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Case Report

Clinical Case Report – Bilateral Idiopathic Juxtafoveal Telangiectasia

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Abstract

Idiopathic juxtafoveal telangiectasia (IJFT) is a rare, acquired or congenital, unilateral or bilateral, condition of the eyes, usually occurring in the fifth or sixth decade of life, caused by dilated or twisting blood vessels (telangiectasia) and malfunctioning capillaries near the juxtafoveolar region in the retina. We present a patient who consults with complaints of decreased visual acuity in both eyes due to his new glasses that were prescribed to him 2 months ago. Intravenous fluorescein angiography (IVFA) and optical coherence tomography (OCT) were used to confirm the diagnosis of this condition. This case displays a typical presentation of bilateral idiopathic juxtafoveal telangiectasia in an 80-year-old man and highlights the importance of technologies enabling more precise diagnosis and better patient care.

Keywords: *Bilateral; Idiopathic; Juxtafoveal Telangiectasia*

Introduction

Idiopathic juxtafoveal telangiectasia (IJFT) is a rare, acquired or congenital, unilateral or bilateral, condition of the eyes, usually occurring in the fifth or sixth decade of life, caused by dilated or twisting blood vessels (telangiectasia) and malfunctioning capillaries near the juxtafoveolar region in the retina [1]. This conditions can be divided into 3 subgroups and their presentations ranges from subtle macular changes, such as loss of foveal reflex and grayish appearance of the macula, to more prominent findings including localized leakage and intraretinal retinal pigment epithelium (RPE) hyperplasia with associated retinal thickening [2]. In all subgroubs, the ectatic dilations of the retinal capillaries may be associated with a progressive decrease in visual acuity, metamorphopsia, scotomas and/or reading difficulties caused by a compromised central vision.

Initially, patients commonly have a visual acuity of 20/40, which may deteriorate gradually because of subsequent retinal atrophy, intraretinal edema or the development of secondary neovascular membranes [3]. Consequently, irreversible vision loss will occur if left untreated [2]. Fluorescein angiography (FA) and optical coherence tomography (OCT) are frequently used to confirm the diagnosis of this rare condition.

This case displays a typical presentation of Juxtafoveal telangiectasia in an 80-year-old man and highlights the importance of new technologies enabling more precise diagnosis and better patient care.

Case Presentation

An 80-year-old male consulted an optometric clinic with complaints of decreased visual acuity in both eyes, due to his new glasses that were prescribed to him 2 months ago. His last visual exam was done 2 months prior to his consultation. Further ocular history is unknown. His past medical history is listed in table 1 and his medications are listed in table 2. He is allergic to Aspirin, Lisinopril and Niacin. His Hemoglobin A1C was 6.5 and his Blood pressure was at 152/70. Presented best corrected visual acuity (BCVA) were OD: 20/200 and OS: 20/80-2, non-improvable with pinhole OU. Pupils, Motility and Confrontation fields were unremarkable OU. Intraocular pressures by Tonometry Applanation were OD: 17 mmHg and OS: 12 mmHg. Slit Lamp examination of the lens showed the presence of +2 nuclear sclerotic cataract (NSC) OU, trace posterior subcapsular cataract (PSC) on axis in the OD and +1 PSC on axis in the OS. Dilated fundus

examination demonstrated that the optic nerve had a 0.30 ratio OU without any neovascularisation of the disk (NVD) OU. In figure 1 and 2, the macular region of both eyes, suggest the presence of juxtafoveal telangectasia as well as circum-foveal glistening deposits, OD > OS. These findings are typical of IJFT but can also be found in Retinal venous occlusions, Diabetic retinopathy, Radiation retinopathy, Sickle cell maculopathy, Irvine–Gass syndrome, Ocular ischemic syndrome/carotid artery obstruction, Hypertensive retinopathy, Polycythemia vera retinopathy, Localized retinal capillary hemangioma and Crystalline retinopathy. To rule out all other ocular conditions besides Juxta-foveal telangiectasia, an Allen-watsky test was performed and the results were negative OU. In addition, Macular SD-OCT images that were taken OU showed Macular microcystic changes OD > OS with surrounding increased retinal thickness OU (figure 3 and figure 4). These findings are also typically found in IJFT. The periphery was flat and within the normal limits OU.

	Degeneration of lumbar or lumbosacral intervertebral disc
	History of Peptic ulcer disease
	Lactose intolerance
	Peripheral vascular disease leg/foot
	Essential hypertension
	Congestive obstructive pulmonary disease (COPD)
	Subclavian steal syndrome
	Unspecified nensorineural hearing loss
	Coronary artery disease (CAD)
	Essential and other specified forms of tremor
	Hyperglycemia
	Aneurysm (aorta/abdominal)
	Left inguinal hernia
	Methicilin resistant staphylococcus aureus
	Idiopathic sleep related non-obstructive alveolar hypoventilation
	Dyslipidemia
	Vitamin D deficiency
	Personal history of noncompliance with medical treatment
	Abnormal glucose
	Chronic kidney disease
	Falls
	Abnormality of gait
	Muscle weakness
	Macrocytosis

