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Case Report

Central Retinal Artery Occlusion in a Patient with Sickle Cell Disease Treated with Recombinant Tissue Plasminogen Activator

Slaven Pikija[1], Marieta V. Peycheva[2,4], Rousan Aghayan-Ugurluoglu[3], Bernhard Ganser[1], Eugen Trinka[1]

[1] *Department of Neurology, Christian Doppler University Hospital, Paracelsus Medical University, Salzburg, Austria*

[2] *Department of Neurology, Faculty of Medicine, Medical University of Plovdiv, Plovdiv, Bulgaria*

[3] *University Clinic for Ophthalmology and Optometry, Christian Doppler University Hospital, Paracelsus Medical University, Salzburg, Austria*

[4] *Research Institute at Medical University of Plovdiv, Plovdiv, Bulgaria*

Corresponding author: Marieta V. Peycheva, Department of Neurology, Medical University of Plovdiv, 66 Peshtersko shosse Blvd., 4000 Plovdiv, Bulgaria; Email: mpeitcheva@yahoo.com; Tel.:+359 32 602 282

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Abstract

Central retinal artery occlusion is a rare ophthalmological complication of sickle cell disease, usually provoked by additional contributing risk factors and its treatment remains controversial. We describe a patient with sickle cell disease and a spontaneous central retinal artery occlusion of his left eye that probably has good result from intravenous thrombolysis. We want to add sickle cell disease as rare etiological cause of central retinal artery occlusion and perpetuate evidence of intravenous recombinant tissue plasminogen activator administration in this condition.

Keywords

central retinal artery occlusion, intravenous thrombolysis, sickle cell disease

INTRODUCTION

Sickle cell disease (SCD) is a hemoglobinopathy first described by Pauling and his group in 1949.[1] There are several types of the disease dependent of the inheritance of the hemoglobin S allele - HbAS, HbSS, HbSC, HbS - beta thalassemia. SCD is demonstrated with intermittent vaso-occlusive events that can affect any organ system and chronic hemolytic anemia.[2] The most common ophthalmological manifestation of SCD is the proliferative retinopathy.[3] Central retinal artery occlusion (CRAO) with the association with sickle cell disease is a rare complication, usually described with additional contributing risk factors and its treatment remains controversial.[4]

CASE REPORT

The patient is a 52-year-old African-American man. He was admitted to the hospital due to a sudden loss of vision in his left eye, which began at 2 am. The patient clearly confirmed the beginning of his complaints, denying a provocative moment. Apart from lost vision, he had no other complaints. There were no known concomitant diseases or history of familial diseases, no intoxication and no concomitant drugs. Somatic condition was stable – afebrile, normal auscultation of the heart, normal auscultation of the lungs, physically normal abdomen, no swelling of the limbs. On neurological examination, we found left amaurosis with no other focal symptoms, mRS 1, NIHSS 0. Cerebral computed tomography in the first hours did not show any pathological findings including ischemic lesions. An ophthalmologist examined the patients and an occlusion of the left central retinal artery was

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diagnosed. The visual acuity of the left eye was possible only in inferior temporal zone for counting fingers and the visual acuity of the right eye was 1.0. A fundoscopy performed later revealed a cherry red spot in the macula in left retina and old ischemic zone temporal inferior on right retina ***(Fig. 1)****. Intraocular pressure of both eyes was normal.*

Three hours and 17 minutes from onset of the symptom, a decision was made to start thrombolysis. The patient received intravenously 0.9 mg/kg of rtPA (recombinant tissue plasminogen activator), total of 81 mg with 10% as bolus and the remainder over one hour. After thrombolysis, the vision in the left eye was completely restored. The patient confirmed that his vision has begun to improve 2 hours after the onset of the symptom.

Detailed laboratory investigations proved that the patient had sickle cell carrier status, heterozygous inheritance with normal level of hemoglobin: Hb 13.2 g/dl (13.3-17.7), Er 4.39 T/l (4.50-5.90), Hct 37.3% (40.0-52.0), WBC and Plt - normal, TSH 1.87 μU/mL (0.50-4.20), coagulation normal, AT III normal, glucose 105 mg/dl (70-100), LDL-H 126 mg/dl (less than 115), lipoprotein A 212 nmol/l (less than 75), urine normal, normal levels of protein C and S, lupus and antiphospholipid antibodies, normal CRP, and normal cholinesterase.

