Original Article

Ophthalmic Horizon
Volume : 16, Issue : I, March 2021; 3-10

Demographic and Clinical Profile of Coats Disease in a Tertiary Eye Care Centre of Bangladesh

Dr. Jasmin Ahmad[1]

[1] Associate Professor, Institute of Community Ophthalmology, Chattogram

ABSTRACT

Aims: To evaluate the demographic and clinical pattern of Coats disease along with outcome after standard management presenting in a tertiary eye care centre.

Methods: A retrospective record based case series of 20 eyes of 20 cases of Coats disease presented at Chittagong Eye Infirmary & Training Complex (CEITC) from January 2008 through June, 2019. The demographic features were noted. Data included detail history of systemic and ocular disease, findings of ocular examination such as, best corrected visual acuity (BCVA), slit lamp biomicroscopy, indirect ophthalmoscopy, Fundus Fluorescein Angiography (FFA). An internet search (medline search) and review of current literature on Coats disease was performed.

Results: Among the total patients, 12 (60%) of them were male and 8 (40%) were female with the mean age of 24.75 (± 1.645) years along with the range of 4 to 55 years. Eleven (55%) of affected eye were right and 9 (45%) were left. Among 20 affected eyes, 16(80%) had 4 quadrant involvement, 2 (10%) had 3 quadrant and remaining 2 (10%) had 2 quadrant involvement. According to severity of disease, 4 (20%) eyes showed 5th stage of disease, half of the eyes 10 (50%) showed stage 3B, 2 (10%) eyes were in stage 3A, 3 (15%) were in 2B and remaining 1 (5%) eye in 2A. Seventeen (85%).eyes presented with severe visual impairment (BCVA less than 6/60) and 3 (15%) presented with moderate visual impairment (BCVA equal or better than 6/60). Intra ocular pressure was normal in all cases.

Conclusion: Most of the cases were in paediatric group and with advance stage of disease. So, parents and physician awareness is needed that may have an impact in early presentation which in turn influence the outcome.

Keywords : Coats disease; telangiectasia, aneurism, intra retinal exudation, exudative retinal detachment.

Introduction

George Coats first reported the disease as uniocular retinal disease in a male child with retinal telangiectasia with massive intra and sub retinal exudations, hemorrhage in 1908[1]. Later on in 1955 Reese[2] coined the name as 'Coats disease' characterized by telangiectasia of retinal vessel that gradually progress to huge intra retinal and sub retinal deposition of exudates and ultimately progress to retinal detachment. Usually young male, other wise healthy suffering from Coat's disease[3,4]; females are affected occasionally at 3:1 ratio. It is unilateral in 75% cases[3,4].

Manuscript Received : 19.02.2021
Revision Accepted : 02.03.2021

Correspondence to:
Dr. Jasmin Ahmad; DO, FCPS
Associate Professor
Institute of Community Ophthalmology
E-mail: drjasmin19@gmail.com

The hallmark of Coats' disease is the development of telangiectasia in the retinal periphery which can lead to intra and subretinal deposits of exudates, gravitate to the macula[5]. Dense macular deposits resulting in poor visual prognosis[5,6]. Another important fundus finding is sub total or total retinal detachment[4,5].

The main features include unilateral painless blurred vision, leukocoria, strabismus, retinal telangiectasias[4,5]. The anterior segment is usually unaffected, but may develop features of anterior ischemic syndrome[3,5]. Vitreous remain clear unless there is haemorrhage[5], proliferative vitreo retinopathy (PVR). Vascular abnormality develops at temporal macula or mid periphery with leakage and deposition of heard exudates[3,5]. The disease may end up with neo-vascular glaucoma, painful blind eye and eventually phthysis bulbi[3,5].

Ophthalmic Horizon
Volume : 16, Issue : I, March 2021; 3-10

The disease is not hereditary though it seems congenital and not associated with systemic disease; mostly (60% to 70%) presents in first decade of life[3,4,5].

Shield et al[6] gave a staging classification of the disease based on clinical features of severity (Table1). Stage 1 is characterised by telangiectasia only; stage 2, by telangiectasia and exudation; stage 3, by telangiectasia, exudation and retinal detachment; stage 4, by total retinal detachment with glaucoma; and stage 5, by advanced end-stage changes and phthisis bulbi.