Table 1*: Patient's past medical history.*

	Amlopidine besylate 10mg TAB; one tablet by mouth every day
	Gabapentin 300mg CAP; one capsule by mouth BID
	Isosobide mononitrate 30mg SA; one tablet by mouth every day for chest pain or pressure (replaces nitro-patches)
	Metoprolol tartrate 50mg TAB; one tablet by mouth every 12 hours,
	Nitroglycerin 0.3mg SL TAB; dissolve one tablet under the tongue every 5 minutes as needed up to 3 doses for chest pain
	Omeprazole 20mg EC CAP; one capsule by mouth BID

	Primidone 50mg TAB; 4 tablets by mouth BID
	Rosuvastatin CA 40mg TAB; one tablet by mouth every day for cholesterol
	Acetaminophen 500mg TAB; 500mg by mouth
	Calcium citrate 315mg/VIT D 200 UNT TAB; 2 tablets by mouth BID
	Coenzyme Q10 [OTC] CAP/TAB; 1 CAP/TAB by mouth every day
	Fish oil 1000MG ORAL CAP; 1000mg by mouth every day
	Multivitamin therapeutic CAP/TAB; 1 one capsule by mouth
	Nattokinase CAP/TAB; 100mg every day

Table 2: *Patient's medications.*

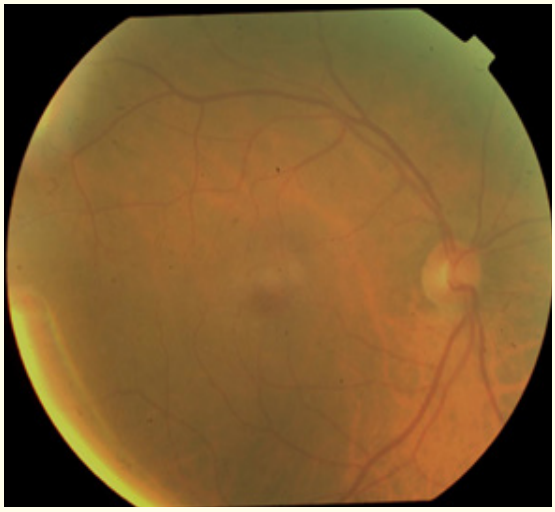

Figure 1: *OD showed juxtafoveal telangiectasia and circum-foveal glistening deposits.*

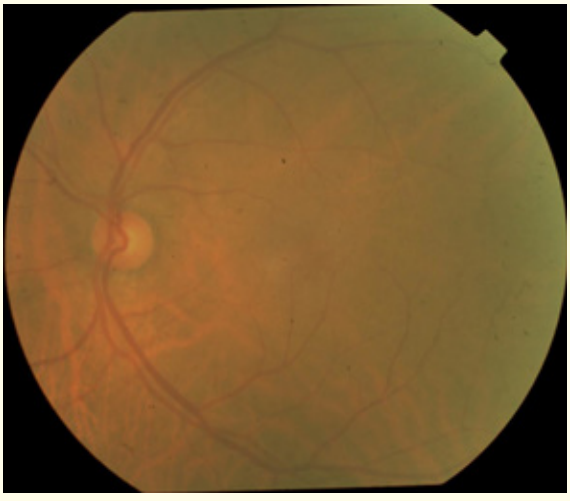

Figure 2: *OS showed less juxtafoveal telangectasia and circum-foveal glistening deposits.*

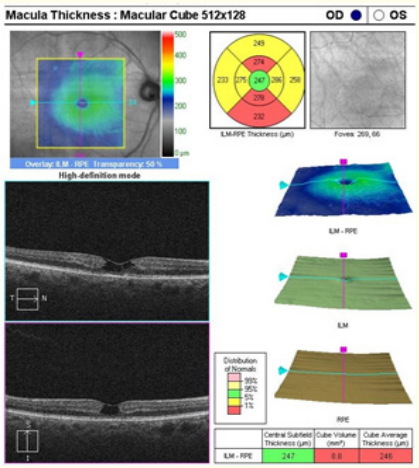

Figure 3: *Macular SD-OCT OD showed underlying macular microcystic changes OD > OS and surrounding increased retinal thickness.*

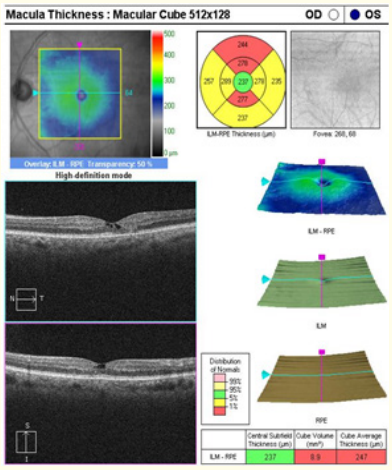

Figure 4: *Macular SD-OCT OS showed underlying microcystic macular changes OS< OD and surrounding increased retinal thickness.*

Impression/Plan

Decreased BCVA OD>OS was most likely secondary to juxtafoveal telangiectasia OU; OD>OS. The patient was referred the same day to Retina clinic for an evaluation and for an Intravenous fluorescein angiography (IVFA). In addition, due to the patient's visually-significant cataracts OU, he was advised to consider cataract extraction after his Retina referral. Following the IVFA, ophthalmologists determined that the fluorescein angiography (FA) OD demonstrated punctuate hyperfluorescence surrounding the fovea with late leakage and an enlarged foveal avascular zone (FAZ) (figure 5). The OS demonstrated similar findings (figure 6). With the results from the OCT and the IVFA, the diagnosis of bilateral IJFT OU, OD > OS was confirmed.

