edema of the retina, which subsided by the end of the patient hospital care. Nevertheless we cannot totally exclude the possibility of a nearly complete occlusion.

Although the condition is likely to be exceedingly rare, it should be considered by anyone evaluating a patient with PPCM. Early recognition can lead to a more effective treatment and limit the extent of the disturbances of vision.

In conclusion, this report is a preliminary experience, which supports that permanent blindness after occlusion of the central retinal artery may be expected and is encountered in peripartum cardiomyopathy.

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