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Case Report

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# RANIBIZUMAB EFFECT ON CHOROIDAL NEOVASCULARIZATION IN STARGARDT DISEASE – A CASE REPORT

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## ABSTRACT

This case report demonstrates the short-term effects of multiple Ranibizumab injections in managing choroidal neovascularization in a fifteen-year-old female patient diagnosed with Stargardt disease. The patient had ten days history of decreased vision in the left eye before treatment. After five months of follow-up and three intravitreal Ranibizumab injections, the visual acuity was stabilized, and choroidal neovascularization was resolved.

**KEYWORDS:** Stargardt Disease, Choroidal neovascularization, CNV, Ranibizumab, Anti-Vascular Endothelial Growth Factor Therapy, Anti-VEGF.

## BACKGROUND

Stargardt Disease, also known as Fundus Flavimaculatus, Stargardt's Macular Dystrophy and Juvenile Macular Degeneration, was first described by a German ophthalmologist back in 1909 in patients from similar families with unique characteristics of inherited ophthalmic disease.[1,3] The etiology of Stargardt disease is linked to ATP Binding Cassette Subfamily A Member 4 (ABCA4) gene mutation.[4] Stargardt disease is the most widespread inherited macular dystrophy illness.[2] Several studies estimated the incidence of Stargardt disease to be between 8 to 13 every 100,000 individuals.[1,4]

Due to the high heterogeneity of Stargardt cases, it is hard to generalize one clinical or phenotypic presentation of this disease.[4,5] However, it is well known to affect the central vision of patients. Currently, there is no known cure for this autosomal recessive disease.[6] Early onset Stargardt is generally linked with poor prognosis compared to late-onset Stargardt cases.[5]

One of the rare complications of Stargardt disease is choroidal neovascularization (CNV). CNV could induce disciform lesions if untreated, reducing visual acuity.[7] Previous studies reported different approaches for CNV management in Stargardt disease. Laser photocoagulation, Photodynamic therapy (PDT), and intravitreal Anti-vascular endothelial growth factor therapy (anti-VEGF) were all reported with various outcomes.[1,4,6]

## CASE REPORT

A 15 years old female patient was referred to our department with ten days history of gradually decreased vision in her left eye with no other complaint. The patient was not known to have any previous medical or surgical illnesses. The patient and her caretakers denied any history of trauma, prior hospital admissions, wearing eyeglasses, or any history of previous blurred or decreased vision before the current complaint. The patient's vaccinations were up to age and she did not have a family history of a similar complaint. Her general physical exam was within normal limits. On ophthalmic physical examination of her left eye, numerous fundus yellow-whitish flecks were observed in the posterior pole. The macula had a cystic lesion filled with clear fluid. No other abnormalities were observed in the physical examination. The cornea was clear, the anterior chamber was deep and quiet, her pupil round, regular and reactive and the lens was clear. Also, the intraocular pressure in her left eye was within the normal. The physical examination of the right eye was unremarkable and the intraocular pressure was within normal limits. Her left eye fluorescein angiography, electrophysiology of the retina (ERG) and optical coherence tomography (OCT) showed clear signs of classic juxtafoveal CNV associated with cystoid macular edema (Fig. 1 and 2). Her visual acuity was counting fingers in the left eye.

The CNV management options, possible complications, prognosis and expected outcome of each option were reviewed and discussed with the patient and her taker. After careful consideration, the treatment plan was to

treat the CNV with intravitreal anti-VEGF. The patient was scheduled for intravitreal Ranibizumab (Lucentis®, Genentech, California, USA; Novartis, Basel, Switzerland) injection in her left eye. Ranibizumab injection (0.05 ml/0.5 mg) was administered in her left eye three times at one-month intervals between the injections with no complications. The total follow-up period until writing this case report was five months.

After five months of follow-up, including three monthly Ranibizumab injections, visual acuity was stabilized at 6/12 in the left eye, the fluorescein angiography did not reveal any signs of leakage in the left eye and the OCT did not show any subretinal fluids and these follow-up findings were consistent with CNV closure (Fig. 3).

According to our hospital policy, the patient and her caretaker signed a comprehensive informed consent form.