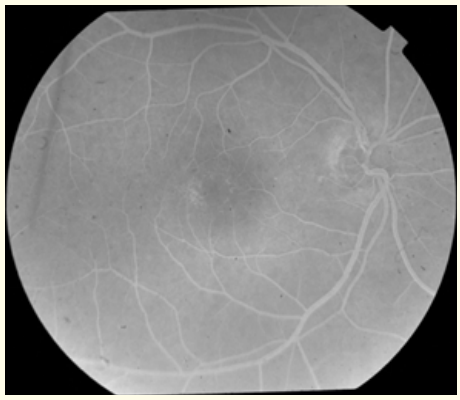

Figure 5: *FA OD showed punctuate hyperfluorescence surrounding the fovea with late leakage and an enlarged FAZ compare to the OD.*

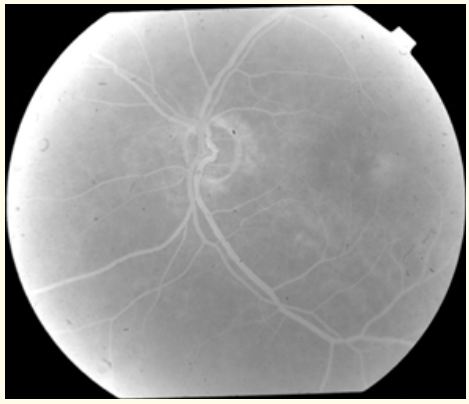

Figure 6: *FA OS showed less punctuate hyperfluorescence surrounding the fovea with late leakage and an enlarged FAZ.*

Clinical Course

The diagnosis was discussed with the patient and the patient was explained that the major cause of decreased vision was most likely the bilateral IJFT. The patient's cataracts played a smaller role in his decreased vision. The patient is under observation for the time being and will be treated with Anti-vascular endothelial growth factor (VEGF) (ranibizumab or bevacizumab) OU if he develops worsening macular edema or if the condition becomes proliferative. Follow up will be in 3 months in Retina clinic. Prior to cataract extraction, the patient was referred to low vision because he wishes to see what improvement he can achieve with low vision aids.

Discussion

FA helps delineate complex abnormal vascular capillaries and demonstrates leakage from telangiectatic vessels [2]. On the other hand, the OCT often shows the presence of paracentral foveal cysts which are described as hyporeflective pockets within the retina related to local hypoxia or photoreceptor degeneration. This can also contribute to the decrease in visual acuity [4]. Results from the patients FA and OCT coincided with the theoretical clinical findings and were key tools in the diagnosis of this rare ocular condition. For the time being, no neovascular membranes were observed. However, due to the unpredictable natural course of the disease a 3-month follow-up would be required and treatment with Anti-vascular endothelial growth factor (VEGF) (ranibizumab or bevacizumab) should be considered if the macular edema worsens or if the condition becomes proliferative. A study has shown that, in one reported patient with Juxtafoveal telangiectasia, a single intravitreal injection of anti-VEGF agents (Bevacizumab) resulted in an increase in VA from 20/50 to 20/20, with significant and sustained decrease in both leakage on FA and cystoid macular edema on the OCT for up to 12 months [4]. Patients benefit

functionally and morphologically from anti-VEGF therapy (ranibizumab or bevacizumab) if there is sign of proliferation. Otherwise, when the condition is non-proliferative and primarily unilateral, with visible telangiectatic retinal capillaries, macular edema and lipid deposition/exudate. laser photocoagulation may reduce exudation and stabilize vision [5].

This case demonstrates the importance of being equipped with the latest technology in order to have fast and efficient results enabling primary eye care professionals to have enhanced patient care. In this clinical case, for example, with a normal funduscopic examination, underlying microcystic macular changes with surrounding increased retinal thickness would have easily been missed. Yet these changes were clearly portrayed with the macular SD-OCT and a rapid diagnosis was established. A recent study used Polarization-sensitive spectral-domain optical coherence tomography (PS-SD-OCT) and SD-OCT to described characteristic morphologic changes in idiopathic juxtafoveal telangiectasia [3]. During the 1st decade of the millennium, the OCT is considered to be one of the top ten must have innovations in eye care [6]. Thus, in today's practice, equipping yourself with the latest ophthalmic technology benefits all parties.

Conclusion

This case demonstrated the clinical presentations found in a bilateral case of idiopathic juxtafoveal telangiectasia by the use of the OCT and IVFA. Furthermore, this case proves how diagnostic tools will help primary eye care professionals see ocular structures *in vivo* that are otherwise undetectable. Consequently, diseases will be diagnosed earlier and treatment will be administrated faster.

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