Natural History of Juxtafoveal Retinal Telangiectasia

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Idiopathic juxtafoveal retinal telangiectasia (IJRT) has been considered a separate clinical entity since it was first described by Gass in 1968. In 1993, Gass and Blodi examined 140 such cases seen at Bascom Palmer Eye Institute, Miami over a 28-year period and established a classification of these entities with subgroups and stages. In recent years, newly recognized manifestations have expanded and refined the clinical spectrum of these macular vasculopathies. Furthermore, the use of high-speed angiography and optical coherence tomography (OCT) have provided a better understanding of the nature of the vascular abnormalities and their secondary effects in the macula, to some degree paralleling histopathological observations described in the ophthalmic literature.

There are few reports on the long term natural history of IJRT. We describe a retrospective series of 35 eyes of 20 patients with IJRT with special emphasis on visual acuity.

**Methods**

We analyzed the case records of 35 eyes of 20 patients diagnosed with IJRT between January 1, 1998 and December 31, 2004. The standard for recruitment was unilateral or bilateral presence of abnormal juxtafoveal vessels documented by fundus fluorescein angiography (FFA). At the follow-up visits, patients were questioned about visual symptoms. Each patient underwent complete ophthalmologic examination, including biomicroscopy and indirect ophthalmoscopy. Colour fundus photographs were taken during follow up visits. Vision was assessed by using the standard eight steps Snellen test charts; measurements were made on the first and follow-up visits and recorded in the clinical records. Comparative visual change between visits was reported as lines of loss or gain. We assessed the clinical photographic and fundus fluorescein angiography (FFA) characteristics of eyes with type II A disease at first visit and during follow-up using the stages of development of parafoveal telangiectasis as described by Gass and Blodi: stage 1 is characterized by no biomicroscopic abnormality, but staining and leakage at the level of retinal pigment epithelium (RPE); Stage 2, by slight retinal graying and capillary telangiectasis visible only by FFA; stage 3 by parafoveal dilated and right angled venules; stage 4 by RPE hyperplasia within the retina; stage 5 by subretinal neovascularisation.

**Results**

There were 8(40 %) male and 12 female (60 %) patients. Age in years ranged from 41 to 72 (Mean 49.65). Ten patients (50 %) were diabetic and 8(40 %) were hypertensive. Of the 20 study patients, 5 had Gass type IA disease and 15 had Gass type IIB disease. Follow-up period varied from 4 months to 102 months (mean 35.6 months). Best corrected visual acuity (BCVA) remained same in 16 eyes(45.71 %), deteriorated by 1 or more lines in 11 eyes (31.42 %), deteriorated by 3 or more lines in 3 eyes (8.57 %), improved by 2 lines in 4 eyes (11.42 %), improved by
1 line in 1 eye (2.85%). The cause for deterioration included progression of IJRT in 9 (64.28%) eyes, progression of cataract in 4 (28.57%) eyes, branch retinal vein occlusion (BRVO) in 1 (7.14%) eye. Two eyes showed improvement in BCVA as a result of cataract surgery, 1 eye because of resolution of macular edema, 2 eyes as a result of regression of corneal disease. None of the eyes had diabetic retinopathy in the beginning, but 6 eyes (15%) developed changes of mild NPDR and 4 eyes (10%) moderate NPDR during follow-up. None of these eyes had features of diabetic macular edema/maculopathy.

Type I B: Unilateral idiopathic, focal juxtafoveal telangiectasis

There were 5 patients (3 males and 2 females) who had type I B disease. Mean age was 51.4 years. All these patients had unilateral focal area of capillary telangiectasis in the parafoveal area as evidenced by fluorescein angiography. Of these 5 eyes, best corrected visual acuity (BCVA) dropped by 1 line in 1 eye (20%) due to progression of IJRT, remained stable in 3 eyes (60%) one of which had poor vision due to BRVO, which was confirmed by FFA (Patient 2; Table 1). BCVA improved by 2 lines in 1 eye (20%) as a result of resolution of macular edema in eye (20%) over a period of 72 months follow-up (Patient 1; Table 1).

Type IIA: Bilateral idiopathic acquired juxtafoveal telangiectasis

Thirty eyes of 15 patients were classified as having type IIA disease. There were 5 male (33.33%) and 10 female patients (66.66%) BCVA remained stable in 13 eyes (43.33%). BCVA dropped by 1 line in 7 eyes (23.33%) (all due to progression of IJRT). deteriorated by 2 lines in 3 eyes (10%) (all due to progression of cataract), deteriorated by 5 lines in 2 eyes (6.66%) (one due to development of SRNVM and other due to BRVO). One eye (3.33%) showed deterioration by 10 lines due to progression of cataract to maturity. Three eyes (10%) showed improvement in BCVA by 2 lines (2 eyes as a result of cataract surgery and 1 eye because of regression of corneal oedema). BCVA improved by 1 line in 1 eye (3.33%) due to regression of corneal problem.