Control ophthalmologic examination two days later showed normal visual acuity of both eyes. Fluorescein angiography of the left retina demonstrated normal vascularization but the optical coherence tomography of the same eye described ischemic changes ***(Fig. 2)****.*

Extensive search for cardioembolism was pursued but none was found. Ultrasound examination of the carotid arteries did not give any data for atherosclerosis and on transorbital approach, the flow of the left central retinal artery was preserved with no spot typical for a clot ***(Fig. 3)****.*

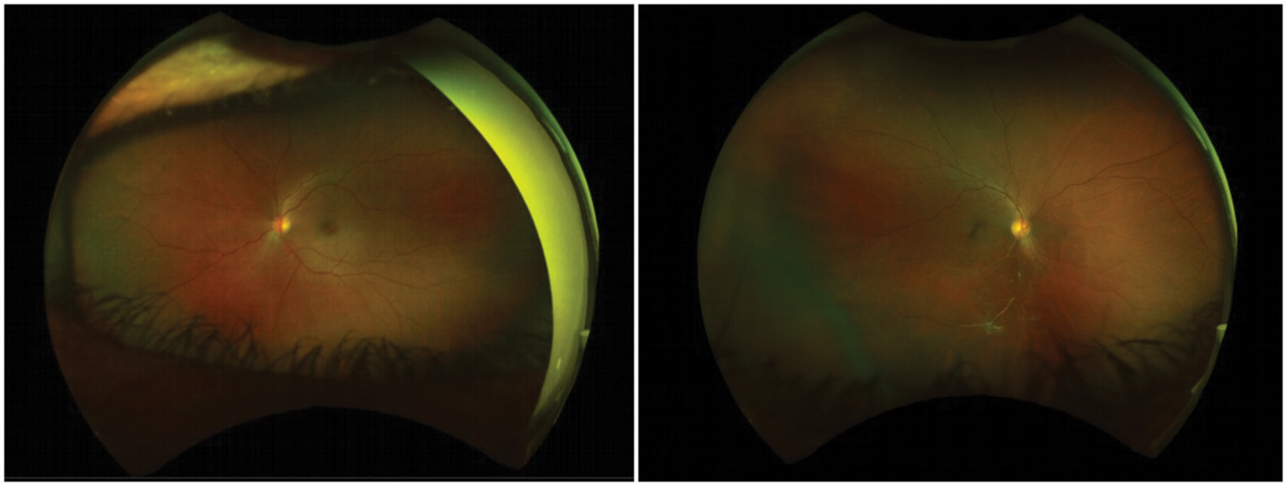

Figure 1. A fundoscopy with a cherry red spot in the macula (left) of the left eye and old ischemic zone temporal inferior on right retina (right).

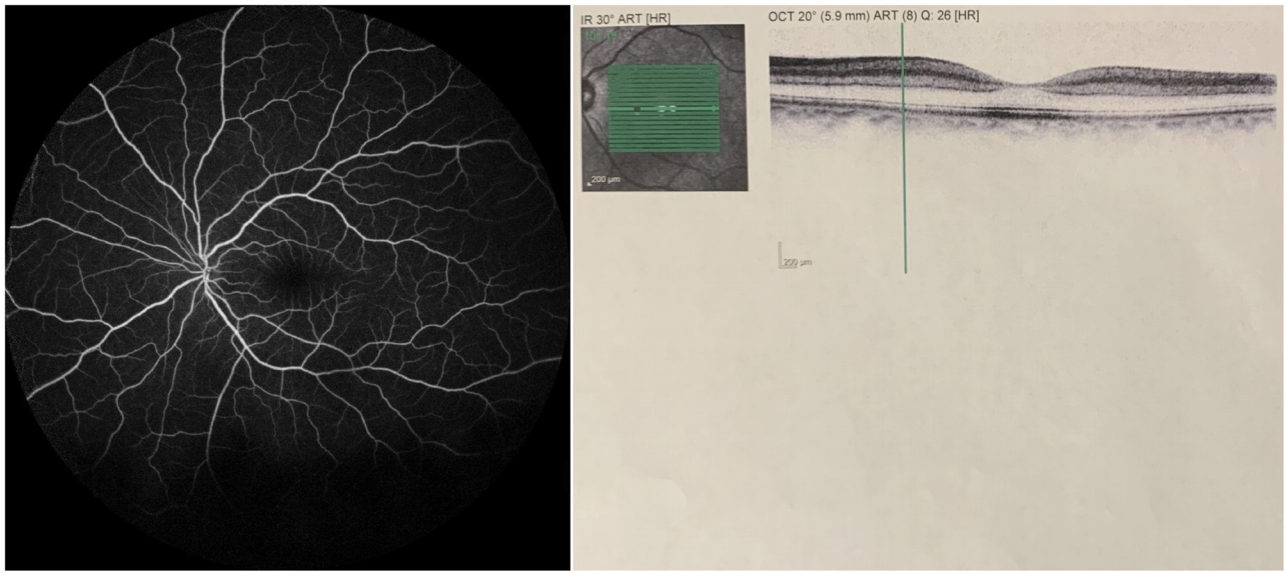

Figure 2. Fluorescein angiography (left) and optical coherence tomography (right) of the left retina.

