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Characterization of idiopathic macular telangiectasia type 2 by fundus fluorescein angiography in Indian population

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PURPOSE. *To characterize the variations in fundus fluorescein angiography (FFA) features in idiopathic macular telangiectasia (IMT) type 2.*
METHODS. *Retrospective observational study. The authors included all the patients with IMT type 2 who underwent FFA during the period from January 2004 to December 2005. The main outcome measures were subretinal neovascularization and number and distance of telangiectasia from the center of the foveal avascular zone.*
RESULTS. *The mean age of 21 patients included in the study was 53.0±7.7 years. There were 11 women and 10 men. Six of the 21 patients had a history of diabetes mellitus. Subretinal neovascularization (SRN) was observed in 10 (23.8%) eyes. Subretinal plaque of pigment hyperplasia in the macula was found in 7 (21.9%) eyes, and crystalline yellowish deposits on the retinal surface were seen in 19 (59.4%) eyes. Only one eye had visible retinal telangiectasia. Most had more than 10 telangiectatic lesions in the macular area and the temporal macula was most commonly involved. The mean distance from the center of the foveal avascular zone (FAZ) up to which telangiectasia could be observed was 1340±400 μm. The maximum distance from the FAZ at which telangiectasia was identified was 2530 μm.*
CONCLUSIONS. *In IMT type 2, telangiectasia may be seen farther from the parafoveal area. (Eur J Ophthalmol 2008; 18: 587-90)*

KEY WORDS. *Fundus fluorescein angiography, Idiopathic macular telangiectasia type 2*

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INTRODUCTION

In 1993, Gass and Blodi established a classification of cases that had retinal capillary telangiectasia in the perifoveal area without any known cause (1). They described these cases as idiopathic juxtafoveolar retinal telangiectasis, and divided them into groups and stages. Among the three groups, type 2 is the most common and is further subdivided into two subgroups, A and B. There are very few cases of Type 2B that have been reported in the literature, which is characterized by the young age at onset, and may have a familial occurrence.

Recently, Yannuzzi et al coined a new term for this disease based on newly recognized clinical and imaging characteristics, referring to the disease as idiopathic macular telangiectasia (IMT) (2). They proposed a new classification which does not include the type 3 group from the original classification due to its rarity, and also because macular ischemia is the primary abnormality in this group. The original classification of type 1 (aneurysmal telangiectasia) and type 2 (perifoveal telangiectasia) has been retained in the new classification. Type 2 has been further subdivided into nonproliferative (exudation and foveal atrophy) and proliferative disease (subretinal neovascular-

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ization [SRN] or fibrosis).

Type 2 IMT is more common than type 1 disease. It is characterized by onset in middle age with bilateral occurrence of telangiectasia mainly in the temporal perifoveal area (2). The associated macular changes in type 2 IMT include perifoveal loss of transparency, RPE atrophy, pigment epithelial hyperplasia, crystalline deposits at the vitreoretinal interface, right-angled vessel, and SRN. Type 2 IMT can be associated with visual impairment either due to foveal atrophy or SRN. Based on this new classification, we retrospectively analyzed the fundus photographs and fundus fluorescein angiograms (FFA) to determine the nature of lesions, and to specifically look for the number and location of telangiectatic vessels from the fovea.

METHODS

Medical records of patients clinically diagnosed with retinal telangiectasia by International Classification of Diseases (ICD-9) and who underwent FFA from January 1, 2004, through December 31, 2005, were retrieved. Demographic data and medical history including diabetes mellitus were recorded. Fundus photographs and FFAs of the patients were reviewed. Further, classification of type 2 IMT patients into subgroups of nonproliferative and proliferative perifoveal macular telangiectasia was performed based on Yannuzzi's modification of the Gass-Blodi classification.

Digital FFA (FS 180, Zeiss, Jena, GmbH, Germany) of each patient were reviewed. The number of telangiectasia in each eye was noted and grouped into categories of less than 5, 5 to 10, 10 to 20, or >20 telangiectasias. The location of the telangiectasia with respect to the fovea – temporal, superior, nasal, and inferior – was recorded. The distance of the most peripheral (farthest) telangiectasia from the center of the fovea was also noted. For this purpose, a frame of the angiogram showing the central 30-degree field was used, and the distance was calculated in microns using the Zeiss Visupac® software. Late phase angiograms were also evaluated for any leakage of dye into the foveal avascular zone (FAZ).

Color fundus photographs were reviewed for retinal hemorrhage, pigment hyperplasia, retinal pigment epithelial atrophy, crystalline deposits at vitreoretinal interface, and subretinal neovascularization or disciform scar. Mathematical and statistical analysis was performed with Microsoft Excel.

RESULTS

Fifty-two eyes of 26 patients were newly diagnosed with type 2 IMT and had undergone FFA during the study period. Five patients were not included in the study due to poor FFA quality that precluded appropriate analysis.

