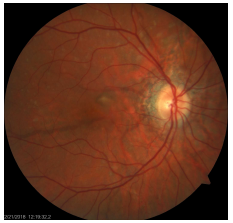
Choroidal neovascularization secondary to Grondblad-Strandberg syndrome: case report

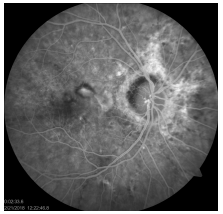
Objective: To report a clinical case of choroidal neovascularization secondary to Gronblad-Strandberg syndrome in a young patient.

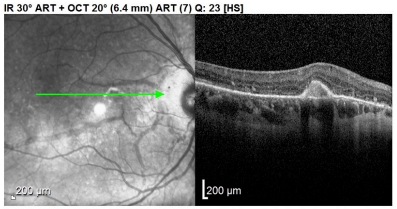
Introduction: Grondblad-Strandberg syndrome is characterized by the development of skin lesions described in the appearance of yellowish confluent papules, mainly in skin folds, known as elastic pseudoxanthoma. It occurs due to the progressive degeneration of elastic tissue related to the most frequent autosomal recessive genetic disease in females.Angioid stretch marks are commonly related to this disease. Dehiscences of the bruch membrane are secondary to abnormal fragility of elastic tissue and due to the proximity of the fovea the macula is more susceptible to complications such as choroidal neovascularization. This complication occurs in 72 to 86% of the patients, with bilateral involvement in 50% of the cases.

Case report:

S.R.F.S, 37 years old, female, presented in March 2019 in a routine ophthalmological evaluation at Institutos Visão, worsening of visual acuity (VA) in both eyes, mainly in the right eye (OD), referring to metamorphopsia during reading. The AMSLER grid exam shows distorted DO temporal vision, even maintaining a 20/20 VA in both eyes. In the anamnesis, he also referred to characteristic flexural skin lesions in the neck of undetermined beginning. Biometrics and tonometry without changes.Fundscopy showed angioid streaks on both optic discs, presence of subretinal hemorrhage with central exudation and choroidal neovascularization in OD. It evolved three months later, with worsening of metamorphopsia and decreased visual acuity, 20/40 -2 in RE. In fluorescent retinography it presents in DO: macula with hyperfluorescence increasing during the exam and choroidal neovascularization. On examination of the macula OCT in OD: she had macular edema with a slum thickness of 406 microns. OE without changes.







Na retinografia simples em OD: *estrias angióides ,* ***retinografia fluorescente em OD*** *: mácula com hiperfluorescência aumentando ao decorrer do exame e neovascularização de coróide .* ***No exame de OCT de mácula em OD:*** *apresentava edema macular com espessura foveal de 406 micra. OE sem alterações .*

Conclusion:

Complications of the disease can generate serious repercussions, such as blindness, in addition to being a hereditary condition that should be investigated in the patient's family. The patient in question presented a brother and cousin with elastic pseudoxanthoma. Treatment referred to as laser photocoagulation or photodynamic therapy with verteporfin is complicated by recurrences and poor visual results. Currently, intravitreal injections of anti-VEGF are promising in the treatment of these patients, and this was the treatment of choice for this case.The patient received 3 intravitreal injections of anti-VEGF, with an interval of 30 days, with significant reduction of macular edema (thickness 236 microns OD), but with return of the liquid after drug suspension. (thickness 429 microns). Due to the healing of the neovascular process and the permeability defect was maintained, the intravitreal application of dexamethasone was chosen in an attempt to improve the clinical condition and prevent recurrent macular edema).