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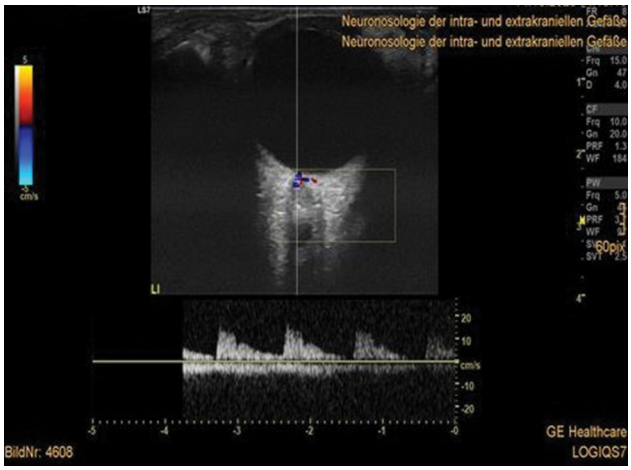

Figure 3. Transorbital approach for ultrasound investigation presents a preserved flow of the left central retinal artery.

A control cerebral MRI 5 days later was without abnormalities. The patent was discharged from the hospital with normal vision and in a stable physical and neurological condition. The etiology of the stroke was presumed to be due to sickle cell trait.

DISCUSSION

The prevalence of sickle cell trait in African-American population is 5-10% and traditionally considered as a benign condition by ophthalmologists.[3] Cases with spontaneous central retinal artery occlusion in patients with SCD are rare and usually additional contributing factors, such as trauma or concomitant systemic illness play a role in the pathogenesis.[4,5] In our case, the patient did not know about his sickle cell heterozygous carrier status and the central retinal artery occlusion was the first manifestation of the disease that occurred without a provocative moment. Risk factors like cardioembolism, atherosclerosis, diabetes, vasculitis, and systemic diseases were excluded.

Acute central retinal artery occlusion is an ophthalmologic emergency that leads to severe and permanent visual loss in more than 90% of cases.[6,7] SCD is a rare cause of CRAO as in most of the cases thromboembolism is the prevailing pathological mechanism.[8] Clinical signs and symptoms of CRAO are sudden loss of vision, visual acuity of counting fingers or worse, retinal edema (ischemic retinal whitening), a cherry red spot, retinal arteriolar attenuation, a retinal embolus in up to 40 and associate signs like headache, scalp tenderness, contralateral sensory or motor deficit.[9]

According to the ophthalmological guidelines, the treatment remains controversial with insufficient efficacy.[10] Conservative treatment with acetazolamide, ocular massage, and blood transfusion when Hb S level drops to 30% could be applied but usually with no effect. Selective ophthalmic artery catheterization with thrombolysis is tried in some centers with no benefits and the studies are stopped because of the higher rate of adverse reactions.[11] Meta-analysis of intravenous fibrinolytic therapy in CRAO[12] and a prospective interventional case series[6] showed a recovery rate of 31.8%, especially within the first 4.5 hours after symptoms onset. Rare complications are hemorrhagic events or subsequent deterioration of visual acuity. Our patient had a transient CRAO of his left eye that probably was affected beneficially from the intravenous rtPA therapy. We also want to add SCD as rare etiological cause of CRAO and perpetuate evidence of intravenous rtPA administration in this condition. The patient was treated in a center with significant experience with CRAO thrombolytic therapy (unpublished data from 11 CRAO cases).

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Competing Interests

The authors have declared that no competing interests exist.

Author contributions

All authors had substantial contributions to conception and design, acquisition, analysis, and interpretation of data, drafting the manuscript or revising it critically for important intellectual content. All authors approved the final version of the manuscript.

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Окклюзия центральной артерии сетчатки у пациента с серповидноклеточной анемией, получавшего лечение рекомбинантным тканевым активатором плазминогена

Славен Пикиджа[1], Мариета В. Пейчева[2,4], Русан Агхаян-Угурлуглу[3], Бернхард Гансер[1], Еуген Тринка[1]

[1] *Отделение неврологии, Университетская больница „Кристиан Доплер“, Медицинский университет Парацельса, Зальцбург, Австрия*

[2] *Кафедра неврологии, Факультет медицины, Медицинский университет – Пловдив, Пловдив, Болгария*

[3] *Университетская клиника офтальмологии и оптометрии, Университетская больница Кристиана Доплера, Медицинский университет Парацельса, Зальцбург, Австрия*

[4] *Научно-исследовательский институт, Медицинский университет – Пловдив, Пловдив, Болгария*

Адрес для корреспонденции: Мариета В. Пейчева, Кафедра неврологии, Факультет медицины, Медицинский университет – Пловдив, бул. „Пещерско шосе“ № 66, Пловдив, Болгария; Email: mpeitcheva@yahoo.com; Тел.:+359 32 602 282

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Резюме

Окклюзия центральной артерии сетчатки является редким офтальмологическим осложнением серповидно-клеточной анемии, обычно провоцируемым дополнительными сопутствующими факторами риска, и её лечение остается спорным. Мы описываем пациента с серповидно-клеточной анемией и спонтанной окклюзией центральной артерии сетчатки левого глаза, которая, вероятно, имеет хороший результат внутривенного тромболизиса. Мы хотим добавить серповидно-клеточную анемию в качестве редкой этиологической причины окклюзии центральной артерии сетчатки и рекомендовать применение внутривенного введения рекомбинантного тканевого активатора плазминогена в этом состоянии.

Ключевые слова

окклюзия центральной артерии сетчатки, внутривенный тромболизис, серповидно-клеточная анемия