Wide-field angiography for diagnosis and follow-up of Eales disease: a case series

Angiografia de amplo campo para diagnóstico e monitoramento da doença de Eales: série de casos

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ABSTRACT | Wide-field angiography enables assessing peripheral areas with better quality and gives greater deep focus, which improves the image periphery. Some studies have proposed the usefulness of these angiographic systems in inflammatory diseases of the retina. However, few studies have evaluated this technique in Eales disease. We present a case series in which 5 eyes of 3 patients with Eales disease were evaluated by using retinal fluorescein angiography with 30°, 50°, and 150° lenses in a laser-scanning ophthalmoscope. These cases highlight the usefulness of wide-field fluorescein angiography in the diagnosis and follow-up of peripheral ischemic retinal areas in Eales disease, which enables better follow-up than possible with conventional fluorescein angiography images.

Keywords: Fluorescein angiography; Eales disease; Retina/ pathology; Retinal diseases; Vision disorders

RESUMO | A angiografia de campo amplo permite avaliar áreas periféricas com melhor qualidade e proporciona maior foco profundo, melhorando a imagem da periferia. Alguns estudos têm proposto a utilidade desses sistemas angiográficos nas doenças inflamatórias da retina. No entanto, poucos estudos avaliaram esta técnica na doença de Eales. Apresentamos uma série de casos em que 5 olhos de 3 pacientes com doença de Eales foram avaliados usando angiografia de fluoresceína da retina com lentes de 30º, 50º e 150º em um oftalmoscópio de varredura a laser. Esses casos destacam a utilidade da angiografia com fluoresceína de campo amplo no diagnóstico e no acompanhamento das áreas isquêmicas periféricas da retina na doença de Eales, permitindo um melhor acompanhamento do o possível com imagens por angiofluoresceinografia convencional.

Submitted for publication: May 18, 2018

Accepted for publication: December 29, 2018

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Descritores: Angiografia com fluoresceína; Doença de Eales; Retina/patologia; Doenças retinianas; Transtornos da visão

INTRODUCTION

Eales disease (ED) is an idiopathic and occlusive inflammatory vasculopathy that primarily affects the retinal periphery^(1,2). The clinical diagnosis is based on the presence of perivascular phlebitis, non-perfusion tissue, and retinal neovascularization^(2,3). Presently, wide-field lens systems have been designed to capture images >150° of the retina in combination with confocal scanning laser ophthalmoscopes^(4,5). Wide-field angiography (WFA) enables assessment of peripheral areas with better quality and gives greater deep focus, which provides an improved image of the periphery⁽⁵⁾. Some studies have proposed the usefulness of these angiographic systems in inflammatory diseases of the retina, macular degeneration, and occlusion of the central vein of the retina, among others⁽⁴⁻⁶⁾. However, few studies have evaluated this technique in ED⁽⁷⁾. The aim of this study is to report the usefulness of WFA for diagnosis and follow-up in three cases of ED.

CASE REPORTS

Five eyes of three patients diagnosed with ED were evaluated. We ruled out the possibility of other systemic diseases that cause retinal vasculitis and proliferative vascular retinopathy mimicking ED (such as tuberculosis, sarcoidosis, syphilis, systemic lupus erythrematosus, toxoplasmosis, diabetes mellitus, sickle cell disease) after proper history, examination, and investigations (chest x-ray, hemoglobin, hematocrit, total red and white blood cell counts, differential count, platelet count, erythrocyte sedimentation rate, reticulocyte count,

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Funding: No specific financial support was available for this study. Corresponding author: Walter A. Tellez.

fasting and postprandial blood sugar, sickle cell preparation, serum angiotensin-converting enzyme, and antinuclear antibody). They underwent traditional fluorescein angiography (TFA) and WFA, corrected visual acuity, biomicroscopy, and fundoscopy.

