Cilioretinal artery occlusion in a previously healthy adolescent diagnosed with infective endocarditis

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Abstract

Retinal arterial occlusions are rarely seen among the young[1] and out of all types of retinal arterial occlusions, the cilioretinal artery involvement is uncommon as the incidence of cilio-retinal artery is 6.9% to 49.5% in population[2]. Hence when a retinal arterial occlusion is seen in a young patient, the cause for it is to be investigated. This case report focuses on an instance where an embolic phenomenon caused by infective endocarditis leading to retinal arterial occlusion.

Key words: infective endocarditis, cilio-retnal artery occlusion

Case report

A 14 year old who was treated for Dengue fever initially in December 2017 at the National Hospital, Kandy, later developed low grade fever and cough nearly a month later. During this period she has developed multiple painless red patches over the plantar surface of the feet, which were later identified as Janeway lesions. Upon investigation the blood cultures were positive for Methicillin-Susceptible Staphylococcus aureus and the transesophageal echocardiogram has revealed large friable, highly mobile vegetation attached to the base of posterior mitral valve leaflet of 27 mm into 8 mm size. Another vegetation of 11 mm into 3 mm size at the septal leaflet base of the tricuspid valve along with Grade I mitral regurgitation and mild tricuspid regurgitation was seen apart from that. With the above findings she was diagnosed with infective endocarditis according to the modified Duke's criteria[3] and was started on intravenous antibiotics and planned for vegetectomy and valve repair. While the patient was waiting for surgical management, she developed a sudden onset visual blurring in the left eye. On assessment she had reduction of the central vision in left eye and the visual acuity was 20/40, while the right eye was normal. The dilated fundoscopic examination revealed that the patient had left sided cilio-retinal artery occlusion with the evidence of two Roth spots. Spectral Domain Optical Coherence Tomography images revealed increased macular thickness, with increased reflectivity in inner retinal layers and reduced reflectivity in outer retinal layers. Upon diagnosing left cilio-retinal artery occlusion, ocular massage was performed and she was started on systemic carbonic anhydrase inhibitors to increase the retinal perfusion pressure. After ocular massage along with acetazolamide, the intraocular pressure was 6 mmHg and for IE she was treated with intravenous antibiotics and underwent vegetectomy and was on follow up pertaining to visual recovery. With time her vision improved to 20/20 and the Fundus Fluorescein Angiography (FFA) performed 6 months after the onset of symptoms showed recanalization of the previously blocked cilio-retinal artery. Fundus assessment after 6 months showed complete resolution of macular oedema and the visual field was improved.

Janeway lesions involving the soles.

