Neovascular Membranes Associated With Idiopathic Juxtafoveolar Telangiectasis

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Objective: To report the visual outcome in patients with a neovascular membrane (NVM) associated with idiopathic juxtafoveolar telangiectasis (IJFT).

Methods: We performed a retrospective, noncomparative analysis of 26 eyes of 16 patients with an NVM associated with bilateral IJFT (Gass classification group 2A). Eyes were divided into 2 groups: group WO (n=11) included eyes with IJFT without evidence of an NVM on initial examination; eyes in group W (n=15) had an NVM at the initial diagnosis of IJFT. In group WO, the initial visual acuity and the time between the initial examination to the diagnosis of an NVM were evaluated. Characteristic fundus findings, including the presence or absence of a chorioretinal anastomosis, intraretinal pigmentary plaques, and crystalline deposits, as well as the final visual acuity were reviewed for both groups.

Results: The initial visual acuity for eyes in group WO ranged from 20/20 to 20/70 (median, 20/30); in group W, from 20/20 to 4/200 (median, 20/70). The average time from initial diagnosis of IJFT to the development of an NVM was 73 months (range, 5-142 months). In group WO, chorioretinal anastomosis and concurrent perivascular retinal pigment epithelial hyperplasia were observed before the development of an NVM. The final visual acuity for all eyes ranged from 20/40 to 2/200 (median, 20/200). Eighty-one percent of eyes (21/26) had a final visual acuity of 20/200 or worse.

Conclusions: The stable final visual acuity in patients with an NVM associated with IJFT is generally poor, with 80% of eyes in this series having a final visual acuity of 120/200 or worse. In patients with IJFT, the presence of a chorioretinal anastomosis and retinal pigment epithelial hyperplastic plaques always preceded the development of an NVM.

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DIOPATHIC juxtafoveolar telangiectasis (IJFT) is characterized by the presence of an area of ectatic and incompetent retinal capillaries in the foveolar region, in the absence of other known causes for retinal telangiectasis. Idiopathic juxtafoveolar telangiectasis may be congenital or acquired, and has been classified by Gass and Blodi into 3 groups. Under Gass’s classification, group 2A describes patients with acquired, typically bilateral areas of occult capillary telangiectasis who generally seek care because of complaints of mild blurred vision in the fifth or sixth decade of life. Vision loss generally occurs slowly over many years, but may be rapid with the development of a neovascular membrane (NVM).

The natural course of eyes with NVMs associated with IJFT is not well known. Therefore, the potential benefits and indications for treatment of the NVM are poorly defined. One previous report by Park et al found a mean final visual acuity of 20/70, in which they questioned the role of laser photocoagulation of the fibrovascular tissues given the relatively mild level of vision loss. The purpose of our study was to review the natural course of NVMs in eyes with IJFT and the associated visual outcomes in these patients. Serial fundus photographs were evaluated for characteristics that may predict the development of an NVM. The potential role of treatment of these lesions, including its indications, timing, and methods, is also discussed.
PATIENTS AND METHODS

We reviewed the charts of all patients with the diagnosis of IJFT with an associated NVM who were seen at the Bascom Palmer Eye Institute, Miami, Fla, between May 1, 1966, and January 31, 1996. The eyes included in this series have been previously reported by Gass and Blodi; however, in that report, the outcomes were not evaluated. The patient medical records, including the clinical chart, as well as serial fundus photographs and fluorescein angiograms were reviewed. One patient was excluded because the NVM was treated with focal laser photocoagulation. We reviewed 26 eyes of 16 patients with an NVM in association with IJFT who received no treatment. The eyes were divided into 2 groups on the basis of the presence or absence of an NVM at the initial diagnosis of IJFT. Group WO included eyes that developed an NVM after the initial diagnosis of IJFT; eyes in group W had an NVM at initial examination.

The patients' age, sex, and medical history were reviewed for both groups. The initial visual acuity and time between initial diagnosis of IJFT and the development of an NVM were examined for the eyes in group WO. Serial fundus photographs and fluorescein angiograms were reviewed for eyes in group W to attempt to establish a pattern of progression leading to the development of an NVM. The final stable visual acuity and length of follow-up were reviewed for both groups.

The patients' medical history was significant for hypertension in 1 patient, diabetes mellitus in 7, thyroid disease in 1, chronic obstructive pulmonary disease in 1, and coronary artery disease in 1. One additional patient had a history of a positive glucose tolerance test but was not treated for diabetes mellitus. Of the 7 patients with diabetes mellitus, none showed retinal changes consistent with diabetic retinopathy; specifically, microaneurysms, blot hemorrhages, and neovascularization of the disc or elsewhere were absent.

