Neovascular glaucoma in neurofibromatosis type 1 (NF1)
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Introduction: Glaucoma in Neurofibromatosis (NF) usually occurs early in life, in patients with unilateral orbito-facial involvement. It results from a combination of factors including infiltration of the angle by diffuse neurofibromas, of the ciliary body causing mechanical angle closure, congenital angle abnormalities and congenital ectropion uvea. Neovascular glaucoma and anterior segment neovascularization in NF1, secondary to retinal vascular lesions is rare [1] and this report highlights the spectrum of retinal vascular findings that may be seen with this association.

Objective: To describe the clinical findings in three patients with neurofibromatosis 1 (NF1) and retinal vascular abnormalities that resulted in neovascularization of the iris, and to describe the histopathological abnormalities in one case. Material and methods: Retrospective case series of patients with NF1 at tertiary care center presenting with iris neovascularization secondary to retinal vascular abnormalities. Histopathological examination of an enucleated specimen from one eye.

Results: Three children whose age ranged from 5-10 years at presentation, had unilateral retinal vascular abnormalities (two vasoproliferative lesions and one diffuse retinal ischemia) and iris neovascularization. Two cases presented with acute elevation of intraocular pressure and neovascular glaucoma. Histopathology of the enucleated eye revealed ectropion uvea, and angle closure secondary to a fibrovascular membrane, diffuse inner retinal ischemic changes and a retinal detachment. Two cases, one with the vasoproliferative lesion and the eye with retinal ischemia showed progressive changes and went blind in the affected eye. One case with a vasoproliferative tumor, who initially received Bevacizumab intracameraly, retained useful vision after controlling the neovascular glaucoma with an Ahmed implant and undergoing cryo and laser ablation of the retinal vascular abnormalities.

Conclusions: A variety of retinal vascular lesions occurring in NF1 are capable of producing neovascularization of the iris, ectropion uvea and neovascular glaucoma. Although complications of such abnormalities can lead to total vision loss in some cases, retention of useful vision is possible with timely treatment of the retinal lesions and associated neovascular glaucoma [1-3]. This report highlights the need for careful examination of the retina with particular attention to vascular abnormalities and peripheral retinal tumors, in young patients with NF1.