Demographic data

Forty-two eyes of 21 patients with type 2 IMT were included. The mean age of patients was 53.0±7.7 years. The mean age of diabetics with macular telangiectasia was 58.8±8.0 years compared to 50.5±6.6 years among non-diabetics. The mean age of patients without SRN was 52.4±7.8 years, whereas the mean age of patients with SRN was 56.2±8.1 years. There were 11 women and 10 men. Six out of the 21 patients had a history of diabetes mellitus. All the patients were Asian Indians.

Fundus fluorescein angiogram

Proliferative changes (Tab. I).—SRN was seen in 9 out of 52 eyes (17.3%) and 1 out of 52 eyes (1.9%) had a disciform scar (Fig. 1). The lesion types were variable. Six out of 9 eyes (66.7%) had a predominantly classic lesion, 2 out of 9 eyes (22.2%) minimally classic, and 1 out of 9 eyes (11.1%) had an occult lesion. Three patients out of 7 patients (42.8%) had bilateral proliferative changes, including one eye with a disciform scar, whereas 4 out of 7 patients (57.8%) had unilateral SRN.

Nonproliferative changes (Tab. I).—Of the remaining 32 eyes without SRN, 3 eyes of 2 patients had dot hemorrhage in the retina, and of these 2 patients, 1 had a history of diabetes mellitus. Subretinal plaque of pigment hyperplasia in the macula was observed in 7 (21.9%) eyes

TABLE I - FUNDUS FEATURES OF PATIENTS WITH TYPE 2 IDIOPATHIC MACULAR TELANGIECTASIA

Fundus feature	No. (%) of eyes	No. (%) of patients
Proliferative change	10 (17)	7 (33.3)
Subretinal pigment plaque	7 (11.5)	5 (23.8)
Retinal pigment epithelium atrophy	21 (35)	11 (52.4)
Crystalline yellowish deposits	19 (32)	10 (47.6)
Visible retinal telangiectasia	1 (1.5)	1 (4.7)
Diabetic retinopathy	2 (3)	1 (4.7)

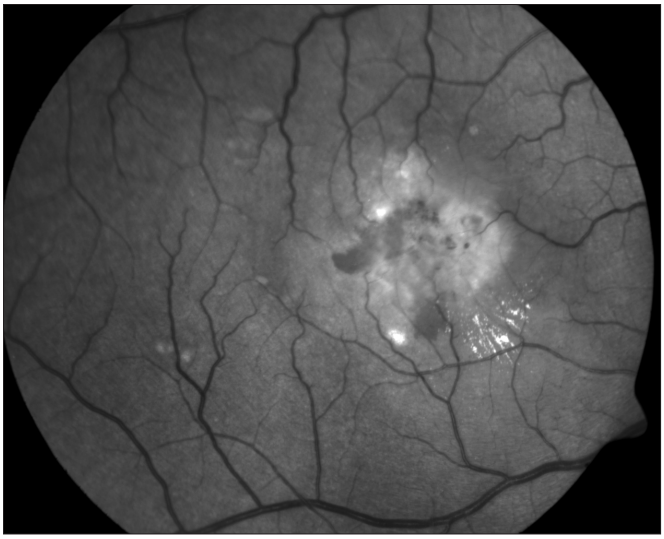

Fig. 1 - *Red-free photograph of a patient with type 2 idiopathic macular telangiectasia showing subretinal hemorrhage and exudates due to subretinal neovascularization.*

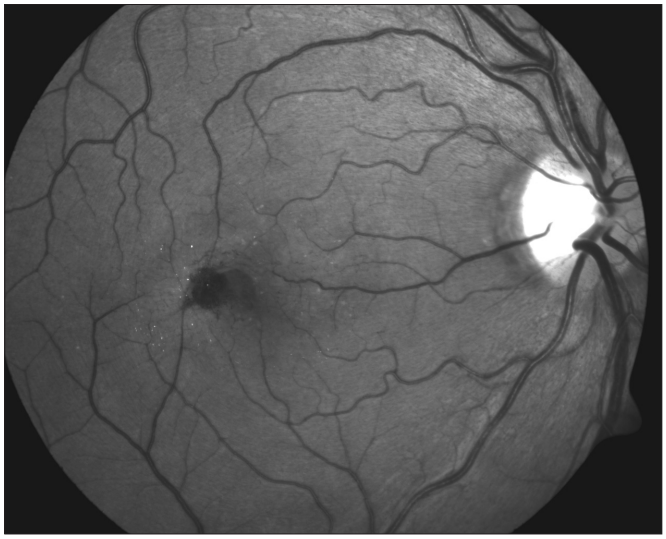

Fig. 2 - *Red-free photograph of the right eye of a patient with subretinal pigment plaque with crystalline deposits on the vitreoretinal interface.*

(Fig. 2). Retinal pigment epithelial atrophic changes were identified in 21 (65.6%) eyes. Crystalline yellowish deposits on the retinal surface were present in 19 (59.4%) eyes (Fig. 3). Among the 6 patients with diabetes mellitus, one patient had minimal nonproliferative diabetic retinopathy with only a few microaneurysms bilaterally. Only one of the 42 eyes with type 2 IMT had visible retinal telangiectasia.