A Staurenghi lens (150°) associated with the confocal scanning laser ophthalmoscope (Spectalis HRA; Heidelberg Engineering, Dossenheim, Germany) was used to capture a wide-field angiographic image. All patients had impaired visual acuity, bilateral ED in 2 patients, a mean follow-up time of 12 months [standard deviation (SD)=3.27 months], vitreous hemorrhages in two different eyes, association with systemic disease of pulmonary tuberculosis in one patient, and significant improvement in their visual acuity after treatment with vascular endothelial growth factor inhibitors, laser photocoagulation, or surgical treatment (Table 1).

Case 1

A 28-year-old male patient with no relevant medical history underwent ophthalmic evaluation because of a sharp decrease in visual acuity in the right eye. He presented with initial visual acuity of 20/200 in the right eye (OD) and 20/20 in the left eye (OS). The fundoscopic examination showed moderate vitreous hemorrhage in OD (Figure 1A). Wide-field imaging revealed areas of neovascularization and peripheral ischemia in OD and OS, respectively (Figure 1B). He was diagnosed with bilateral ED and received antiangiogenic treatment and laser photocoagulation. Finally, visual acuity improved to 20/20 in both eyes. Although we prevented progression of the disease in OS, the improvement in OD was because of reabsorption of the hemorrhagic vitreous.

Case 2

A 23-year-old male patient presented with initial visual acuity of 20/30 in OD and 20/20 in OS. The reason for consultation was a decrease in visual acuity in OD. Wide-field images showed retinal neovascularization and areas of tractional fibrous proliferation (Figure 2). He was diagnosed with unilateral ED associated with pulmonary tuberculosis and received treatment based on laser photocoagulation. Finally, visual acuity was 20/20 bilaterally.

Case 3

A 29-year-old woman presented with initial visual acuity of 20/25 in OD and 20/800 in OS. She sought an ophthalmological consultation for control of a vitrectomy performed 10 years earlier and was without

Table 1. Ophthalmological characteristics, associated diseases, and treatment of patients with ED

Cases	Age	Sex	Eye	Visual acuity	Fundoscopy	Diagnosis	Associated systemic disease	Treatment	Time of follow-up (months)	Last visual acuity	Last control
Case 1	28	м	OD	20/200	Hemorrhagic vitreous	ED	Mantoux +, inactive tuberculosis	Antiangiogenic (bevacizumab 1.25 mg/0.05 ml for 3 months, 1 injection each month in each eye)	12	20/20	Laser applied retina
			OS	20/20	Laser applied retina, peripheral ischemia	ED				20/20	Normal optic nerve, macula with no alteration, and panretinal photocoagulation
Case 2	23	М	OD	20/30	Hemorrhagic vitreous, inferior tractional fibrous proliferation	ED	Pulmonary tuberculosis	Laser photocoagulation and periocular triamcinolone	8	20/20	Laser applied retina
			OS	20/20	Laser applied retina	Retina and optic nerve with no alteration				20/20	Retina and optic nerve with no alteration
Case 3	29	F	OD	20/25	Laser applied retina, stable	ED	None	Laser photocoagulation in OD and phacoemulsifi- cation in OS	16	20/25	Laser applied retina, peripheral vitreous band without proliferation
			OS	20/800	Laser applied retina, focal activity	Cataract and ED				20/100	Laser applied retina

symptoms. Wide-field angiographic examination identified areas of focal activity in OS due to old bilateral ED reactivated (Figure 3), which were treated by laser photocoagulation in OD and phacoemulsification in OS. Eventually, visual acuity became 20/25 in OD and 20/100 in OS. The visual acuity improvement in the OS was because of phacoemulsification of the cataract.