Table-01: Severity Stage Classification

Stage of disease	Retinal findings
1	Retinal telangiectasia only
2	Telangiectasia and exudation
2A	Extrafoveal exudation
2B	Foveal exudation
3	Exudative retinal detachment
3A	Subtotal detachment
3A1	Extrafoveal
3A2	Foveal
3B	Total retinal detachment
4	Total retinal detachment+Glaucoma
5	Advance end stage

From Shields JA, et al[6]

Differantial diagnosis include Retinoblastoma, Hamartoma and veso proliferative tumor, Retinal detachment, Persistent hyperplastic primary vitreous, Congenital cataract, Norrie's disease/ FEVR, Eale's disease/Vasculitis, Tumour with exudation, Idiopathic juxtafoveal telangiectasia (Type 1)[3,5].

FFA provide early detection of peripheral vascular abnormalities as typical "Light bulb" aneurisms[3,5], vascular tortuacity and leakage, capillary non perfusion (Classic features)[5].

This study was aimed to provide a demographic features, clinical menifestations of Coats disease presenting in a tertiary eye care centre.

Methodology

It is a retrospective, consecutive series of Coats disease study. The medical records of 20 eyes of 20 consecutive patients, who were diagnosed as a case of Coats disease at Chittagong Eye Infirmary & Training Complex (CEITC) from January 2008 through June, 2019 were reviewed retrospectively. This study was approved by institutional review board.

We divided the participants in two age group, pediatric age group (Upto age 18 years) and adult age group. The demographic features like age, sex were noted. The data included recorded history, ocular findings like best corrected visual acuity (BCVA), anterior segment slit lamp examination findings, tonometric findings. Features of Coat's disease were noted from the standard fundus drawing made at the first visit and fundus photographs like telangiectasia and aneurismal abnormalities of vessels, intra retinal and sub retinal hard exudates, exudative retinal detachment. Number of retinal quadrants were affected with lesion along with stage of classification of disease were recorded. Findings of available FFA like "Light bulb" aneurismal vascular abnormalities, telangiectasias, with vascular leakage and area of capillary non perfusion were noted.

Data analysis was done by SPSS software verson 20.00 statistical package(IBM Corpotion).

An internet search (Medline via the PubMed platform) and review of current literature on Coats disease was performed.

Results

20 eyes of 20 patients presented with Coats disease were included in the study. Most, 12 (60%) of them were male and 8 (40%) were female (Fig 1).

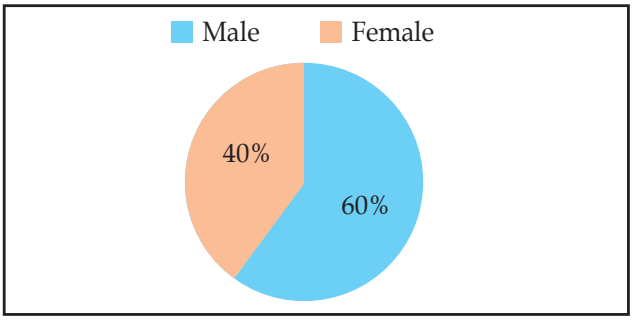

Figure-01: Percentage idstribution of gender of study patients

The mean age of the responders was 24.75 (± 1.645) years with the range of 4 to 55 years; among them 12 (60%) were in paediatric group (upto age 18) years and 8 (40 %) in adult group (Fig 2).

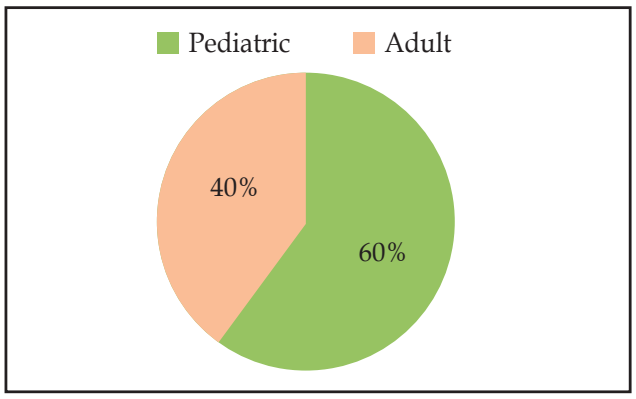

Figure-02: Percentage distribution of age group * paediatric group (upto age 18 years) adult group (More than 18 years of age)

The presenting symptom of all our cases was blurred vision, none of them gave history of squint or leukocoria.

None of our patient had any systemic disease or did not give any positive family history.