Group WO included 11 eyes that developed an NVM after the initial diagnosis of IJFT. The initial visual acuity for eyes in group WO ranged from 20/20 to 20/70 (median, 20/30). The time between the initial diagnosis of IJFT and the development of the NVM ranged from 5 to 142 months (mean, 73 months). A review of serial fundus photographs for eyes in group WO showed the development or presence of a chorioretinal anastomosis in all eyes before the development of the NVM. Hyperpigmented plaques were also noted in all eyes before the diagnosis of an NVM. Two eyes in group WO with stage 3 IJFT were treated with focal laser photocoagulation for persistent macular edema associated with the IJFT. Both eyes developed an NVM, one at 1 month and the other at 3 months after the laser treatment.

Group W included 15 eyes with an NVM at initial examination, with a median visual acuity of 20/70 (range, 20/20 to 4/200). A chorioretinal anastomosis was seen in all eyes in group W, and all but 1 eye showed hyperpigmented plaques at the site of the NVM.

The NVMs found in these patients were most commonly located temporal to the fovea in association with hyperplastic plaques and appeared to originate from the retinal vasculature as opposed to the choroidal vasculature (Figure). Initially, the NVMs enlarged with concurrent retinal edema and subretinal hemorrhage before becoming a cicatrix. Once in the cicatricial stage, the membranes tended to contract.

Groups WO and W each had a median final visual acuity of 20/200 (range, 20/200 to 2/200 and 20/40 to 5/200 in groups WO and W, respectively). The stable final visual acuity for all eyes ranged from 20/40 to 2/200 (median, 20/200). There were no identifiable characteristics of the NVM that predicted a better visual outcome. Twenty-one (81%) of the eyes in this series had a final visual acuity of 20/200 or worse. The average length of follow-up was 107 months (range, 14-212 months).

COMMENT

Vision loss in patients with IJFT is generally mild and occurs over many years. The development of an NVM may occur late in the disease process and lead to more dramatic vision loss. The natural course of an NVM associated with IJFT is not well understood, and therefore the utility of treatment is not well defined.

Idiopathic juxtafoveolar telangiectasis involves the presence of ectatic retinal capillaries in the absence of other known causes of retinal telangiectasis, such as diabetic retinopathy, radiation retinopathy, carotid occlusive disease, or following a branch retinal vein occlusion. In 1982, Gass and Oyakawa originally proposed a classification system of idiopathic juxtafoveolar retinal telangiectasis. This classification system was revised in 1992 by Gass and Blodi and includes 3 classification groups. Under the current classification, group 2A includes patients with bilateral IJFT. Group 2A represents the most common type of IJFT, and patients are usually first examined in the fifth and sixth decades of life because of the complaint of mild vision loss in one or both eyes. Idiopathic juxtafoveolar telangiectasis is an acquired disease that usually occurs bilaterally and has no sex predilection. The presence of exudation from the telangiectatic vessels is also characteristically absent.

Gass and Blodi described 5 stages in the progression of group 2A IJFT. In stage 1, there is evidence of juxtafoveolar telangiectasis on fluorescein angiography, but with no abnormality seen on clinical examination. Biomicroscopic evidence of the disease begins in stage 2 with graying of the parafoveal retina; also, minimal telangiectatic changes may be visible. Patients most commonly are first seen with stage 3 disease and a complaint of mild vision loss that appears to be secondary to progressive foveal atrophy. Stage 3 is clinically characterized by the presence of right-angle–draining retinal venules or chorioretinal anastomosis. Stage 4 is characterized by the development of plaques of hyperplastic retinal pigment epithelium that are commonly associated with the chorioretinal anastomosis. The final stage, stage 5, involves the development of an NVM, which is believed to originally develop from the retinal
Patient Characteristics

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VA indicates visual acuity; WO, eyes without a neovascular membrane at initial examination; and W, eyes with a neovascular membrane at initial examination.

Idiopathic juxtafoveolar telangiectasis with associated neovascular membrane (NVM). A, Color fundus photograph of the NVM. B through D, Corresponding fluorescein angiograms showing progressive hyperfluorescence of the NVM. B, Note the retinal vessel (arrow) filling before the appearance of the NVM.
vasculature but may also include connections to the choroidal vasculature or choroidal neovascularization.1,2 Superficial retinal crystalline deposits of unknown origin have also been described in eyes with IJFT and have been reported to occur in 40% to 45% of eyes.3,5,6 The significance of these refractile deposits is unknown and they may be seen in stages 2 through 5 in the progression of the disease.

In our series, all patients had the characteristic findings on initial examination that were described by Gass and Blodi7 for patients with group 2A IJFT. All eyes included in our series also had an NVM present (stage 3). Patients in group WO of this series did not have an NVM present at the time the patient was initially diagnosed as having IJFT. All eyes in group WO that had adequate serial fundus photographs available showed the progression of disease as described by Gass and Blodi. The development of a chorioretinal anastomosis in the area of the juxtafoveal telangiectasis was followed by the development of hyperpigmented retinal pigment epithelial plaques before the presence of an associated NVM in all eyes in group WO. Also, all eyes with an NVM at the initial diagnosis of IJFT (group W) had a chorioretinal anastomosis with hyperplastic plaques present, except 1 eye that did not show a hyperplastic plaque. No eyes in our series with stage 5 IJFT had crystalline deposits present, although 1 patient had crystalline deposits present in the opposite eye without an NVM.