A majority of the eyes had >10 telangiectatic lesions. The temporal macula was involved in 31/32 (96.8%) eyes. The inferior macula was least commonly affected (11 out of 32 eyes, 34.3%), and was the predominant site of lesion in only one eye. The mean distance from the center of the FAZ up to where the telangiectasia were observed was 1340±400 mm. The maximum distance from the FAZ to telangiectasia observed in an eye was 2530 mm. Leakage of fluorescein into the FAZ was present in 19 eyes.

There was no correlation between age and number of lesions, distance of the lesions from the FAZ or SRN, although the number of subjects in this study is too small to provide any meaningful statistical analysis.

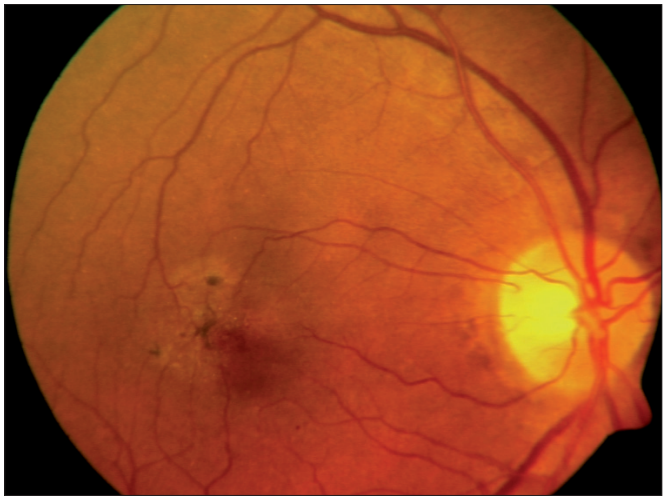

Fig. 3 - *Color fundus photograph of a patient with type 2 idiopathic macular telangiectasia showing perifoveal halo, retinal pigment epithelium (RPE) atrophic changes, RPE hyperplasia, and yellowish crystalline deposits on the retina.*

DISCUSSION

Idiopathic macular telangiectasia, Leber miliary aneurysms, and Coats disease are three types of retinal telangiectasias that may represent a continuum of the same disease. Group 1 IMT patients are primarily male; the telangiectasia is unilateral in most cases and easily visible. It is suspected that this form of IMT may be a mild presentation of Coats disease. Leber miliary aneurysms represent a more severe form of telangiectasia. The nomenclature of parafoveal telangiectasia has been recently changed to macular telangiectasia (2). The present study demonstrates that the telangiectasia can be present

far from the fovea, but still within the anatomic macula (3600 µm). Hence, the term macular telangiectasia seems to be appropriate.

In this study, SRN was seen in approximately one-fourth of the patients. This finding is higher than the reported incidence in the literature, which is from 14% to 17% (2, 3). The present study reveals a slightly higher incidence of SRN in Indian patients with IMT which may be due to geographic variation. However, this difference should be interpreted with caution as this study was performed at a tertiary care center, introducing selection bias, and due to the small sample size of the study. The sample size was also inadequate to perform a statistical test of significance among the age difference seen in patients with respect to diabetic status and SRN. The higher age at diagnosis of patients with diabetes mellitus may be due to a referral delay; however, this could not be evaluated in the present study. Retinal hemorrhage and pigment hyperplasia were uncommon in this study, whereas RPE atrophic changes were seen in majority of the patients.

Crystalline deposits at the vitreoretinal interface have been described as inconsistent but characteristic findings in IMT (2). The incidence of this feature has not been well reported in the literature. We found crystalline deposits in about 60% or our nonproliferative eyes with IMT. Subretinal plaques of pigmentation are characteristic, but only occurred in approximately 20% of our eyes with nonproliferative lesions.

We used the lesion size calculation program in the Zeiss Visupac software to measure the distance of the farthest telangiectasia from the center of the FAZ. The distance of the telangiectasia from the center of FAZ was large but limited to within the arcades, although changes of a lesser degree in the retinal capillaries have been described in the peripheral fundus as well by light and electron microscopy where there were no clinically detectable abnormalities (4). Most of our eyes with type 2 IMT had more than 10 telangiectatic lesions, and a significant percentage had more than 20 lesions. The smaller number of lesions and lesser distance from the center of the FAZ could represent an earlier stage of the disease, and more lesions along with a larger spread of the lesions could signify a later stage in the natural history of the disease. Prospective studies to study the natural course of IMT, such as the Mactel study (https://web.emmes.com/study/mactel/), may be able to confirm this hypothesis.

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