Among affected eyes, 11 (55%) were right and 9 (45%) were left (Fig 3).

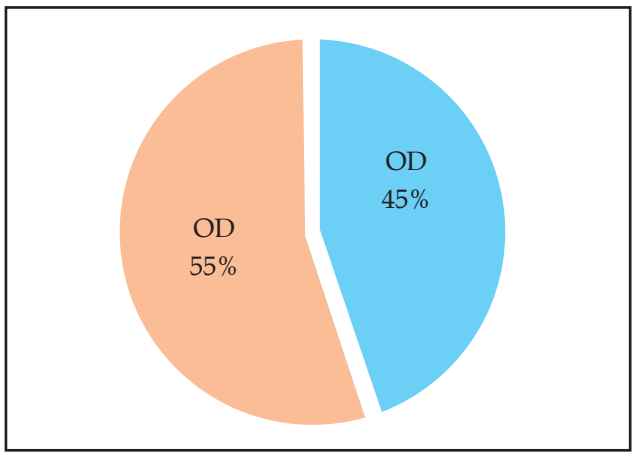

Figure-03: Percentage distribution of eye affected *OD = Oculus Dexter (Right eye), OS = Oculus Sinister (Left eye)

Among 20 affected eyes, 16 (80%) had all 4 quadrants involvement, 2 (10%) had 3 quadrants and remaining 2 (10%) had 2 quadrants involvement. None had disease confined to single quadrant (Fig: 4).

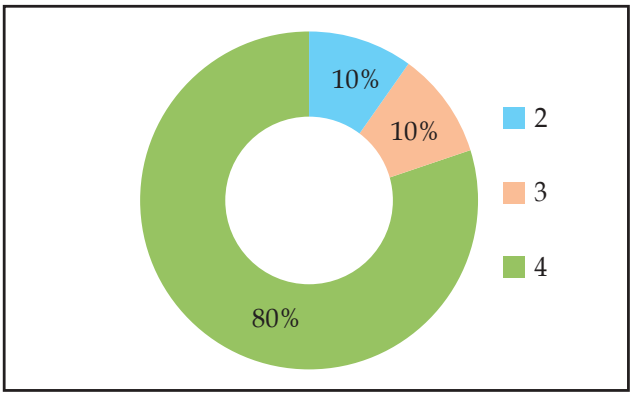

Figure-04: Percentage distribution of number of retinal quadrants affected by disease

According to severity of disease, 4 (20%) eyes showed 5th stage of disease, 10 (50%) eyes showed stage 3B, 2 (10%) eyes were in stage 3A, 3 (15%) were in 2B and remaining 1 (5%) eye in 2A (Fig 5).

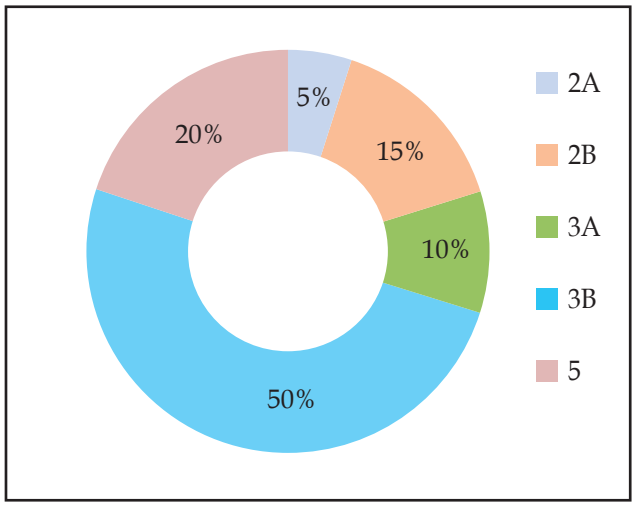

Figure-05: Percentage distribution of different stages of Coats disease of presenting series

None of cases presented with functional vision (6/6 to 6/18) or mild (6/9 to 6/18) visual impairment. Three (15%) presented with 6/60 that was moderate (6/24to 6/60) visual impairment. Seventeen (85%) eyes presented with severe visual impairment (BCVA 3/60 to presence of perception of light), among them 2 (10%) eyes

had BCVA 3/60; 7(35%) had CFCF (counting finger close to face); 3(15%) had hand movement; 5 (25%) had perception of light . None of our cases presented with NPL (No perception of light) (Fig 6).