An association between systemic diseases, such as diabetes mellitus, and IJFT has not been well established. The presence of parafoveal telangiectasis and diabetic retinopathy in patients has been previously reported,7 but the eyes in this series had other manifestations of nonproliferative diabetic retinopathy. Although 7 of the 16 patients in this series had a diagnosis of diabetes mellitus and 1 additional patient had a history of a positive glucose tolerance test, none of these patients had any typical manifestations of diabetic retinopathy. Also, the development of subretinal NVM is uncharacteristic of the manifestations of diabetic retinopathy. Although familial cases of bilateral IJFT have been reported,8,9 none of the patients in our series were related.

Park et al12 reported the visual outcome in a series of eyes with IJFT and associated fibrovascular tissue. They found little change in the size of the fibrovascular tissue and an average stable final visual acuity of approximately 20/70 for the 11 eyes in their series. Because of the relatively mild vision loss associated with the presence of neovascular tissue and the apparent stability of the membranes, they questioned the utility of treatment in these eyes. In contrast to the previously reported series, we found a median stable final visual acuity of eyes in this series of 20/200, with 80% of eyes having a final visual acuity of 20/200 or worse. Only 3 of 26 eyes maintained a visual acuity of 20/70 or better. Because 80% of the eyes in our series had poor visual acuity with long-term follow-up, we believe that treatment, if available, should be considered before significant vision loss. In our series, there were no identifiable characteristics of the NVM that predicted a better visual outcome.

Previous studies have reported the use of focal laser photocoagulation to areas of macular edema associated with juxtafoveal telangiectasis.1,2,5,9-11 Park et al10 reported the results of 10 eyes treated with grid laser photocoagulation for macular edema in bilateral juxtafoveal telangiectasis and showed that treatment did not improve or stabilize long-term visual acuity. Gass and Blodi7 reported the results of 8 eyes with stages 3 and 4 disease treated with laser photocoagulation; they reported no improvement in vision in 4 eyes and worse vision in 4 eyes. Laser treatment may be complicated by subretinal hemorrhage or the development of an NVM.10,11 The development of an NVM after laser treatment has been reported.10,11 In our series, 2 eyes that were treated with laser for macular edema developed an NVM, one at 1 month and the other at 3 months after treatment. Because previous reports have shown no significant improvement in visual outcomes with focal laser photocoagulation, and because there may be an increased risk of the development of an NVM following treatment, the use of laser photocoagulation for the treatment of macular edema associated with IJFT is not recommended.

The role of treatment of NVMs associated with IJFT is not well defined. The NVMs in IJFT in our series initially grew with associated retinal edema and subretinal hemorrhage before entering a cicatricial stage in which the membrane contracted or remained relatively unchanged. Therefore, intervention would appear to be potentially most beneficial in the early stages of neovascularization. However, to our knowledge, there are no reports in the peer-reviewed literature that discuss the role of focal laser photocoagulation for the treatment of NVM associated with IJFT. Berger et al12 reported the results of 2 eyes that were treated with surgical removal of a subfoveal NVM. The visual outcome in both eyes was poor secondary to marked adherence between the NVM and the sensory retina. Therefore, as they reported, the surgical removal of these membranes may be contraindicated. Photodynamic therapy may play a role in the treatment of subfoveal NVMs in IJFT. Recently, photodynamic therapy has been reported to result in resolution of leakage from an NVM, with no effect on the edema associated with the telangiectasis.13

The natural course of the visual acuity in eyes with NVMs associated with IJFT has not been well defined. Therefore, the indications for treatment of these membranes are also not well established. The use of laser photocoagulation for macular edema associated with IJFT has not been shown to stabilize or improve visual outcomes in these patients and may hasten the development of NVMs. Therefore, this treatment is not recommended. The use of laser photocoagulation or photodynamic therapy for the treatment of NVMs associated with IJFT may improve the visual outcomes in these patients; however, there are insufficient data at this time to support their use in these patients. Eighty-one percent of the eyes in our series with an NVM associated with IJFT had a stable final visual acuity of 20/200 or worse. Therefore, we believe that it is important to investigate the potential role of treatment of these eyes to attempt to prevent significant vision loss.
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REFERENCES

Ophthalmological Numismatics

Anton Elschnig, 1886-1939, was a professor of ophthalmology in Prague. He is best know for his research on the lens capsule, especially the exfoliation of glass-blowers. The lens remnants seen on the capsule following extracapsular cataract surgery are known to this day as “Elschnig’s pearls.”

For his 40th birthday in 1926, a uniface medallic plaquette (Figure) in bronze was created by the artist Henke.

Courtesy of: Jay M. Galst, MD, 30 E 60th St, New York, NY 10022.