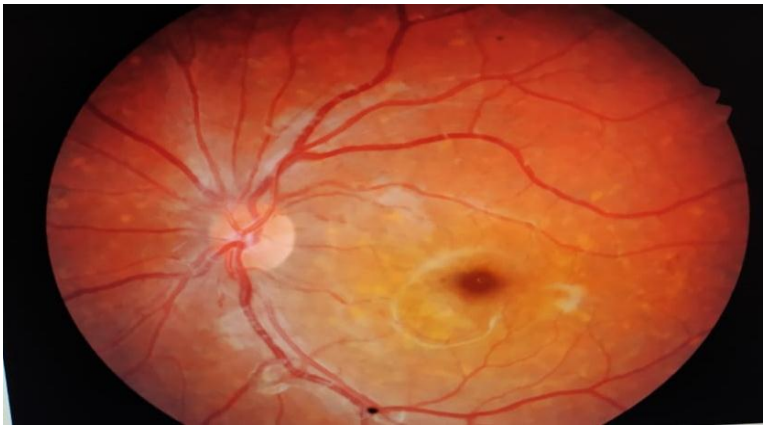

**Figure 1: Fundus photographs of the left eye before treatment.**

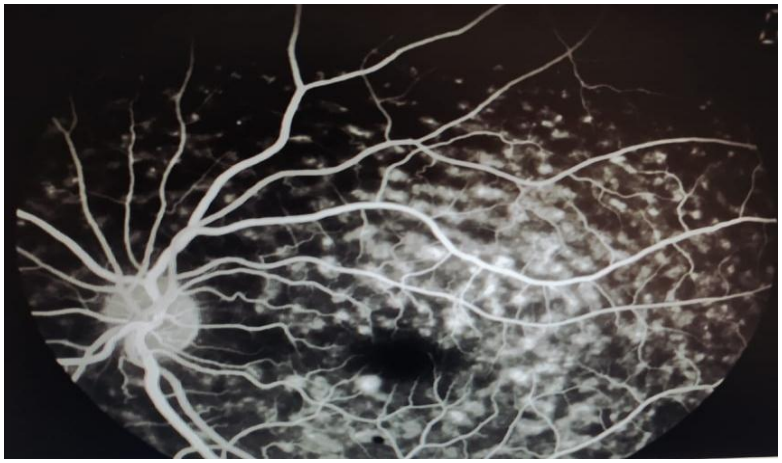

**Figure 2: Fundus fluorescein angiography images of the left eye before treatment.**

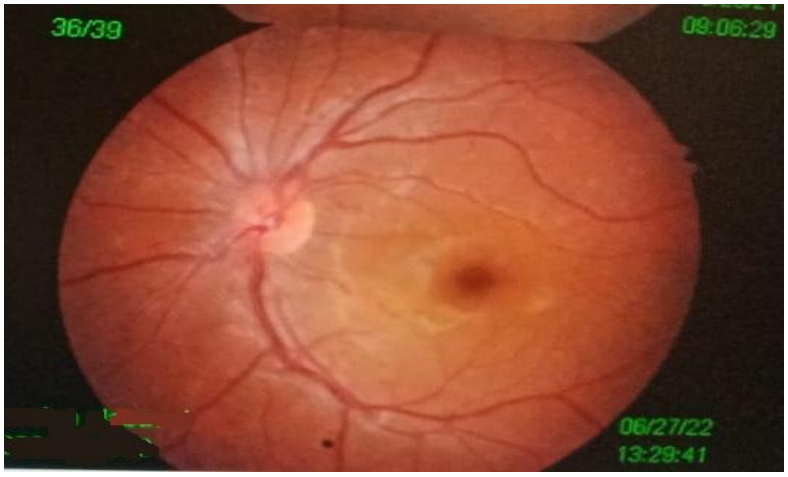


**Figure 3: Fundus photographs of the left eye after treatment.**

## DISCUSSION

This case report shows the effect of anti-VEGF on the management of CNV, a relatively rare complication of Stargardt disease. To the best of our knowledge, only a few previous Stargardt disease case reports focused on CNV management, and anti-VEGF was used only in one of these previous case reports.[6,8] The previous similar case report also revealed a favorable effect of anti-VEGF on the closure of CNV in a Stargardt disease case. However, the follow-up duration of current and previous case reports regarding the impact of anti-VEGF on the closure of CNV was limited to less than six months.

The current case was diagnosed with Stargardt disease based on history, ophthalmic physical examination and investigations. However, genetic analysis was not part of this case assessment and this is one of the limitations of this case report. Nevertheless, the clinical presentation and evaluation findings were typical of Stargardt disease. The typical signs of Stargardt disease are the development of symptoms of atrophic fovea associated with yellow-whitish flecks with numerous shapes. Usually, the onset of Stargardt disease is in the adolescent to early twenties. This inherited disease is generally associated with lipofuscin accumulation in retinal pigment epithelium, which could induce cell death.[9]

Although Stargardt disease is still considered not curable, multiple clinical trials are currently conducted with promising outcomes. The focus of these clinical trials is on genetic treatments, stem cells, and pharmaceutical interventions. Meanwhile, visual aids and restricting the consumption of vitamin A are two common approaches for visual loss management in Stargardt cases.[1,4]

Very limited scientific data is available concerning the efficacy of anti-VEGF in treating CNV in Stargardt disease. This case report had a promising improvement in CNV and visual acuity after multiple Ranibizumab injections. Similar to Querques et al., we assume that it is unlikely to see in the near future any double-blinded clinical trials regarding the effect of Ranibizumab in management of CNV in Stargardt cases due to the rare presentation of this complication and poor natural outcome of this disease.[8]

## CONCLUSION

Taking into consideration the encouraging short-term outcome and the limited number of similar cases, Ranibizumab could effectively manage CNV in Stargardt cases. Due to the high heterogeneity of clinical presentation in Stargardt disease, additional studies are needed to investigate the short and long-term effects of anti-VEGF in the treatment of CNV. In addition, there is a need to understand the pathophysiological consequences of this potential novel treatment for a devastating adolescent ophthalmic disease.

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