

Bilateral Leber's miliary aneurysm in a female black African

Ogugua Ndubuisi Okonkwo, Adekunle Olubola Hassan, Valentina Ideh, Micheal Ekwuoba Gyasi, Olufemi Oderinlo

ABSTRACT

Introduction: Leber's Miliary Aneurysm (LMA) is a primary telangiectasia of the retina blood vessels with characteristic exudation. It is an idiopathic disease, mostly unilateral and seen predominantly in males. This case report represents the first case of Leber's Miliary Aneurysm reported in a Nigerian and perhaps in sub-Saharan black Africa. **Case Report:** A 26-year-old female presented with a bilateral retina disease characterized by multiple retinal vascular aneurysmal dilatations, telangiectasia of the retina blood vessels with right eye macular atrophy and retinal exudation in both the eyes. This resulted in severe loss of vision in the right eye and moderate loss of vision in the left eye. Retinal laser photocoagulation was used to treat the leaking aneurysms in the left eye. **Conclusion:** This case demonstrates that a rare disease such as LMA can occur bilaterally in a female of African descent. Thus, eye specialists practicing in this region should be aware of this possibility.

Ogugua Ndubuisi Okonkwo¹, Adekunle Olubola Hassan¹, Valentina Ideh², Micheal Ekwuoba Gyasi³, Olufemi Oderinlo¹

Affiliations: ¹Consultant Ophthalmic Surgeon, Eye Foundation Retina Institute, Eye Foundation Center for the Prevention of Blindness, Lagos, Nigeria; ²Consultant Ophthalmic Surgeon, Department of Ophthalmology, Prime Optical Center, Benin, Nigeria; ³Consultant Ophthalmic Surgeon, Department of Ophthalmology, St Thomas Eye Hospital, Accra, Ghana.

Corresponding Author: Ogugua Ndubuisi Okonkwo, Eye Foundation Retina Institute, Eye Foundation Center for the Prevention of Blindness, 27 Isaac John Street, GRA. Ikeja, Lagos, Nigeria; Email: o_okonkwo@yahoo.com

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INTRODUCTION

Leber's miliary aneurysm (LMA) is a rather rare primary vascular anomaly characterized by vascular telangiectatic lesions and retina exudation [1]. It falls into the spectrum of Coats like disease. Coats first described this disease in 1908 [2]. In his initial description, retinal vascular telangiectasia and exudation were the predominant features; this clinical feature has been similarly reported by other researchers [3, 4]. This disease like Coats disease, is unilateral and more commonly seen in males [2, 5, 6]. In 1912, Leber described a retinal disorder, characterized by retinal degeneration and multiple miliary aneurysms [7]. He later felt the disease he described was a variant of Coats' disease. Resse in 1956 characterized both diseases as being the same, and felt that LMA was in a state of evolution into Coats' disease [8].

Leber's miliary aneurysm therefore has been characterized as a primary retinal telangiectatic disease of the retina vessels associated with exudation of lipids. The cause is unknown.

In this case report, LMA occurs as a bilateral asymmetric disease in a female Nigerian. Leber's miliary aneurysm has not been reported previously in the African population when search was done using popular search engines including Medline and Google scholar.

CASE REPORT

A 26-year-old female was referred to the retina clinic on account of a three-month history of gradual, painless reduction in left eye vision. The right eye vision had been poor for several years. There was no previous history of trauma or systemic illness. Her family history revealed nothing significant. Systemic review was entirely normal.

Upon examination her visual acuity was reduced for right eye to counting fingers and left eye 6/18. This could not be improved with refraction or use of a pinhole. Intraocular pressure was 10 mmHg in both eyes. The anterior segment examination was normal with reactive pupils in both eyes.

Dilated fundus examination (stereoscopic biomicroscopy) was done using a 90D (Volk) non-contact lens. A binocular indirect ophthalmoscopy using a 20D (Volk) non-contact lens was used for more peripheral retina examination. This examination revealed that the right eye had more chronic features of central geographic chorioretinal atrophy, scarring and pigmentary dispersion (Figure 1). There were arteriolar bulb like aneurysmal dilatations, with surrounding leakage and hard exudates in the superonasal peripheral retina of the right eye (Figure 2). Also, in the right eye were multiple focal whitish irregular fibrotic lesions (these were likely previous aneurysms that had undergone thrombosis and scarring, leaving the white lesions as localized fibrotic tissue (Figure 1)). It was assumed that the central area of chorioretinal atrophy once had hard exudates that were now reabsorbed, following sclerosis and closure of leaking aneurysms.