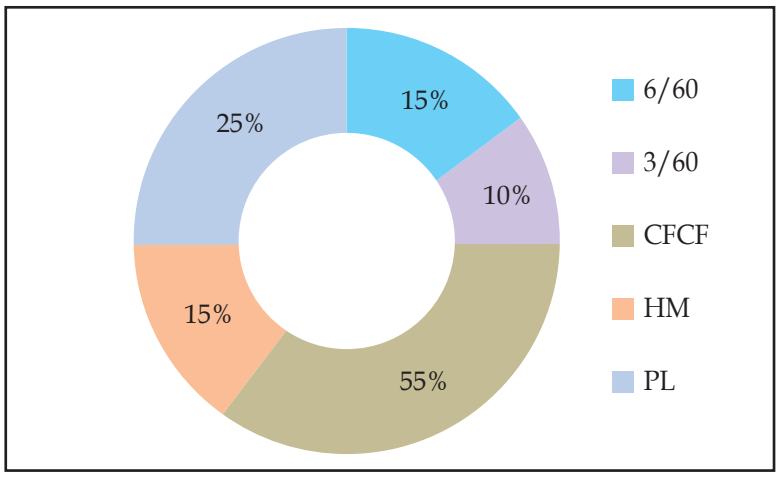

Figure-06: Percentage distribution of presenting BCVA (CFCF: Counting finger close to face; HM: Hand Movement; PL:Perception of light)*

Table-02: Percentage distribution of unaided visual acuity of the respondents

Posterior Segment Findings	Number	Percentage (%)
Telangiectasia	20	100
Intra and subretinal hard exudates	19	95%
Retinal Detachment (sub total)	10	50%
Retinal Detachment (total)	06	30%
Retinal Haemorrhage	03	15%
Macular oedema	01	5%
Macular scar	01	5%
Optic disc neovascularization	01	5%
Vitreous exudates	01	5%
Vitreous haemorrhage	00	0%

NB: Total percentage was more than 100, because some eyes had multiple signs

Anterior segment findings were normal except 3 (15%) had posterior capsular opacity. Posterior segment findings showed all (100%) eyes had telangiectasia mostly involved temporal and infero temporal quadrant (Fig 7,8). The second most common feature was intra and sub retinal exudates found in 19 (95%) eyes; 16 (80%) eyes had exudative retinal detachment (RD), among them 10 (50%) had sub-total RD (Fig 9) and 06 (30%) had total RD (Fig 10) (Table 2) .

Among other retinal features retinal haemorrhage was found in 3 (15%) eyes (Fig 7), optic disc neovascularization in 1 (05%) eye (Fig 11). In this series of eyes 1 (05%) eye had macular oedema (Fig 9) and 1 (05%) had macular scar (Fig 8); 1 (05%) eye had vitreous exudates and no eyes showed vitreous haemorrhage (Table 2).

Ophthalmic Horizon
Volume : 16, Issue : I, March 2021; 3-10

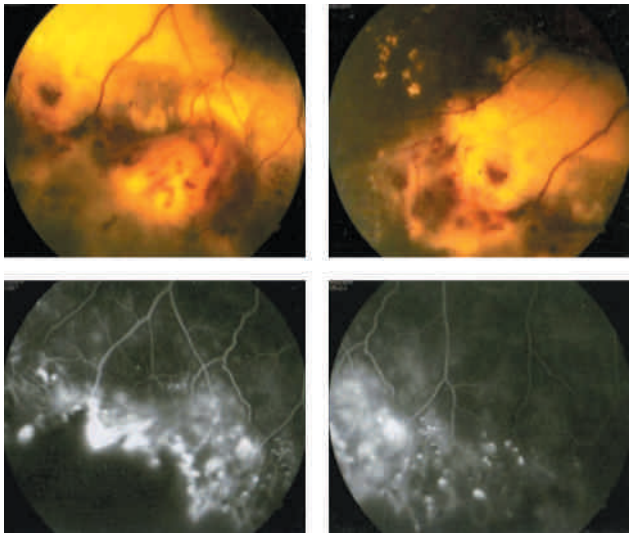

Figure-07: Color fundus photo (upper two) of right eye showed telangiectasia and aneurismal vascular abnormalities with hard exudates and retinal hemorrhage at inferior and temporal periphery; FFA (lower two) showed leakage from telangiectatic vessel and typical "Light bulb" aneurisms was seen with large area of capillary non perfusion at temporal periphery.

