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A CASE OF IDIOPATHIC PERIFOVEAL TELENGIECTASIA

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Abstract : Idiopathic perifoveal telangiectasia or macular telangiectasia type 2 is a rare condition of unknown pathogenesis initially described by Gass and Oyakawa in 1982. We here report a case of Idiopathic perifoveal retinal telangiectasia in a 50 years old female. In Melbourne collaborative cohort study incidence of Idiopathic macular telangiectasia type 2 ranged from 0.0045 percentage to 0.022 percentage and in Beaver Dam eye study prevalence, 0.1 percentage.

Keyword : Idiopathic perifoveal telangiectasia, muller cells, subretinal neovascularization

INTRODUCTION: Retinal capillary telangiectasia is relatively common. Most cases are secondary to another retinal condition, typically involving inflammation or vascular occlusion; examples include diabetic retinopathy and retinal vein occlusion. Primary retinal telengiectasis comprises a group of rare, idiopathic, congenital or acquired retinal vascular anomalies characterized by dilatation and tortuosity of retinal blood vessels, multiple aneurysms, vascular leakage and deposition of hard exudates. The vascular malformations often progress and become symptomatic later in life4. Retinal telangiectasia localized to foveal region in one or both eyes is termed as macular telangiectasia1. There are two basic and distinct forms in Idiopathic macular telangiectasia. Idiopathic macular telengiectasia type 1 otherwise called as aneurysmal telangiectasia2 is a developmental or congenital usually unilateral vascular anomaly, may be a part of the larger spectrum coat's disease1. Idiopathic macular telangiectasia type 2 otherwise termed as juxtafoveal or perifoveal telangiectasia2 is a presumably acquired bilateral form found in middle aged and elderly people with characteristic alterations of the macular capillaries and neurosensory degeneration1.

CASE HISTORY: A 50 years old female, housewife presented with gradual impairment of central vision and metamorphopsia in both eyes since 1 month. No h/o any systemic ailments. General examination and systemic examination were within normal limits. Her visual acuity at the time of presentation was 6/60 NIG NIP in right eye and 6/36 NIG NIP in left eye. On ocular examination both eyes anterior segment was normal. Lens was clear. The intraocular pressure by Goldmann applanation tonometry was 16 mm of Hg in right eye and 14 mm Hg in left eye. On dilated fundus examination, right eye media was clear. Disc was normal with cup disc ratio of 0.3 and major vessels were normal. Dilated capillaries in the macula, blunting of foveal reflex and loss of retinal transparency with grayish discoloration in the perifoveal region (figure 1). Peripheries found to be normal. Left eye fundus examination, Disc was normal with cup disc ratio of 0.3 and major vessels were normal. Dilated capillaries in the macula, blunting of foveal reflex and loss of retinal transparency with grayish discoloration in the perifovea were noted (figure2).

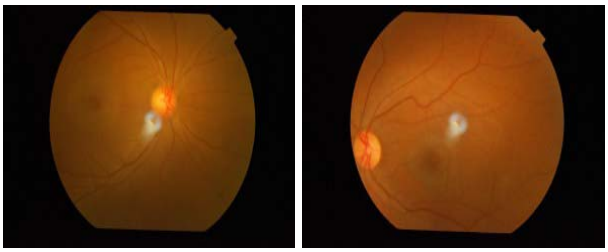

Figure 1 of 50 yrs female IP NO 471560 Fundus picture of right eye Figure 2 of 50 yrs female IP NO 471560 Fundus picture of left eye

Clinical Diagnosis of Idiopathic perifoveal telangiectasia was made. Fundus fluorescein angiography and Optical Coherence Tomography were done to the patient. On Fundus Fluorescein Angiography, right eye there was normal filling pattern of disc and major vessels, telengiectatic capillaries was noted in the macula. There was late leakage from perifoveal capillaries. In left eye there was normal filling pattern of disc and major vessels, telangiectatic capillaries were noted in the macula. There was late leakage from perifoveal capillaries.

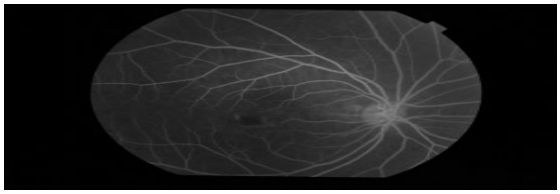

Figure 3 of 50 yrs female IP NO 471560 fundus fluorescein angiography of right eye early phase

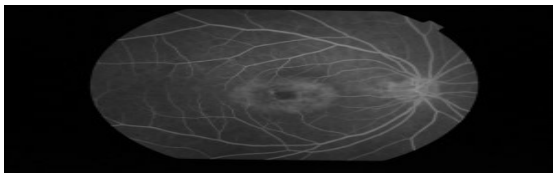

Figure 4 of 50 yrs female IP NO 471560 fundus fluorescein angiography of right eye midphase