DISCUSSION

ED is an idiopathic vasculitis that is difficult to diagnose and treat⁽⁸⁾. FA is an important tool for the detection and follow-up of patients with this disease. However, TFA only has limited vision of the retina because it captures images in a range of 30°-50°, with loss of visibility of the peripheral retina that is the most compromised area

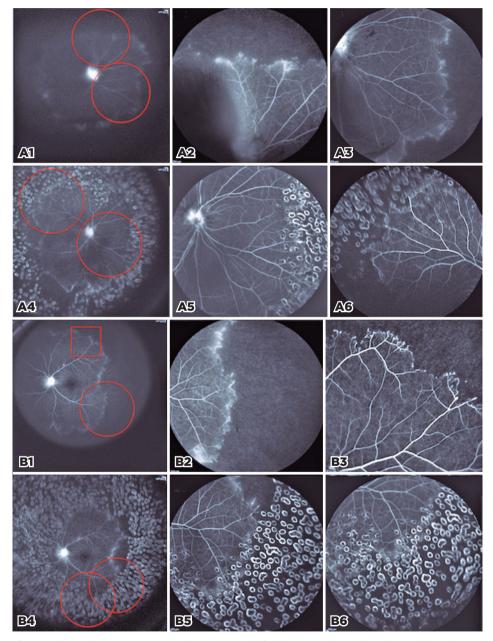


Figure 1. Case 1 OD angiogram. A1 shows 150° wide-field angiography, evidencing peripheral retinal ischemia with arteriolar narrowing; the image areas showing vitreous hemorrhage are diffused. The circles in red show the images with fields of 50° that are shown in detail in A2 and A3. In A4, the same eye after laser treatment is shown. The details of the periphery and the 50° images are visible (A5 and A6). Case 1 OS angiogram. B1 shows a 150° wide-field angiographic image; the square shows a detailed image in a 30° field and the circle shows a 50° field that magnify the areas shown in B2 and B3, respectively. B4 shows the same eye after photocoagulation treatment with wide-field angiography, and the details of zones are marked with circles in B5 and B6.

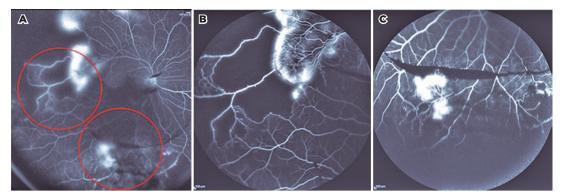


Figure 2. Case 2 angiogram. A shows a 150° wide-field image, with areas of retinal hypoperfusion, hemorrhagic vitreous, and hyperfluorescence in the temporal peripheral retina with the presence of multiple sea fan formations. In B and C, sea fans and hemorrhagic vitreous are shown with more details.

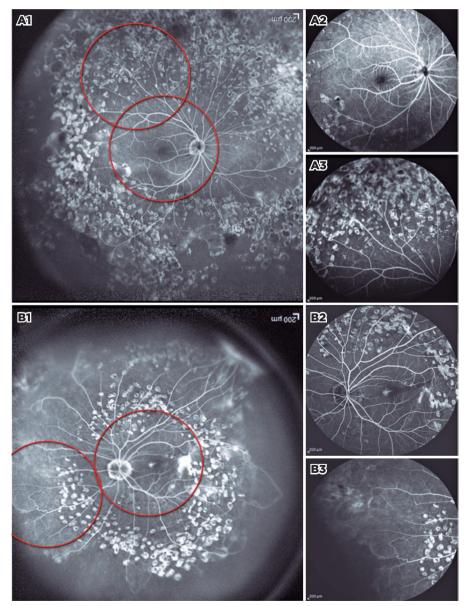


Figure 3. Case 3 angiograms. Al shows the OD wide-field angiography, and A2 and A3 show more details in 50° field images. B1 shows OS wide-field angiographic evidence of peripheral changes in the retina. Those changes are seen with more detail in B2 and B3.

in this disease⁽³⁾. This limitation in the examination has been overcome by the use of lens systems with a greater field amplitude of >150°, which enables peripheral assessment of the retina, as documented in other studies related to peripheral retina diseases^(5,9).