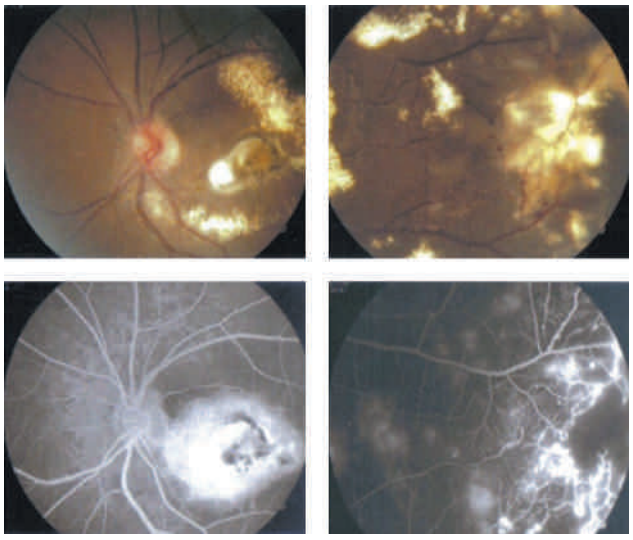

Figure-08: Color fundus photo (upper two) of left eye showed macular scar, telangiectasia and aneurismal vascular abnormalities with hard exudates at temporal periphery. FFA (lower two) showed macular scar staining and leakage from telangiectatic vessel and typical "Light bulb" aneurisms was demonstrated with large area of capillary non perfusion at temporal periphery.

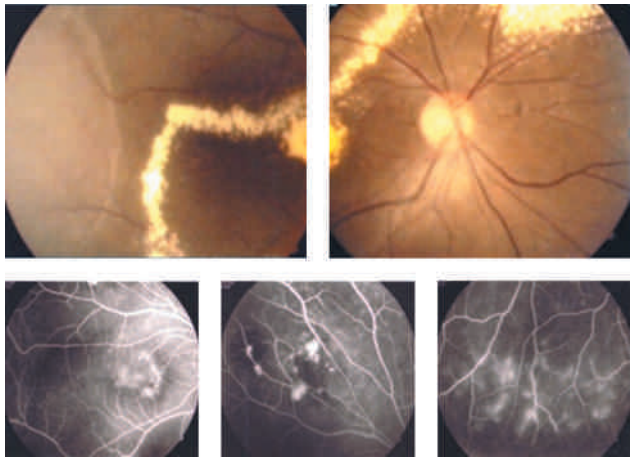

Figure-09: Color fundus photo (upper two)of right eye showed macular oedema with exudates, sub total retinal detachment at temporal periphery. FFA (lower three) showed macular leakage and leakage from telangiectatic vessel and typical "Light bulb" aneurisms was demonstrated supero temporally with leakage from telangiectatic vessel inferiorly.

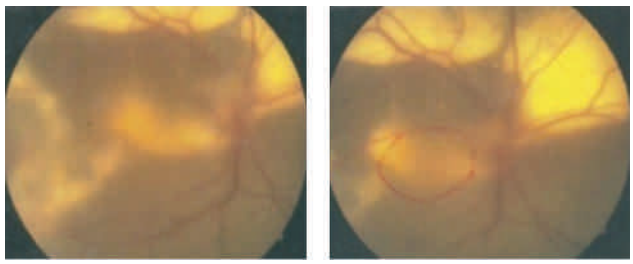

Figure-10: Color fundus photo of right eye showed total retinal detachment with intra retinal exudates, macular deposition of exudates.

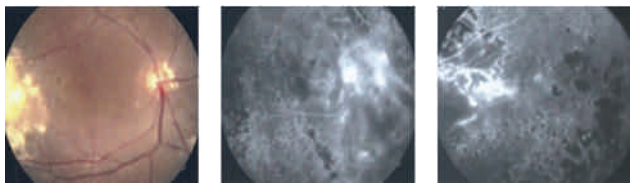

Figure-11: Color fundus photo of right eye showed macular oedema with hard exudates and Optic disc neovascularization on FFA leakage from NVD and telangiectatic vessel at temporal periphery,therer was also capillary non perfusion.

None of our cases developed neovascular glaucoma.

