



---

## **Coats Disease in Nigeria: A Case Series**

**B. Fiebai<sup>1\*</sup>, T. S. Oluleye<sup>2</sup> and I. O. Omaka<sup>3</sup>**

<sup>1</sup>*Department of Ophthalmology, Vitreo Retinal Unit, University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria.*

<sup>2</sup>*Department of Ophthalmology, Vitreoretina Services, University College Hospital, Ibadan, Oyo State, Nigeria.*

<sup>3</sup>*Department of Ophthalmology, Federal Medical Centre, Yenagoa, Bayelsa State, Nigeria.*

### **Authors' contributions**

*This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.*

### **Article Information**

DOI: 10.9734/AJMAH/2018/46010

#### Editor(s):

(1) Dr. P. Veeramuthumari, Assistant Professor, Department of Zoology, V.V.Vanniaperumal College for Women, Virudhunagar, India.

#### Reviewers:

(1) Engy Mohamed Mostafa, Sohag University, Egypt.

(2) Kevin J. Blinder, Washington University, USA.

Complete Peer review History: <http://www.sciencedomain.org/review-history/27988>

**Case Study**

**Received 22 September 2018**

**Accepted 08 December 2018**

**Published 28 December 2018**

---

## **ABSTRACT**

**Aims:** To report a series of four cases of Coats disease in black Nigerians, a rare disorder.

**Study Design:** A case series.

**Place and Duration of Study:** Retina unit of the departments of ophthalmology at the University College Hospital, Ibadan, Oyo State and the University of Port Harcourt Teaching Hospital, Rivers State, Nigeria. Duration of study was 2014-2018.

**Methodology:** Case folders of patients who presented to the retina units of the University College Hospital Ibadan, University of Port Harcourt Teaching Hospital and a peripheral eye hospital between 2014 and 2018, were reviewed. Data collected includes age, sex, presenting visual acuity, findings on fundus examination and reports of ancillary tests.

**Results:** Four eyes of 4 patients were reviewed. There were 2 males and 2 females. 3 of the four cases were found in children while one case presented in adulthood. All eyes presented with profound visual loss with unioocular presentation and exudative retinal detachments at the late stages, with poor visual prognosis.

**Conclusion:** Coat's disease though an uncommon ocular disorder in Nigeria, does exist and may have been underdiagnosed or misdiagnosed. Routine examination of children is pertinent in early

---

\*Corresponding author: E-mail: [bassief@yahoo.com](mailto:bassief@yahoo.com);

diagnosis and prompt treatment to save vision and a good knowledge of its clinical presentation may lead to more case findings.

**Keywords:** Coats disease; leucocoria; retinal telangiectasia; sub-retinal exudates; retinal detachment; Nigeria.

## 1. INTRODUCTION

Coats' disease is a rare, idiopathic ocular disorder resulting from abnormal development of the retina vasculature [1]. It is characterized by progressive intra retinal and sub retinal exudation, retinal hemorrhages and retina telangiectasia [1-4]. The disease was first described in 1908 by George Coats a Scottish ophthalmologist who observed retina fluid accumulation and exudates with aneurysms in young male patients [5]. The exact etiology remains unknown, however mutations in the retinal proteins NDP are believed to exist in diseases involving retinal vasculogenesis [6]. Most cases however are believed to occur sporadically with no specifically identified hereditary pattern. The pathological processes involved, includes a breakdown of the blood retinal barrier of the endothelial cells and loss of pericytes leading to leakage of lipid rich exudates and formation of aneurysms [7,8].

Coats' disease commonly occurs in childhood between the ages of 5-16, presenting more in the first decade [2,4]. However a few presentations in adults have been reported [9,10]. It is more common in males and is usually unilateral [2,11,12]. Presentation is usually protean with the most common being decrease in vision, strabismus and leukocoria [2,4,11-13].

Treatment in the past involved basically the use of ablative lasers but in the last decade the use of anti -vascular endothelial growth factors has emerged as a promising modality of treatment as well [14,15].

To the best of our knowledge, Coats' disease has not been reported before in a Nigerian child, though Mahmoud et al reported a case of a man with the disease [9]. Our aim in this series, therefore is to report four cases of Coats disease seen in four Nigerians.

