IDIOPATHIC JUXTAFOVEOLAR TELANGIECTASIS TYPE 1A. A CASE REPORT

Pandey A[1], Anand A[2], Singh S[3], Chandra A[4]

ABSTRACT

Idiopathic juxtafoveolar telangiectasisis a descriptive term for various disease entities presenting with incompetence, ectasia, and/or irregular dilations of the capillary network affecting only the juxtafoveolar region of one or both eyes. A 65 years male presented with the chief complaint of painless progressive diminution of vision in his left eye for 6 months' duration. Past ocular, surgical, medical, drug and family history was non contributory. The patient was non hypertensive and non diabetic. His best corrected visual acuity was 6/60 right eye and 6/36 left eye. Right eye, fundus was grossly normal whereas, on the left eye, few microaneurysms and circinate pattern of hard exudates were noticed on the perifoveolar region. Fundal reflex was dull and the foveal depression was absent indicating thickening. Fluorescein angiography showed clusters of telangiectatic vessels around exudates, hyperfluorescent dots of microaneurysms with a circinate pattern of leakage in the late phase. Optical coherence tomography showed macular thickening of 487 micrometer with cystoid changes. Injection Bevacizumab 0.2ml was given after final diagnosis of Left Eye Idiopathic Juxtafoveolar Telangiectasis Type 1A. Three types of idiopathic juxtafoveolar telangiectasis has been defined. Its pathophysiology is also less understood and the treatment modalities are not established yet.This case was an incidental finding, as these patients do not oftenly have profound diminution of vision, unless neovascularization has occurred and the clinical features too are very subtle.

KEYWORDS: Idiopathic juxtafoveolar telangiectasis, Microaneurysm, Hard exudate

1. Post Graduate Resident, Department of Ophthalmology, Universal College of Medical Sciences & Teaching Hospital, Bhairahawa, Nepal
2. Vitreo Retinal Surgeon, Lumbini Eye Institute, Bhairahawa, Nepal
3. General Ophthalmologist, Lumbini Eye Institute, Bhairahawa, Nepal
4. Associate Professor, Department of Ophthalmology, Universal College of Medical Sciences & Teaching Hospital, Bhairahawa, Nepal

For Correspondence
Dr. Araniko Pandey
Post Graduate Resident,
Department of Ophthalmology,
Universal College of Medical Sciences and Teaching Hospital
Bhairahawa, Nepal
E-mail: aranikopandey@gmail.com

INTRODUCTION

Retinal capillary telangiectasia or dilated retinal capillaries are usually the result of antecedent retinal vascular inflammatory or occlusive disease. In the macula, diabetic retinopathy, hypertension, venous occlusion, inflammatory diseases, and blood dyscrasias are the usual causative factors. However,there are other forms of telangiectasia that develop in the macula and perifoveolar areas without a known cause.[1]

Most retinal telangiectases are acquired secondary to local or systemic conditions, as mentioned above. Primary retinal telangiectasia is found in Coats' disease, Leber's miliary aneurysms (a localized, less severe form of Coats' disease), idiopathic juxtafoveal telangiectasia, and other angiomatous diseases.[2]

Idiopathic juxtafoveolar telangiectasis (also known as idiopathic parafoveal, perifoveal or macular telangiectasia or telangiectasis) is a descriptive term for various disease entities presenting with incompetence, ectasia, and/or irregular dilations of the capillary network affecting only the juxtafoveolar region of one or both eyes.[3]

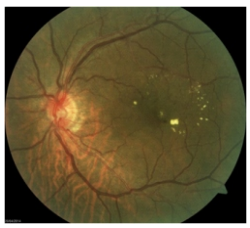
CASE REPORT

A 65 years male from Nepal, presented to the Vitreoretina clinic at Lumbini Eye Institute with the chief complaint of painless progressive diminution of vision in his left eye for 6 months' duration. Diminution of vision was evenly present within his visual field with no associated symptoms.

Past ocular, surgical, medical, drug and family history was non contributory. Also, the patient was non hypertensive and non diabetic.

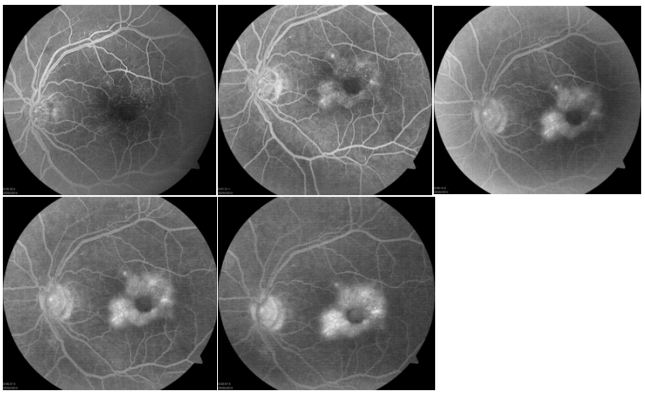
His best corrected visual acuity was 6/60 right eye and 6/36 left eye.The patient was in good general health with no noticeable gross abnormalities. Thorough anterior and posterior segment examination was done which revealed, nuclear sclerosis grade 2 with posterior subcapsular cataract in his right eye and nuclear sclerosis grade 1 in his left eye. Right eye, fundus was grossly normal whereas, on the left eye, few microaneurysms and circinate pattern of hard exudates were noticed on the perifoveolar region. Fundal reflex was dull and the foveal depression was absent indicating thickening. (Figure 1)