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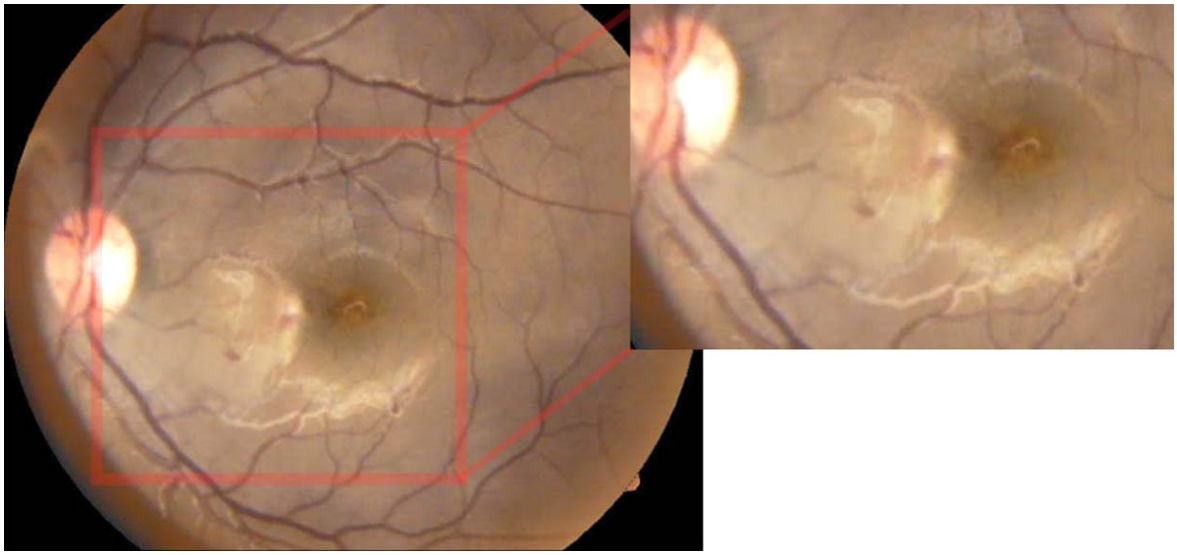
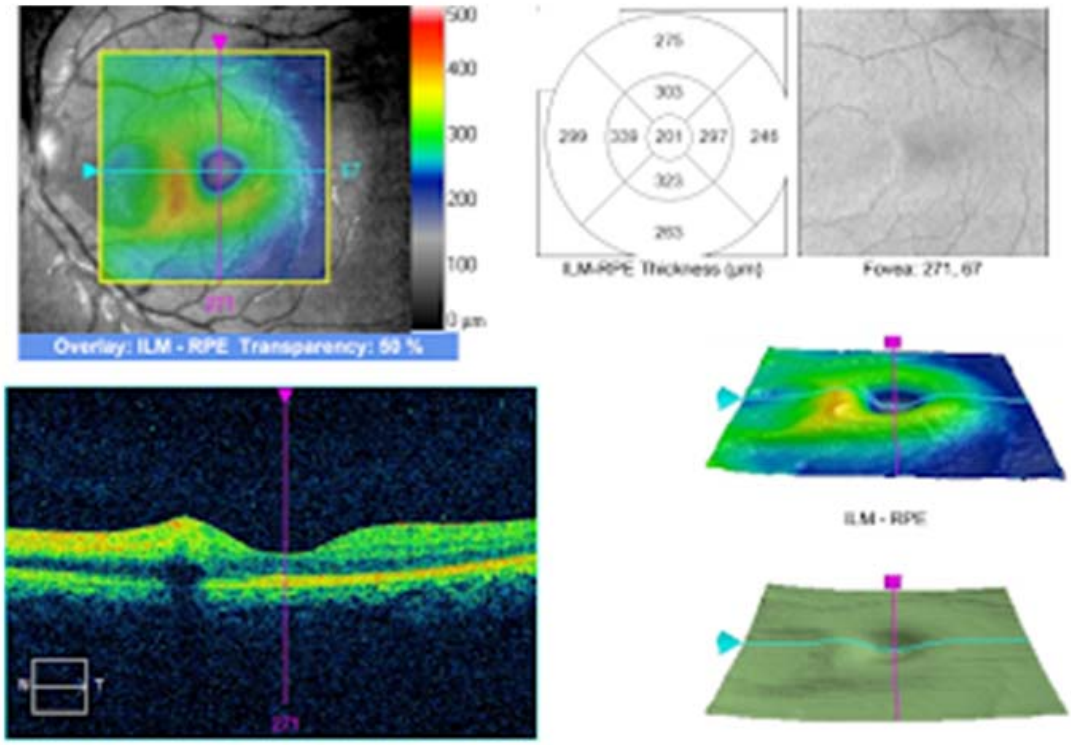


Fundal photograph – Cilio-retinal artery occlusion with two Roth spots.



SD OCT - increased inner retinal thickness.

Resolution of retinal oedema and recanalization of the previously blocked cilio-retinal artery in FFA performed after 6 months.