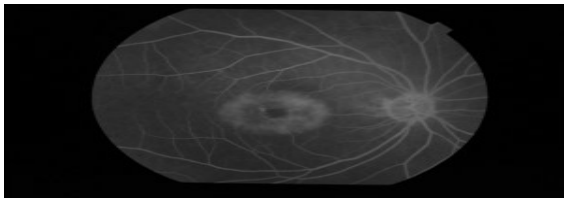

Figure 5 of 50 yrs female IP NO 471560 fundus fluorescein female IP NO 471560 fundus fluorescein

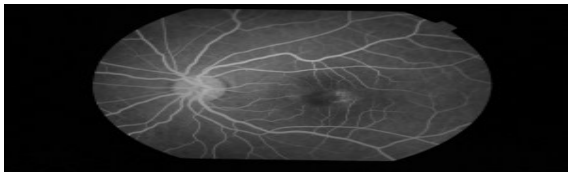

Figure 6 of 50 yrs angiography of right eye late phase angiography of left eye early phase

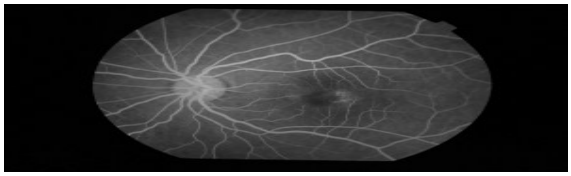

Figure 7 of 50 yrs female IP NO 471560 fundus flourescein NO 471560 fundus fluorescein

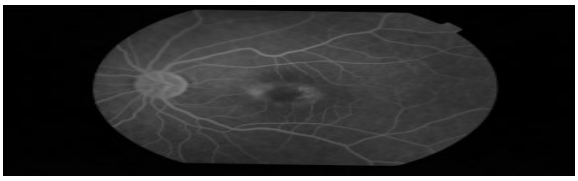

Figure 8 of 50 yrs female IP angiography of left eye midphase angiography of left eye late phase

On Optical Coherence Tomography, right eye enlargement of foveal pit with thinning of retinal layers and hyporeflective intraretinal cavitation were noted (figure 5). Left eye enlargement of foveal pit with thinning of retinal layers and hyporeflective intraretinal cavitation were noted (figure 6).

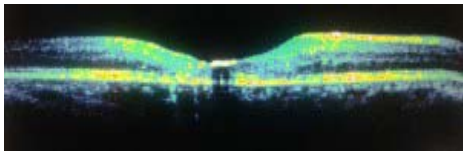

Figure 9 of 50 yrs female IP NO 471560 optical coherence tomography of right eye tomography of right eye

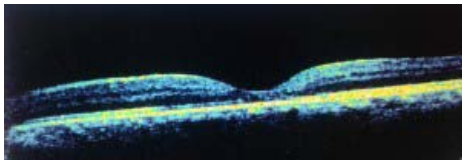

Figure 10 of 50 yrs female IP NO 471560 Optical coherence tomography of right eye tomography of left eye

Diagnosis of Idiopathic perifoveal telangiectasia was confirmed.

Treatment: There is no definite treatment for idiopathic perifoveal telangiectasia. Patient is on monthly follow up to look for the progression of the disease. She was advised to report immediately if she develops any sudden drop in vision which is usually secondary to subretinal neovascularization. If subretinal neovascularization occurs later, that is treated with intravitreal anti vascular endothelial growth factor agents (Ranibizumab or Bevacizumab).

DISCUSSION

Idiopathic macular telangiectasia is otherwise known as idiopathic juxtafoveal telangiectasia includes several, different vascular diseases affecting the capillaries of the posterior pole.

GASS CLASSIFICATION: In 1968 Gass named the Idiopathic macular telangiectasia as Idiopathic juxtafoveolar retinal telangiectasia. He classified it into 3 groups based on historical, biomicroscopic and angiographic findings.

Group 1A: Unilateral congenital parafoveal telangiectasia, which typically occurs in men.

Group 1B: Similar to group 1A with tiny area of capillary telangiectasia confined to one clock hour at the edge of the foveal avascular zone.

Group 2A: Bilateral, acquired, idiopathic parafoveal telangiectasia occurring in the fifth and sixth decades.

Group 2B: Similar to group 2A with subretinal neovascularization.

Group 3A: (Occlusive telengiectasia) Telengiectasis, vascular occlusion and minimal exudation.

Group 3B: Similar to group 3A with neurological disease.

YANNUZZI CLASSIFICATION: In 2006 Yannuzzi introduced the name Idiopathic macular telangiectasia. He simplified the classification into two types by eliminating rare forms of the disease (2B,3A&3B)2.