The left eye, which had more active findings, on clinical examination, had multiple retina arteriolar aneurysmal dilatation and vascular telangiectasia around the temporal arcades as well as nasal and temporal mid peripheral retina. This was responsible for the dense patch of hard exudation and significant edema seen in the posterior pole, and milder degree of leakage seen in the mid periphery (Figures 3 and 4).

A retina fundus photograph and fundus fluorescein angiography (FFA) was obtained. Fundus photography revealed features in keeping with the spectrum of Coats' like retina vasculopathy; having retina telangiectasia, multiple aneurysmal dilatation and exudation as the primary features.

FFA was essential and in general, both retinae presented having multiple macroaneurysms, with some of them leaking, and some others cicatrized. Moreover, all the areas containing macroaneurysms did also have

telangiectatic vessels that were "Coats type". There were areas of non-perfusion surrounding the aneurysms.

The right eye had a central area of wide spread geographic atrophy, pigmentary dispersion {with hyperfluorescence due to window defects on FFA (Figure 5A)}, and chorioretinal scarring. All these may have occurred as a result of previously untreated but resolved retinal exudation. There were obvious areas of peripheral hypofluorescence surrounding the multiple bulb-like aneurysmal hyperfluorescent areas. This was due to non-perfusion (Figure 5B).

The left eye had more actively leaking macroaneurysms and exudation. There were several hyperfluorescent

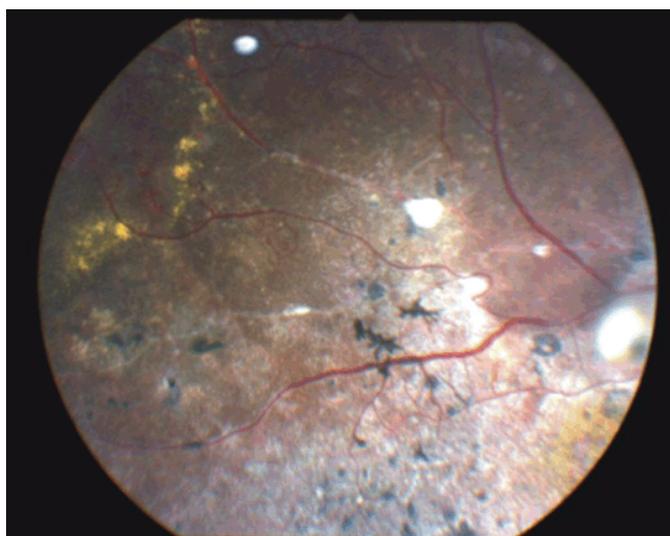


Figure 1: Right eye fundus picture showing significant chorioretinal atrophy involving the macular region. There is presence of significant pigmentary dispersion. The localized whitish round fibrotic lesions, are noticeable which represent previous aneurysm, which now have become thrombotic.

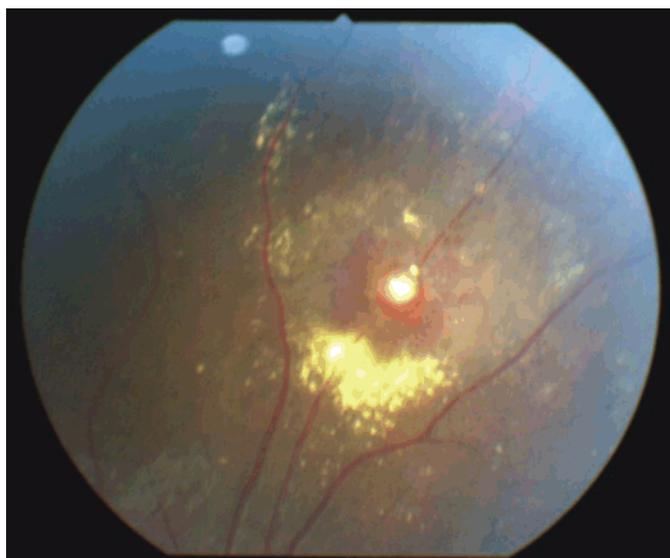


Figure 2: Right eye fundus picture showing peripheral arteriolar bulb like aneurysmal dilatations, with surrounding leakage and hard exudates in the superonasal peripheral retina.

bulbs in the posterior pole, nasal and temporal mid periphery and more peripheral retina due to pooling of dye in the multiple, variously sized macro aneurysms and telangiectatic vessels. Areas of hypofluorescence were seen in the macula due to blocked fluorescence from the parafoveal hard exudation and from areas of non-perfusion (Figure 6).

As this case was seen in the era prior to the clinic acquiring an optical coherence tomography (OCT), an OCT was not available and could not be used for further evaluation of the macular pathology.