Discussion

Retinal artery occlusion is an ophthalmological emergency and it is due to obstruction of the retinal blood flow, due to an embolus causing occlusion or thrombus formation, vasculitis causing retinal vasculature inflammation, traumatic vessel wall damage, or vascular spasm. It can involve any artery that supplies the eye. The main artery that supplies the eye and surrounding structures is the ophthalmic artery. The first branch of the ophthalmic artery is the central retinal artery, and it supplies nerve fibers in the optic nerve as well as the inner layer of the retina. After entering the eye, the central retinal artery divides into superior and inferior branches. In addition to these, the cilio-retinal artery is a branch of the short posterior ciliary arteries, which is a separate branch of the ophthalmic artery[4]. The incidence of cilio-retinal artery is 6.9% to 49.5% in population and it can get occluded resulting in painless reduction of central vision[2]. Retinal artery occlusions are rare under 30 years of age[1]. Embolism is considered the commonest cause of retinal artery occlusion, usually from the carotid arteries, while the heart is the source of emboli in <10% of instances[5]. Considering the type of emboli 74% of them are made of cholesterol, 15.5% of platelet fibrin and 10.5% of calcified material. Infectious emboli from vegetations in infective endocarditis is another cause of retinal arterial occlusion.

Infective endocarditis (IE) is a relatively rare, life-threatening disease, and among survivors it can lead to long-lasting health effects[6]. Once established, IE can involve almost any organ system in the body. IE has an incidence of 1.4 to 6.2 per 100,000 in the lite-rature[7] and causative organisms are commonly Staphylococci (42%), as in this presentation, nearly 40% are caused by Streptococci, and an 8% of cases remain culture negative[8]. IE particularly affects those who have underlying structural heart disease, prosthetic valves and previous history of IE, intravenous drug use and hemodialysis increases the risk[9]. In the setting of bacteraemia with a pathogenic organism, an infected vegetation may form as the end result of complex interactions between invading microorganisms and the host immune system[6]. Cardiac complications of IE include, heart failure, perivalvular abscess, pericarditis, fistulous intracardiac connections and valve insufficiency and systemic embolization takes place in 22-50% of patients. Nearly a 65% of embolic events show the involvement of the central nervous system and the other common sites of systemic embolization are major arterial beds including lungs, coronary arteries, spleen bowel and extremities. Nearly 90% of the central nervous system emboli lodge in the distribution of the middle cerebral artery[10]. Embolic complications are more often when there is involvement of the aortic and the mitral valves with the infection and usually during the first 2 to 4 weeks after initiating antimicrobial therapy[11]. The rate of first embolic events diminishes with time, falling from 13 per 1000 patient-days during the first week of therapy to less than 1.2 per 1000 patient-days after completion of the second week of the therapy[12]. The risk of embolization seems to increase with increasing vegetation size, especially in mitral valve endocarditis with Staphylococcal infection[13].

In retinal artery occlusions, still there is no particular therapy which has been proven based on evidence to show efficacy in improving visual outcomes. A meta-study has suggested that some therapies are better avoided as they may be worse than the natural course[14]. The European Assessment Group for Lysis in the Eye Study (EAGLE study) evaluated clot busting tissue plasminogen activator (tPA) against a placebo. Yet the study revealed that there is a significant risk of symptomatic intracranial haemorrhage without evidence of visual benefit, with the use of tPA, and the study was terminated early[15]. Although a recent meta-analysis of observation studies suggest that there may be mild benefit to intravenous tPA when given within 4.5 hours, yet they also reported several fatalities associated with tPA. It also suggested that the conservative treatments such as ocular massage, paracentesis and hemodilution led to worse outcomes[14]. Ocular massage is a conservative therapy that may theoretically cause the embolus to move more distally and to reduce the area of ischemia[16]. Anterior chamber paracentesis may be performed by removing 0.1-0.4 ml of aqueous humor from the anterior chamber using a small gauge needle. Theoretically this lowers the intraocular pressure and may allow the embolus to move further down the vessel[17]. In addition to this the intraocular pressure can be reduced medically. Increasing carbon dioxide concentration is also proposed to induce vasodilation[18].

Visual field losses induced by retinal arterial occlusions are usually permanent, yet in branch retinal occlusions including cilio-retinal artery occlusions the visual acuity may recover to 20/40 or better in 80% of eyes.

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