Type 1: Aneurysmal telangiectasia (1A&1B)

Type 2: Perifoveal telangiectasia

ANEURYSMAL TELENGIECTASIA also known as visible and exudative idiopathic juxtafoveal retinal telangiectasia under Gass and Blodi Classification, considered as a form of coat's disease that is limited to the macula. It is seen in middle aged males typically present with a unilateral mild blurring of central vision. Slit lamp biomicroscopy reveals arteriolar, capillary and venular aneurysms and telangiectasia which is found in both the superficial and deep retinal capillary circulations. Vascular abnormalities

are noted within 2 disc diameters of center of the fovea, primarily along the temporal parafoveal region, often associated with localized capillary leakage and lipid exudate. Visual loss occurs due to accumulation of foveolar exudate and cystoid macular edema2. Optical Coherence Tomography demonstrates retinal thickening, cystoid macular edema and localized exudative retinal detachment. Fundus Fluorescein Angiography shows telangiectasia and multiple capillary, venular and arteriolar aneurysms and late leakage. There is minimal non-perfusion. Treatment by laser photocoagulation to areas of leakage may be beneficial in preventing visual loss from chronic cystoid macular edema and exudation. Recently intravitreal vascular endothelial growth factor inhibitors may reduce macular edema and improve vision4. PERIFOVEAL TELENGIECTASIA also known as occult and nonexudative idiopathic juxtafoveal retinal telangiectasia under the Gass and Blodi classification2, a bilateral disease occurs in both males and females with slight predilection for the female sex1. Patients typically present in the fifth and sixth decades of life with mild blurring of vision that can progress to foveolar atrophy or subretinal neovascularization. It has the worst visual prognosis. Idiopathic macular telangiectasia type 2 is progressive vasculopathy classified into nonproliferative and proliferative stages but it is rare to observe the complete vasogenic sequence. As vessels course towards fovea, they usually decrease in diameter but in macular telangiectasia type 2 they dilate and make a right-angle turn, diving into deep retinal layers.

Slitlamp biomicroscopy reveals symmetric blunting of foveal reflex, greyish loss of juxtafoveolar retinal transparency, initially temporal to and later surrounds the fovea4, minimal serous exudation and no lipid deposition. Glistening white or yellow-white crystalline deposits may be noted in superficial parafoveal retina in 40% of patients. In some patients, small yellow lesion, 1/3 disc diameter in size, develops within the foveal avascular zone and appears to be represent an inner retinal cavitation on optical coherence tomography. Initially telangiectasia is not visible on biomicroscopy and fluorescein angiography shows mild staining within one disc diameter of the foveola, especially temporally. With progression of the disease there is formation of dilated right angled venules and arterioles, stellate plaques of pigment epithelial hyperplasia, intra and subretinal anastomosis and subretinal neovascularization2.

Neurosensory atrophic changes lead onto lamellar or full thickness macular hole in some patients. Fundus fluorescein angiography in early disease shows bilateral perifoveal telangiectasia with diffuse leakage in late phase but without cystoid macular edema. Neovascularization and cystoid macular edema may be demonstrated in later stage of the disease4. Optical Coherence Tomography shows hyporeflective intraretinal cavitation without foveolar thickening and middle or inner layer hyperreflectivity corresponding to areas of retinal pigment epithelial migration. Pathogenesis of perifoveal telangiectasia is unclear, but may involve dysfunction of parafoveolar muller cells that results in capillary endothelial degeneration, which may lead on retinal capillary proliferation and telangiectasia. Crystalline deposits are thought to represent the foot plates of degenerated muller cells. Cystic cavities on optical coherence tomography represent tissue loss from neurosensory degeneration5. As primary pathogenesis is due to neurosensory degeneration, there is no generally acceptable therapy in non proliferative stage, laser photocoagulation or photodynamic therapy does not appear to improve or stabilize visual acuity in non neovascular macular telangiectasia type2. Treatment with anti-angiogenic agents or intravitreal triamcinolone acetonide decrease leakage on fluorescein angiography, but ineffective visually. Studies have shown that despite angiographic and tomographic effects after intravitreal anti angiogenic agents, there is no functional benefit and these agents are not recommended for the nonproliferative stage6. In proliferative stage anti-angiogenic agents may retard subretinal neovascularization, replacing transpupillary therapy and photodynamic therapy. So patients with Idiopathic macular telangiectasia type 2 need to be followed up regularly, if they developsubretinal neovascularization, treated with intravitreal anti-angiogenic agents (Ranibizumab, Bevacizumab).

CONCLUSION: Idiopathic perifoveal telangiectasia is a rare disease characterized by a nonproliferative stage without any definitive treatment and a proliferative stage with treatment. This case is presented for its rare incidence and to emphasize the importance of regular followup for early diagnosis and treatment of subretinal neovascularization associated with the disease to prevent loss of vision.

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