A diagnosis of asymmetric Leber’s miliary aneurysm (LMA) was made based on these clinical and FFA findings. She was advised to have treatment to the leaking aneurysms in the left eye.



Figure 3: Left eye fundus picture showing multiple actively leaking aneurysmal dilatations and telangiectasia in the macular area. The dense patch of parafoveal hard exudation is also evident.

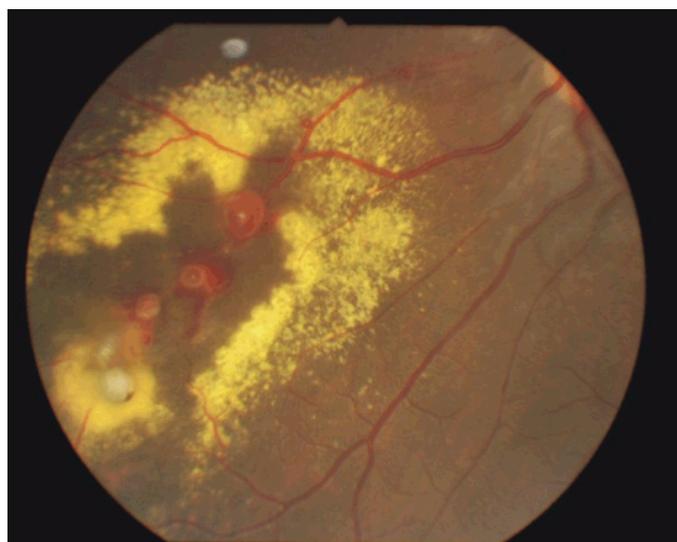
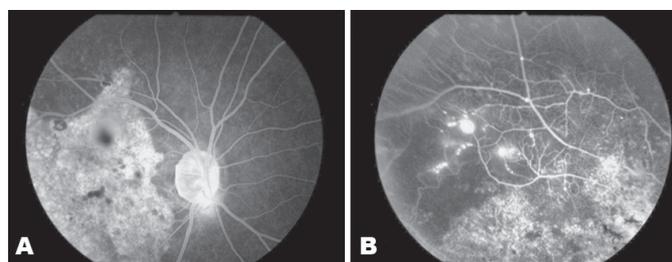


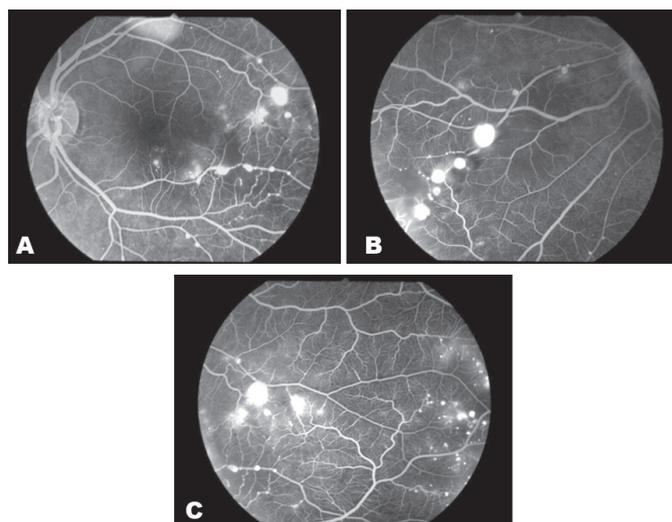
Figure 4: Left eye fundus picture showing similar aneurysmal dilatations in the nasal mid peripheral retina.

She received focal thermal retinal laser photocoagulation to leaking aneurysms using a green (532 nm) laser and had two sessions of retinal laser in the left eye. The retina laser was directed to the para macula aneurysms in the left eye. Also, she had an additional session of trans pupillary thermotherapy (TTT) using the diode (810 nm) laser. This was applied to the same aneurysmal dilations in the extrafoveal area of hard exudation and retina edema in the left eye.

She attended the retina clinic for regular follow up visit but was later lost to follow up after she relocated abroad and could no longer keep up with hospital appointments. Her visual acuity remained unchanged at 6/18 all through the period of follow-up.



Figures 5(A and B): Right eye fundus fluorescein angiography showing the areas of macular hyperfluorescence, which is due to atrophy (window defect). There are areas of localized hyperfluorescence representing dye pooling within the aneurysms. These aneurysms are surrounded by areas of hypoperfusion in the retina periphery, resulting in the hypofluorescence seen.