In ED, it is important to find retinal areas without perfusion but with vascular anomalies between the still vascularized retina and the ischemic retina, which is a common finding in the angiogram mainly in the periphery^(2,3). The absence of confluent retinal flow has been observed in all of the reported cases. The difference in flow between the anteroperipheral retinal binding sites lacking blood flow and the well-irrigated posterior area is well demarcated and is seen in the images (Figures 1, 2), which show multiple vascular alterations, such as microaneurysms, venous rosaries, and remnants of obliterated vessels that are stained with fluorescein.

Although there is a lack of retinal peripheral blood flow, it does not affect the macula generally, which preserves the central vision⁽¹⁰⁾, as observed in Cases 1 and 2; however, in some patients, peripheral ischemia can condition physiopathological changes that are related to a decrease in central vision due to the presence of macular edema, intraretinal hemorrhages, and macular ischemia, among other changes⁽¹⁰⁾, as we observed in the left eye of the patient in the third case (Figure 3, B1-B3).

Neovascularization is common in this disease and is located mainly in the periphery between the perfused retina and the one lacking blood flow⁽²⁾. In our case series, neovascularization was present in all cases and was associated with vitreous hemorrhages in two of the patients. In the patients with hemorrhage, there was a sudden decrease in vision, as shown in the OD of the patients in Cases 1 and 2 (Figures 1, 2). The peripheral presence of neovessels is an important finding for determining the visual prognosis of the patient, so it must be described in the angiographic evaluation to provide effective treatment; therefore, it is useful to have broad field images to determine their location.

Kumar et al.⁽⁷⁾ recently reported the usefulness of WFA on 24 eyes of 17 patients diagnosed as having ED. This was the first published article about a study that evaluated how WFA improves the diagnosis and management of this disease, as reported in other peripheral retinal diseases. The researchers recruited more cases and eyes with ED than those in our case series; however, our case series differs in some respects from that of Kumar et al.; their group followed up the cases from 3 to 10 months (12 to 40 weeks), whereas our study had a follow-up period from 8 to 16 months (32 to 64 weeks). Additionally, they only used WFA to evaluate their cases and stated that simultaneous conventional photography was not possible for a variety of different reasons; in our study, we performed WFA and TFA in every included case, which enabled us to identify the pathological changes broadly by using WFA and evaluate them in detail with the targeted TFA, which is also useful for evaluating retinal changes after proper treatment. Therefore, our case series has some advantages, such as a longer follow-up period and simultaneous WFA and TFA for comparison.

WFA was helpful for the detection of peripheral lesions in the retina that suggested ED in eyes with and without symptoms and also generated changes in the treatment plan for each case. For instance, the patient in Case 1 obtained ophthalmological consultation because of a decrease in visual acuity in OD; however, when examining both eyes with WFA, areas of neovascularization and peripheral ischemia were seen in both eyes, so we decided to apply antiangiogenic agents as treatment. The patient in Case 2 was diagnosed with ED in OD in which WFA showed hemorrhagic vitreous and inferior tractional fibrous proliferation; therefore, we decided to perform laser photocoagulation and apply periocular triamcinolone, which was a different scheme from that used in Case 1 because it has been reported that the application of antiangiogenic agents when there is concomitant proliferation was associated with retinal detachment and hemorrhages. Finally, the patient in Case 3 obtained ophthalmological consultation for routine control of a previous vitrectomy without significant symptoms suggesting ED, but when WFA was performed, we could visualize retinal changes compatible with ED, so we performed laser photocoagulation in OD and only phacoemulsification in OS.

In conclusion, WFA can contribute considerably to the diagnosis and monitoring of ED by detecting some important characteristics of this disease, such as vascular inflammation, recurrent vitreous hemorrhage, and neovascularization, which enables better monitoring of the peripheral areas and can guide changes in the management of the patients.

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