Table-03: Clinical data of 20 patients

SL. No.	Age years	Gender	Eye	Quadrants involved	BCVA	Anterior Segment	Posterior Segment	Stage Classification
1	24	Male	OD	2	3/60	WNL	TV ,HE , sub total RD	3A
2	50	Female	OD	4	CFCF	WNL	TV, HE, ME,	2B
3	10	Male	OD	2	6/60	WNL	Mac Scar	2B
4	18	Male	OD	3	PL	P.S.C	TV , HE , sub total RD	3B
5	18	Female	OS	4	1/60	P.S.C	TV, HE , sub total RD	3A
6	30	Male	OS	3	6/60	P.S.C	TV,HE	2B
7	18	Male	OD	4	HM	WNL	TV, HE	3B
8	16	Female	OD	4	CFCF	WNL	TV, HE , sub total RD	3B
9	50	Female	OS	4	CFCF	WNL	TV, HE, ExRD	3B
10	50	Male	OD	4	CFCF	WNL	TV, HE , Total RD	3B
11	55	Female	OD	4	HM	WNL	ExRD, HE, TV	3B
12	18	Male	OS	4	CFCF	WNL	TV, HE , sub total RD	3B
13	11	Male	OS	4	CFCF	WNL	TV, HE , sub total RD	3B
14	20	Female	OD	4	6/60	WNL	TV, HE , sub total RD	5
15	50	Female	OD	4	HM	WNL	TV, HE , sub total RD	3B
16	13	Male	OD	4	PL	WNL	TV, HE , Total RD	5
17	13	Male	OS	4	PL	WNL	TV, HE , Total RD	3B
18	4	Male	OS	4	PL	WNL	TV, HE , Total RD	5
19	15	Female	OS	4	PL	WNL	TV, HE , Total RD	5
20	12	Male	OS	4	CFCF	WNL	TV, HE , Total RD	2A

* OD = Oculus Dexter (Right eye), OS=Oculus Sinister (Left eye); CFCF: Counting finger close to face; HM: Hand Movement; PL: Perception of light; WNL=With in Normal Limit; PSC = Posterior sub capsular cataract; TV = Telangiectatic vessel; HE = Hard Exudates, RD = Retinal detachment

Discussion

Coats disease is an idiopathic clinical entity of telangiectatic and aneurismal abnormalities of retinal vessel with subsequent deposition of huge intra and or sub retinal exudates and exudative retinal detachment[4,5,7].

The disease is mostly unilateral. Our findings regarding laterality, all cases (100%) were unilateral with 55% right eye and 45% left eye where 95% in Shields et al[4] series were unilateral with 42% right eye and 53% left eye. Nucci et al 8 series of 32 cases; Morris et al[9] series of 55 cases (55% right 45% left), all were unilateral like us.

In our series we got male predominance, 60% were male nearly similar to Shields et al[4] findings that 76% were male, Aparna et al[7] reported 63% were male, 85% were male in Morris et al[12] series.

Most of the patients of Coats disease are diagnosed in the first or second decades of life though it can happen at any age[4]. The mean age of our series was 24.75 (± 1.645) years which is higher than the mean age of Shields et al[4] series which was 11 years.

None of our patient had any systemic disease or did not give any positive family history similar to serieses of Shields et al[4]; Nucci P et al[8]; Morris et al[9] and Budning et al[10].

The presenting symptom of all our cases was blurred vision none of them gave history of squint or leukocoria that is again similar to Shields et al[4] series, their majority of patients presented with decreased vision.

The presenting vision of all our cases were 6/60 to PL (positive percertion of light). None of cases presented with functional vision (6/6 to 6/18) or mild (6/9 to 6/18) visual impairment and none with NPL. In this series 12 (60%) eyes presented with 6/60 to CFCF where as 18% eyes of Shields et al[4] series presented with 6/60 to CFCF; we got 8(40%) had hand movement to perception of light where as Shields et al[4] series showed 58% presented with hand motions to no light perception.

All our cases presented with normal intra ocular pressure. None had neovascular glaucoma, Shields et al[4] series found 8% neovascular glaucoma.

In our series 85% presented with normal anterior chamber findings and 3 (15%) had posterior subcapsular cataract where as 90% patients of Shields et al[4] cases were normal anterior chamber and 8% had cataract, we had double than them.We did not get any cases of iris neovascularization where as Shields et al[4] found 8% and 4% in Morris et al[9] series.

Posterior segment findings included all (100%) eyes had telangiectasia mostly involved temporal and infero temporal quadrant (Fig 7, 8 and 9); Shields et al[4] also showed similar findings. The second most common features were intra and sub retinal exudates were found in 19 (95%) eyes .