Case 10 (Patient 10, Table 1) exemplifies progression of disease to subretinal neovascularisation. This patient was a 43 years old woman who noted gradual loss of central vision first in the right eye and then in the left eye for 6 months. She was neither diabetic nor hypertensive. She was examined in December 2000 and BCVA was 6/36 in right eye and 6/12 in left eye. Parafoveal whitening, RPE proliferation and pigmentation temporal to fovea was seen in right eye. Fluorescein angiography of right eye showed leakage of dye. (Stage 4 disease) Left eye showed retinal graying and dilated and blunted retinal venules. FFA confirmed this and showed leakage. (Stage 3 disease) In September 2001, her vision was stable at 6/36 in right eye and 6/12 in the left eye. There was increased pigmentation and proliferation in the right eye. FFA showed about same amount of intraretinal leakage in both eyes. She was not seen again until March 2007. Her BCVA had dropped to 2/60 in right eye and was stable at 6/12 in left eye. Fundus photograph of right eye (Fig. 1a) showed a scarred CNVM (Stage 5 disease) which was confirmed by FFA (Fig 1c). Left eye fundus photograph (Fig 1b) showed no significant change and FFA...
(Fig. 1d) showed increased intra-retinal leakage (stage 3 disease). OCT examination (horizontal line scan) at this visit showed a scarred membrane in right eye (Fig 1e) and pseudocyst in left eye (Fig 1f).

**Case 15** (Patient15, Table 1) is an example highlighting the slow rate of progression of the disease. This 52 years old man complained of defective vision in both the eyes of 6 years duration. He was a known diabetic. On examination in December 2003, he was missing letters of the Snellen's chart in both eyes and his BCVA was 6/12 in right eye and 6/18 in left eye. Anterior segment examination revealed immature senile cataract in both eyes. Colour fundus photograph of both eyes (Fig. 2a, 2b) showed parafoveal retinal graying and crystals and left eye in addition showed RPE hyperplasia and pigmentation. FFA of both eyes (fig. 2c, 2d) showed telangiectatic capillaries and late intra-retinal leakage of dye in both eyes. He was followed up and in July 2005, his BCVA was stable at 6/12 in right eye and 6/18 in left eye. Anterior segment examination was almost unchanged. Colour fundus photo of right eye showed parafoveal retinal graying and crystals. Left eye colour fundus photograph was significant for the increase in the pigmentation. In June 2007 he presented with drop in visual acuity, BCVA being 6/24 in right eye and 6/36 in left eye. Anterior segment examination revealed significant progression of cataract in both eyes. On colour fundus photography (Fig 2e, 2f), both eyes were status quo. FFA was significant only for the increase in the intra-retinal leakage of dye (Fig 2g, 2h). There was no evidence of subretinal neovascularisation in either eye. The deterioration of visual acuity was mainly due to progression of cataract.

**Case 20** (Patient 20, Table 1) is another example which highlights the very slow progressive nature of the disease. This patient was 44 years old woman who had
noted gradual loss of vision first in right eye and then in left eye since 6 months. She was examined in September 2004. BCVA was 6/18 in right eye and 6/6 in left eye. There was parafoveal graying along with crystals in both the eyes. (Fig 3a and Fig 3b). On FFA, there was late leakage of dye in right eye (Fig. 3c) and minimal leakage in left eye (Fig. 3d). She was followed up and during her last visit in March 2007; BCVA was stable in both eyes, 6/18 in right eye and 6/6 in left eye. (Fig. 3e & f) status quo fundus photograph in both eyes on review

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December 2008  

**Discussion**

There were 5(25%) patients with type 1B disease and 15(75%) patients with type 2A disease confirming that type 2a is the most common form of IJRT. Majority (16 eyes, 45.71%) maintained a stable visual acuity and vision loss in patients with IJRT is generally mild and occurs over many years.

When we analyzed each disease type, there were 5 patients with type IB disease, of which majority (60%) were male and mean age was 51.4 years. This pattern in which middle-aged men are most commonly affected correlates well with the demographic characteristics described earlier. Regarding type 2A disease, there were 15 patients and majority was female. (66.66%). This slight female preponderance was in contrast to an earlier report of equal sex distribution.

Among the eyes in our study that lost vision, progression of IJRT was responsible only in 64.28% of cases. Loss of central vision occurs slowly over many years and is associated with atrophy of the foveolar retina. Visual disturbance in retinal telangiectasia is usually due to vascular leakage, with intra-retinal edema and exudate accumulation and later cystic degeneration. However, the development of neovascular membrane in the vicinity of a black hyperplastic retinal pigment epithelial plaque or a dilated vein passing at right angles into the depth of the retina and indicative of a retinochoroidal anastomosis can lead to rapid and severe visual loss. None of the patients in our series developed macular hole. The two recent reports of full thickness macular hole development in IJRT have opened up a new dimension in the pathogenesis and natural history of IJRT. The pronounced central foveal structural abnormalities (for excavitation) could be due to loss of the structural aspects afforded by Muller cells, particularly the Muller cell cone. Hence loss of Muller cells could be an important factor in the pathogenesis of IJRT.

No intervention was done in any of the patients in our series. Park and associates attempted grid laser photocoagulation for macular edema in IJRT but found that it neither improved nor stabilized long-term visual acuity. The role of any intervention arises only in type 2 stage 5 diseases when there is development of subretinal neovascularisation. Intravitreal injection of triamcinolone acetonide has been found to be of some benefit in few reports, but there are no randomized controlled trials. The potential role of photodynamic therapy with Verteporfin in IJRT with subretinal neovascularisation has been well substantiated in literature. IJRT is a slowly progressive disease and visual acuity remains stable for quite a long time.
Conclusion

IJRT has favourable prognosis unless there is development of subretinal neovascular membrane 12.

References

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