## 2. PRESENTATION OF CASES

### 2.1 Case Reports

#### 2.1.1 Case 1

A 12 year old girl presented in the retina clinic of a tertiary hospital with complaints of poor vision

in the right eye since childhood. Systemic history was unremarkable. Visual acuity at presentation was 'Light perception on the Right eye and 6/6 on the left with a right exotropia. Intraocular pressure in both eyes was within normal limits. There was a relative afferent pupillary defect (RAPD). Fundoscopy revealed a leukocoria in the right eye and massive exudation with retinal haemorrhages and telangietatic vessels. This was classified as a Stage 3A disease [16]. The differential diagnosis considered in this case included retinopathy of prematurity and toxocara canis. The lack of a history of low birth weight and supplemental oxygen administration and the unilateral nature ruled out the former and in the latter, there was no history of contact with an infected animal.

Intravitreal Avastin was offered however the patient was lost to follow up.

#### 2.1.2 Case 2

A 25 year old man presented to the retina clinic of a tertiary facility with a history of poor vision since childhood in the right eye. There was no associated systemic disease. Visual acuity assessment was 'Counting Fingers' in the right eye and 6/6 in the left eye. The intraocular pressure in both eyes was within normal limits. The left eye was essentially normal, however pupillary examination of the right eye revealed a subtle RAPD. Fundoscopy of the right eye showed subretinal exudation with dilated vessels. The left eye was within normal limits. We classified this as a Stage 3A disease [16]. Familiar exudative vitreoretinopathy (FEVR) was a differential in this case but the presentation was unilateral.

#### 2.1.3 Case 3

A 9 year old boy was referred to the retina clinic of a peripheral hospital from the general ophthalmology clinic with complaints of poor vision in the right eye of 2 years duration with a positive history of nyctalopia. Systemic history was unremarkable.

Presenting visual acuity in the Right eye was 'Perception of Light' with good light projection in



**Fig. 1. Patient 1 with Leucocoria, massive exudation and haemorrhage**

all quadrants and 6/5 in the left eye. The intraocular pressure was 12 mmHg in the right eye and 14 mmHg in the left eye. The globe was normal in both eyes with no strabismus. Other findings in the left eye were generally unremarkable and findings were limited to the right eye only. There was a relative afferent pupillary defect. The lens was clear with no opacity. There was xanthocoria from a shallow retinal detachment and extensive subretinal exudates with an area of telangiectasia in the inferotemporal quadrant. Optical coherence tomography showed intraretinal and subretinal exudates and a B scan ultrasonography showed a shallow retinal detachment. According to Shield's classification, this patient was classified as stage 3A. The differential diagnosis here included retinopathy of prematurity and retinoblastoma. However, history and ultrasonography ruled these out.

fundus fluorescein guided laser photocoagulation. Patient however was lost to follow up and returned 6 months later with a visual acuity of 'no perception of light'.

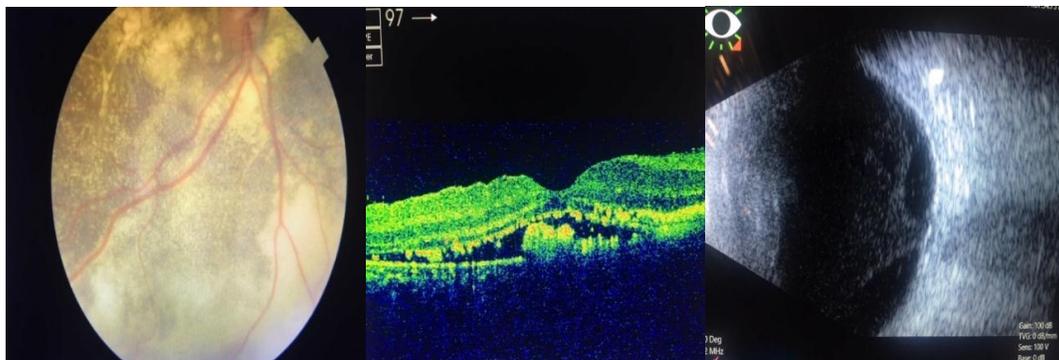


**Fig. 2. Fundus picture of patient 2 showing sub- retinal exudation**

#### 2.1.4 Case 4

The modalities of treatment offered to the patient included intravitreal injection of an anti vascular endothelial growth factor agent with a follow up

The last case in our series is a 14 year old girl who presented with a one year history of



**Fig. 3. Fundus picture of patient 3 showing extensive subretinal exudation. OCT showing intraretinal, subretinal exudation and retinal detachment. Bscan showing retinal detachment B scan of patient 3 showing retinal detachment**



**Fig. 4. Fundus picture of Patient 4 showing massive retinal exudates and Bscan on the right showing sub retinal exudates**