Figures 6(A–C): Left eye fundus fluorescein angiograms showing areas of multiple aneurysms in the macular, nasal and temporal mid periphery and peripheral retina seen as hyperfluorescent bulb like lesions. Hypofluorescence is noted due to blocked fluorescence from the hard exudates present in the macular. There are hypofluorescent areas resulting from hypoperfusion.

DISCUSSION

Leber's miliary aneurysm represents one of the three known primary retinal telangiectasia. Juxtafoveal telangiectasia, Leber's miliary aneurysm, and Coats' disease are all well described and are considered to belong to the same clinical spectrum of primary retinal telangiectasia [9, 10]. They are a group of rare, idiopathic congenital, retinal vascular anomalies characterized by dilation and tortuosity of retinal vessels, formation of multiple aneurysms, varying degrees of leakage and deposition of hard exudates in form of lipids. Though Leber's miliary aneurysm and Coats' disease were initially described as separate diseases, most authorities now recognize them as variable expressions of the same disease spectrum, as both diseases are characterized by presence of vascular anomaly. Leber's miliary aneurysm is seen as the milder form of the disease while Coats' disease is the more severe end of the spectrum. Both are therefore currently grouped under the common name of Coats disease. [8, 10].

Retinal telangiectasia always involves the capillary bed, although arteries and venules may also be affected. In the case presented, arteries seem to be most affected as indicated by the clinical examination and on fluorescein dye testing. The vascular malformations frequently progress and may become symptomatic later in life as a result of hemorrhage, edema or lipid exudation. The characteristic features include mostly telangiectasia and retina exudation. It is mostly a unilateral disease with male predominance. Female affection is uncommon and bilateral disease though rare [11], has been previously reported and noted to be asymmetric in a female patient as in our case [12]. There is usually no systemic effect and association reported.

Leber's miliary aneurysm, which is taken to represent a milder form of the spectrum, does not have severe exudation and hemorrhaging as a characteristics; as is seen in the typical Coats' disease which could present with exudative retina detachment and significant visual loss [6]. In LMA, most patients are asymptomatic at presentation with minimal exudation. Vision becomes affected when exudates affect the macula as seen in our case.

Also, in our case the right eye already had evidence of chronic disease affecting the macular. The leakage and exudation was considered to have started in the right eye at an unknown time; undetected by the patient. Central involvement of the left eye by leakage and lipid exudates raised the alarm following a noticeable loss of vision. The bilateral and asymmetric presentation in female patient is rare. This case demonstrates that such rare congenital disease can be seen in a Nigerian and a black African.

Treatment is often initiated when exudation involves the macular resulting in reduced vision as seen in our case. Though treatment of leaking aneurysm is generally done with retina laser photocoagulation [12, 13], and

trans scleral cryotherapy to peripheral lesions [14], there are reports of intravitreal anti Vascular Endothelial Growth Factor (VEGF) use to treat macular edema [15]. There has been a report of spontaneous resolution of the macular edema with return of good vision [16]. The right eye of our case had a spontaneous resolution of macular exudates we believe, but visual recovery was limited by the extensive chorioretinal atrophy that occurred. The goal of treatment is to control exudation by the leaking aneurysms and to prevent further progression to visually threatening complications. It is important to treat the entire area of abnormal vessels. Fluorescein angiography is helpful in visualizing the full extent of leaking aneurysms and non-perfusion, as demonstrated in this case. Multiple treatment sessions may be required to ensure complete treatment of all leaking aneurysm [17], as observed in our case too, in which three separate sessions of retinal laser were performed at intervals to the left eye.

Adequate vascular closure, and ablation of the ischemic peripheral retina should be done to prevent progression to neovascular complications, especially in Coats' disease. Despite adequate treatment, resolution of the macular edema and hard exudates may take several months.

CONCLUSION

To conclude, Leber's miliary aneurysm though rare can present as a bilateral disease in a female of African descent, indicating that the gene pool and other causative factors necessary for its occurrence does exist amongst the black African population. This also suggests that other primary retina telangiectasia including coats diseases can occur in a black African population. Since this finding has not been previously reported, Ophthalmologist and retina specialist practicing in this region should be aware of this possibility and should be able to identify such a rare case when it occurs.

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Author Contributions

Ogugua Ndubuisi Okonkwo – Substantial contributions to conception and design, Drafting the article, Revising

it critically for important intellectual content, Final approval of the version to be published

Adekunle Olubola Hassan – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Valentina Ideh – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Micheal Ekwuoba Gyasi – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Olufemi Oderinlo – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None

Consent Statement

Written informed consent was obtained from the patient for publication of this study.

Conflict of Interest

Authors declare no conflict of interest.

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