Figure 1: Fundus Picture, Left Eye



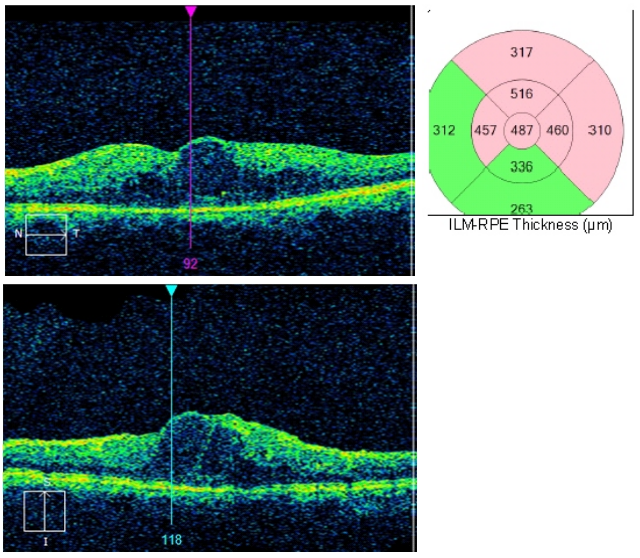
Fluorescein fluorescein angiography (FFA)was performed with 3 ml of 2% fluorescein sodium injected through a peripheral vein and the photographs were taken with a Carl Zeiss Meditec non-mydriatic Visucam Pro NM fundus camera. It showed clusters of telangiectatic vessels around exudates, hyperfluorescent dots of microaneurysms with a circinate pattern of leakage in the late phase (Figure 2).

Figure 2: Serial Picture of FFA, left eye



Optical coherence tomography (OCT) was done using, CIRRUS™ HD-OCT 500from Carl Zeiss Meditec, which showed macular thickening of 487 micrometer with cystoid changes (Figure 3).

Figure 3: OCT macula of left eye



Injection Bevacizumab 0.2ml was given after final diagnosis of Left Eye Idiopathic Juxtafoveolar Telangiectasis Type 1A. Left eye was planned for cataract surgery with intraocular lens implantation.

DISCUSSION

The term *idiopathic juxtafoveolar retinal telangiectasis* (IJFT) was coined by Gass and Oyakawa in 1982, who proposed the first classification of these entities into four groups based largely on their

clinical and fluorescein angiographic (FA) features. In 1993, Gass and Blodi further updated this classification, by subdividing IJFT into three distinct groups I, II, and III (also known as groups 1, 2, and 3), with two subgroups in each (A and B), based on demographic difference or clinical severity. (Table 1)[3]

Table 1: Gass and Blodi Classification of IJFT

Table 3. Classification of Macular Perifoveal Telangiectasia

IJRT*	IMT† Classification	IMT† Description
Group 1	Type 1	Aneurysmal telangiectasia
1A: visible and exudative IJRT	NA	NA
1B: visible, exudative, and focal IJRT	NA	NA
Group 2	Type 2	Perifoveal telangiectasia (nonproliferative or proliferative)
2A: occult and nonexudative IJRT	NA	NA
Stage 1: occult telangiectatic vessels	NA	NA
Stage 2: loss of transparency without clinically evident telangiectatic vessels	NA	NA
Stage 3: prominent dilated right-angle retinal venules	NA	NA
Stage 4: retinal pigment hyperplasia into the retina	NA	NA
Stage 5: SRN from proliferation of intraretinal capillaries	NA	NA
2B: juvenile occult familial IJRT	NA	NA
Group 3	NA	NA
3A: occlusive IJRT	NA	NA
3B: occlusive IJRT with CNS vasculopathy	NA	NA

According to Gass, IJFT can be divided into three groups based upon phenotype.Type I is typically a unilateral disease characterized by parafoveal dilation of capillaries, microaneurysms, leakage, and lipid deposition; type II is the most common form of IJFT and typically presents with bilateral juxtafoveal telangiectasias with minimal exudate; type III is extremely rare and is characterized by occlusive telangiectasia.[4]

The MacTel Project, which is under way in 26 centers around the world, set out to observe the natural disease course, follow the progression and conduct a genetic study of patients and families to identify genes and genetic variants.[5] The very subtle nature of the early findings in MacTel means the diagnosis is often missed by optometrists and general ophthalmologists. No new information has emerged about the condition since its clinical features were first well described by Dr. J. Donald Gass in 1982. There is much work to be done to understand the disease better, to raise its profile and to search for treatments.[6]

No treatment has been established. Laser treatment is not helpful. Anti-vascular endothelial growth factor (VEGF) drugs for subretinal neovascularization can be used for subretinal neovascularization and these patients can do well. Recent publications on intravitreal anti-VEGF injections, namely bevacizumab, report on possible short-term visual acuity increase in some cases of IJFT IIA.[7,8]

Hence, we report this case of IJFT, which was an incidental finding, as these patients do not oftenly have profound diminution of vision, unless neovascularization has occurred and the clinical features too are very subtle. Its pathophysiology is also less understood and the treatment modalities are not established yet. Plentiful of research work is going on worldwide regarding IJFT, and we must be aware of "the calling" to take up this challenge.

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