

Unilateral Central Retinal Artery Occlusion in Eisenmenger Syndrome

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Abstract

Background: To report a case of unilateral central retinal artery occlusion in a patient with Eisenmenger syndrome.

Methods: Full ophthalmic examination, physical examination and laboratory examination were performed.

Results: Ophthalmic examination revealed unilateral central retinal artery occlusion. Laboratory examination revealed hemoglobin 22.3 g/dl, hematocrit 65.9%, and pO₂ 42 mmHg.

Conclusion: The unilateral central retinal artery occlusion in a patient with Eisenmenger syndrome is rarely reported in the literatures.

Keywords: Eisenmenger syndrome, Central retinal artery occlusion

Introduction

Eisenmenger syndrome is defined by pulmonary hypertension accompanied by a reversed or bidirectional shunt through a large intracardiac defect [1,2].

Patients with Eisenmenger syndrome have congenital cardiac defects (most commonly, atrial septal defect, ventricular septal defect, or patent ductus arteriosus) with left-to-right shunting of blood. The resultant increased pulmonary artery pressure creates pulmonary vascular fibrosis and occlusion of pulmonary capillary beds. This increased resistance eventually reverses the intracardiac shunting of blood from left-to-right to right-to-left, thus bypassing the pulmonary circulation. As blood continues to bypass the lungs, the patient becomes hypoxic and develops erythrocytosis and resultant hyperviscosity.

We treated a patient with the Eisenmenger syndrome associated with an unoperated on congenital ventricular septal defect who developed unilateral central retinal artery occlusion.

Material and Methods

A 24-year-old Asian boy with Eisenmenger syndrome secondary to a large congenital ventricular septal defect presented with a 2-hour history of decreased vision in right eye. Medical history was notable for prominent pulmonary hypertension and severe shortness of breath, but no symptomatic coronary artery disease, dizziness, or syncope. The patient was oxygen dependent and unable to ambulate secondary to dyspnea. The patient developed secondary erythrocytosis with hematocrit levels between 56% and 65.9%.

Results

Ophthalmic examination revealed corrected visual acuity of R.E.:20/200 and L.E.:20/20, normal intraocular pressure, no evidence of iris neovascularization. Funduscopy demonstrated retina edema and cherry-red spot in right eye (Figure 1) and retinal vessels dialation and tortuosity, small blot retinal hemorrhages in left eye (Figure 2). Fluorescein angiography showed delayed artery filling and late leakage in the macula of the right eye.

Physical examination revealed a harsh III/VI systolic murmur. Extremities showed mild clubbing. Chest x-ray demonstrated cardiomegaly. Electrocardiogram showed right ventricular hypertrophy. Laboratory examination revealed hemoglobin 22.3 g/dl, hematocrit 65.9%, and pO₂ 42 mm Hg.

Discussion

Krarup described a patient with multiple cardiac defects and Eisenmenger syndrome who developed bilateral rubeosis iridis with spontaneous hyphemas and minimal retinal findings [3]. An additional report by Harino and associates described two patients with atrial septal defects and Eisenmenger syndrome, whose findings included microaneurysms, blot hemorrhages, and capillary dilation in the temporal peripheral retina [4]. One of these patients developed bilateral rubeosis iridis. Rodriguez reported a patient with Eisenmenger syndrome who developed bilateral visual loss secondary to central retinal vein occlusion [5]. Our patient developed unilateral central retinal artery occlusion.

In adults, the most common causes of cyanotic congenital heart disease are tetralogy of Fallot and Eisenmenger syndrome [6]. Patients with cyanotic congenital heart disease have arterial oxygen desaturation



Figure 1: Ophthalmoscopic appearance of right eye: retinal edema and cherry-red spot.

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Figure 2: Ophthalmoscopic appearance of left eye: retinal vessels dilation and tortuosity, small blot retinal hemorrhages.

resulting from the shunting of systemic venous blood to the arterial circulation. The hemostatic changes associated with Eisenmenger syndrome may lead to thromboembolic events, cerebrovascular complications, or the hyperviscosity syndrome [7]. As erythrocytosis caused by arterial desaturation develops in patients with Eisenmenger syndrome, the blood viscosity increases commensurately, and blood flow and oxygen transport eventually decrease. As demonstrated by our patient, individuals with Eisenmenger syndrome and associated

secondary polycythemia may develop unilateral retinal artery occlusive disease.

As demonstrated by our patient, individuals with ES and associated secondary polycythemia may develop unilateral retinal artery occlusive disease.

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