We found 16 (80%) eyes of 20 cases had all 4 quadrants involvement which was much more higher than 36% of Dalvin et al[10] series. They reported 15% had 3 quadrants 10% had 2 quadrants involvement, but in our cases 10% had 3 quadrants and 10% had 2 quadrants involved. In our series none had disease confined to single quadrant again similar to the Shields et al[4] findings; where as 16% of Dalvin et al[10] had single quadrant involvement.

We found 16 (80%) eyes had exudative retinal detachment (RD) similar to the findings of Shields et al[4] which was 81%. Among our cases 06 (30%) had total RD but Shields et al[4] series showed 47% total RD (Fig 11, Table 2).

Retinal bleeding is rare sign of Coats disease[4]. We found this in 3 (15%) eyes, similar to the findings of Shields et al[4] that was 13% (Fig 7).

Retinal neovascularization in response of retinal ischemia may develop though it is rare feature We get neovascularization of disc in 1 (05%) eye (Fig 12) similar to 3% of Shields et al[4] series,but none in Aparna et al[7] reported series. In this series, 1 (05%) eye had macular oedema (Fig 10) similar to 4% of Dalvin et al[10] series.

This study series showed 5% eye in 2A; 15% were in 2B; 10% eyes were in stage 3A; 50% eyes showed stage 3B; 20% eyes showed 5th stage of disease ((Fig 5). One report of 55 eyes by Morris et al[9] showed 20% in stage 2A; 42% were in stage 2B; 27% were in stage 3A; 8% were in stage 3B.

We found no eyes in stage 1 and stage 4 where as Morris et al[9] did not find any eyes in stage 1 or stage 5 and only four (7%) eyes in stage 4.

To our knowledge, it is the first report on coats disease in this country.

The limitation of the study: It was a retrospective study with small sample size.

Conclusion

Coats disease is a rare clinical entity but it has a very poor devastating anatomical and functional features resulting in severe visual loss demanding awareness between parents and physician.

Disclosure: The authors report no financial or conflict of interest in any concept or product reported in the present review.

Financial Support & Sponsorship : Nil
Conflicts of interest : There are no conflicts of interest.

References

01. Coats G. Forms of retinal diseases with massive exudation. Roy lond ophthalmol hosp rep. 1908; 17: 440-525.
02. Reese AB. Telangiectasis of the Retina and Coats' Disease: The Eleventh Sanford R. Gifford Lecture. American journal of ophthalmology. 1956 Jul 1;42(1): 1-8.
03. Sigler EJ, Randolph JC, Calzada JI, Wilson MW, Haik BG. Current management of Coats disease. Survey of ophthalmology. 2014 Jan 1;59(1):30-46.
04. Shields JA, Shields CL, Honavar SG, Demirci H. Clinical variations and complications of Coats disease in 150 cases: the 2000 Sanford Gifford Memorial Lecture. American journal of ophthalmology. 2001 May 1;131(5):561-71.
05. Sen M, Shields CL, Honavar SG, Shields JA. Coats disease: an overview of classification, management and outcomes. Indian journal of ophthalmology. 2019 Jun;67(6):763-71.
06. Shields JA, Shields CL, Honavar SG, Demirci H, Cater J. Classification and management of Coats disease: the 2000 Proctor Lecture. American journal of ophthalmology. 2001 May 1;131(5):572-83.
07. Aparna Ramasubramanian,Carol L Shields. Bevacizumab for Coats' disease with exudative retinal detachment and risk of vitreoretinal traction. Br J Ophthalmol. 2012 Mar;96(3):356-9. doi: 10.1136/bjophthalmol-2011-300141.
08. Nucci P, Bandello F, Serafino M, Wilson ME. Selective photocoagulation in Coats' disease: ten-year follow-up. European journal of ophthalmology. 2002 Nov;12(6):501-5.
09. Morris B, Foot B, Mulvihill A. A population-based study of Coats disease in the United Kingdom I: epidemiology and clinical features at diagnosis. Eye. 2010 Dec;24(12):1797-801.
10. Dalvin LA, Udyaver S, Lim LA, Mazloumi M, Atalay HT, Khoo CT, Shields CL. Coats disease: clinical features and outcomes by age category in 351 cases. Journal of pediatric ophthalmology and strabismus. 2019 Sep 24;56(5):288-96.