**Table 1. Patients baseline data and classification of four eyes of four patients**

Patient	Age	Sex	Eye	Initial BCVA	Presenting complaint	Classification	Planned treatment
1	12	Female	Rt	LP	Loss of vision, Leucocoria	Stage 3A	Anti -Vegf
2	25	Male	Rt	CF	Loss of vision	Stage 3A	Anti -Vegf
3	11	Male	Rt	LP	Loss of vision	Stage 3 A	Anti-Vegf/Laser
4	14	Female	Lt	LP	Loss of vision, strabismus	Stage 3 A	Anti Vegf

KEY: Rt- Right eye, Lt- Left eye, BCVA- Best corrected visual acuity, LP – Light Perception  
Anti- Vegf- Anti vascular endothelial growth factor, Classification of Coats disease [16]

uniocular loss of vision and strabismus in the left eye. Examination revealed a presenting VA of 6/5 in the right eye and ‘Light perception’ in the left eye. The right eye was unremarkable. There was 30 degrees exotropia with a relative afferent pupillary defect in the left eye. Fundus examination of the left eye revealed widespread retinal exudation. Ocular B-scan revealed a shallow retinal detachment with subretinal opacities. Case 4 was also classified as a stage 3A disease [16]. Differential diagnosis considered here was FEVR but the presentation in our patient was unilateral.

In all our cases we were unable to examine other members and perform chromosomal studies.

**Table 2. Classification of Coats Disease [16]**

Stage	Findings
1	Retinal vessel telangiectasia and aneurysm
2A	Exudate, extra foveal
2B	Exudate, foveal
3A	Subtotal exudative retinal detachment
3B	Total exudative retinal detachment
4	Secondary glaucoma
5	End- stage (pre-phthisis disease)

### 3. DISCUSSION

Most cases of Coats disease reported in literature are in Caucasians or Asians. Only one case has been reported in Nigeria and one in

Tanzania [9,17]. The 4 cases presented here seen in black Nigerians, are in keeping with the diagnosis of Coats disease and seen over 4 years. Though Coats’s disease commonly occurs in childhood, adult onset has been reported [10]. Our patients were within the age range of 12-25 years. The second patient in our series was an adult but had a history of poor vision since childhood so he could fall within the typical pattern of presentation had he presented earlier.

Coats disease is reported to be commoner in males with a ratio of 3:1 and a unilateral presentation in 95% of cases [2,18,19]. The 4 cases in our series had an equal sex distribution and the disease was unilateral in all cases with the contralateral eye in very good ocular health.

Fundus examination in all four cases revealed widespread retinal exudation with retinal telangiectasia, aneurysm (light bulbs) typical of Coats disease [2,13,16,19,20].

The diagnosis of Coats disease is largely done from clinical examination though some ancillary test may be required to confirm the diagnosis. Ocular B scans in our 3<sup>rd</sup> and 4<sup>th</sup> cases showed exudative retinal detachments and sub retinal exudates respectively (Figs. 3 and 4) which were consistent with the findings on funduscopy. The OCT picture showed in Fig. 3 revealed intra retinal and sub retinal exudates.

The most recent classification Coats disease by Shields et al. was used in our series and all four patients were classified as Stage 3A [16]. Perhaps the late stage at presentation is a factor that points to an underdiagnosis of this disorder which may not be such a rarity after all in Africans. Indeed some may have been misdiagnosed as retinoblastoma and managed as such as was the case in the report from Tanzania [17].

Like other retinal vascular diseases, the armamentarium of treatment for Coats disease has also expanded to include the use of anti-vascular endothelial growth factors. Other modalities of treatment used in the past include cryo therapy, laser photocoagulation, scleral buckling and pars plana vitrectomy [9,14,15,21]. Studies have shown success in terms of regression of the subretinal fluid and re attachment of the retina with the use of anti Vegfs alone or in combination with other treatment modalities [9,14,15,21].

All our patients were offered intravitreal antiVegfs, as an initial form of treatment, however none of them took up the treatment. Subsequent treatment would have been based on our findings after administration of the ant- Vegfs. AntiVegf was offered despite the poor prognosis for vision, to down regulate the vegf load and cause regression and prevent neovascularization and Phtisis bulbi.

#### 4. CONCLUSION

Coat's disease though an uncommon ocular disorder in Nigeria, does exist and may have been underdiagnosed or misdiagnosed. Routine examination of children is pertinent in early diagnosis and prompt treatment to save vision and a good knowledge of its clinical presentation may lead to more case findings.

#### CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

#### ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the authors.

#### COMPETING INTERESTS

Authors have declared that no competing interests exist.

#### REFERENCES

1. Do DV, Haller JA. Coats disease. In Ryan SJ, Ed. Retina. China: Elsevier Mosby. 2004;1417-23.
2. Shields JA, Shields CL, Honavar SG, Demirci H. Clinical variations and complications of Coats' disease in 150 cases: The 2000 Sanford Gifford Memorial lecture. AM J Ophthalmol. 2000;131:561-571.
3. Cербulescu BJ, Stefan C. Coats disease. Oftalmologia. 2003;58(3):21-5.
4. Mishra A, Aggarwal S, Shah S, Negi P, Bharwada R, Desai N. An interesting case of Coats' disease. Med J Armed Forces India. 2015;71(4):384-388.
5. Coats G. Forms of retinal disease with massive exudation. R Lond Ophthalmol Hosp Rep. 1908;17:440-525.
6. Black GC, Perveen R, Bonshek R, Cahill M, Clayton-Smith J, et al. Coats' disease of the retina (unilateral retinal telangiectasis) caused by somatic mutation in the NDP gene: A role for Norrin in retinal angiogenesis. Hum Mol Genet. 1999;8(11):2031-2035.
7. Fernandes BF, Odashiro AN, Maloney S, Zajdenweber ME, Lopes AG, Burnier MN Jr. Clinical-histopathological correlation in a case of Coats' disease. Diagn Pathol. 2006;1:24.
8. Fernandes H, Umashankar T, Richie AJ, Hedge S. Coats disease- disease of the eye rarely encountered by pathologists. Indian J Pathol Microbiol. 2018;61:98-100.
9. Mahmoud AO, Kolawole O, Ibrahim T, Olokoba L. Coats disease in a Nigerian man with a favourable structural outcome following retinal laser photocoagulation. Ann Afr Med. 2013;12:61-2.
10. Smithen LM, Brown GC, Brucker AJ, Yannuzi LA, et al. Coats' disease diagnosed in adulthood. Ophthalmology. 2005;112:1072-1078.
11. Shienbaum G, Tasman WS. Coats disease: A lifetime disease. Retina. 2006;26:422-424.
12. Rishi P, Rishi E, Uparkar M, Sharma T, Gopal L, Bhende P, et al. Coats' disease: An Indian perspective. Indian J Ophthalmol. 2010;58:119-24.

13. Ghorbanian S, Jaulim A, Chatziralli IP. Diagnosis and treatment of Coats' disease: A review of the literature. *Ophthalmologica*. 2012;227:175-182.
14. Villegas VM, Gold AS, Berrocal AM, Murray TG. Advanced Coats' disease treated with intravitreal bevacizumab combined with laser vascular ablation. *Clin Ophthalmol*. 2014;8:973–97.
15. Scheffler AC, Berrocal AM, Murray TG. Advanced Coats' disease. Management with repetitive aggressive laser ablation therapy. *Retina*. 2009;29(1):127.
16. Shields JA, Shields CL, Honavar S, et al. Classification and management of Coats disease: The 2000 proctor lecture. *Am J Ophthalmol*. 2001;131:572-583.
17. Rugwizangoga B, Mwabili T, Scanlan T, Meyer P, Kitinya J. Coats' disease in Tanzania: First case report and literature review. *Afr Health Sci*. 2014;14(3):763–768.
18. Shields JA, Shields CL, Review. Coats disease: The 2001 LuEsther T. Mertz lecture. *Retina*. 2002;22:80-91.
19. Grosso A, Pellegrini M, Cereda MG, Panico C, Staurengi G, Sigler EJ. Pearls and pitfalls in diagnosis and management of coats disease. *Retina* 2015;4:614-23.
20. Ochi R, Sato B, Mimura M, Morishita S, Fukumoto M, Kida T, Sugawara J, Ikeda T. A case of Coats' disease with spontaneous retinal reattachment after total detachment. *Case Reports in Ophthalmology*. 2015;6(2):200–203.
21. Cai X, Zhao P, Zhang Q, Jin H. Treatment of stage 3 Coats' disease by endolaser photocoagulation via a two-port pars plana non vitrectomy approach. *Graefes Arch Clin Exp Ophthalmol*. 2015;253(7):999–1004.

© 2018 Fiebai et al.; This is an Open Access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

*Peer-review history:*  
*The peer review history for this paper can be accessed here:*  
<http://www.sciencedomain.org/review-history/27988>