Laser photocoagulation, intravitreal anti-VEGF, and vitreous surgery for the treatment of juxtapapillary retinal capillary hemangioma

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Juxtapapillary retinal capillary hemangiomas (JRCHs) are benign vascular tumors located on or adjacent to the optic nerve head. A 19-year-old girl presented with epiretinal membrane (ERM) associated with an elevated and round vascular tumoral mass located in the juxtapapillary region of her left eye. She was subsequently diagnosed with isolated JRCH. A combined approach with laser photocoagulation and intravitreal bevacizumab injection was used to facilitate shrinkage of the tumor preoperatively and pars plana vitrectomy was used to remove the tumor and ERM. A small remnant of tumoral mass remained intact and did not show any growth for 7 years.

Key words: Capillary hemangioma, epiretinal membrane, juxtapapillary, optic nerve, retina

Juxtapapillary retinal capillary hemangiomas (JRCHs) are benign vascular tumors located on or adjacent to the optic nerve head. They often occur in association with Von Hippel–Lindau (VHL) disease but may appear sporadically as an isolated pathology.[1] Treatment is required if there is a reduction in visual acuity (VA) or a progression in tumor size. VA is related to the growth of tumor and secondary complications including exudation, subretinal fluid accumulation, macular edema, and exudative retinal detachment. Glial proliferation appearing within the tumoral mass may also lead to development of epiretinal membrane (ERM) formation and tractional retinal detachment. Actually, the treatment options in JRCH are rather restricted and challenging due to close proximity of the mass to the optic nerve.[1]

Herein, we present a 19-year-old girl who presented with ERM associated with JRCH and was successfully treated with a combination of laser photocoagulation, intravitreal bevacizumab injection, and pars plana vitrectomy (PPV).

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Case Report

A 19-year-old girl was referred to our clinic complaining of progressive reduced vision in her left eye for the past several months. Her best corrected VA was 20/20 in the right eye and 20/400 for the left eye. Biomicroscopic examination of both eyes and fundus examination of right eye were completely normal. Fundoscopy of the left eye revealed a 2.0 × 1.5 mm sized, elevated and round circumscribed, orange tumoral mass which was located in the inferotemporal juxtapapillary region. The tumor was covering more than half of the optic nerve head and protruding mainly to the vitreous and a thick ERM obscuring the foveal zone was also noted [Fig. 1a]. No additional pathology was observed within the peripheral retina.

Fundus fluorescein angiography of the lesion showed signs of early filling with late leakage implying the presence of a vascularized lesion. Optical coherence tomography showed an elevated hyperreflective mass shadowing the inner structures. The ERM extended to the fovea causing diffuse retinal thickening and subretinal elevation adjacent to the optic disc [Fig. 1b]. Systemic workup of the patient and the ophthalmological screening did not reveal any relevant finding of VHL or any other disease. A diagnosis of isolated JRCH was made.

A combined approach was adopted for treatment. In the initial step of treatment, 532 nm argon laser photocoagulation (ALP) was performed with a long exposure time (0.5–1 second) for a deeper penetration. Sufficient energy (150–300 mjoules) to obtain the paleness of the lesion was applied using small spot sizes (50–200 microns). Three consecutive sessions of ALP were applied 5 days apart. An injection of intravitreal bevacizumab was given within 5 days following the last laser session. Apparent regression of the tumor was observed within a few days [Fig. 2a-c]. As the final step of the treatment, 20-gauge PPV (OG) was performed on the first week following intravitreal injection. In surgery, upon lifting the hyaloid, the fibrotic glial capsule of the hemangioma was carefully displaced from the optic nerve with an end-gripping forceps. The external segments of the tumor were partially vitrectomized and the remaining lesion was undermined from its retinal stalk. Consequently, the epimacular membrane was carefully removed. Slight bleeding could easily be stopped by increasing the bottle height and the operation was ended following fluid air exchange. Postoperative improvement was excellent with complete visual recovery except a paracentral scotoma, which persisted most probably due to the loss of ellipsoid zone layer on the nasal side of the fovea [Fig. 3a and b]. Best corrected VA is still 20/20 on both eyes. No growth or

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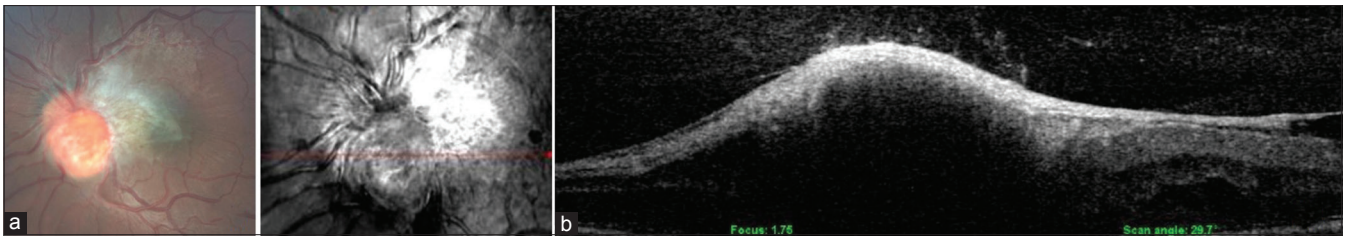

Figure 1: (a). At presentation, fundus examination of the left eye revealed an elevated and round circumscribed, orange tumoral mass which was located in the inferotemporal juxtapapillary region. It was covering more than half of the optic nerve head. (b). Optical coherence tomography showed an elevated hyperreflective mass shadowing the inner structures. The thick ERM extended to the fovea causing diffuse retinal thickening and subretinal elevation adjacent to the optic disc

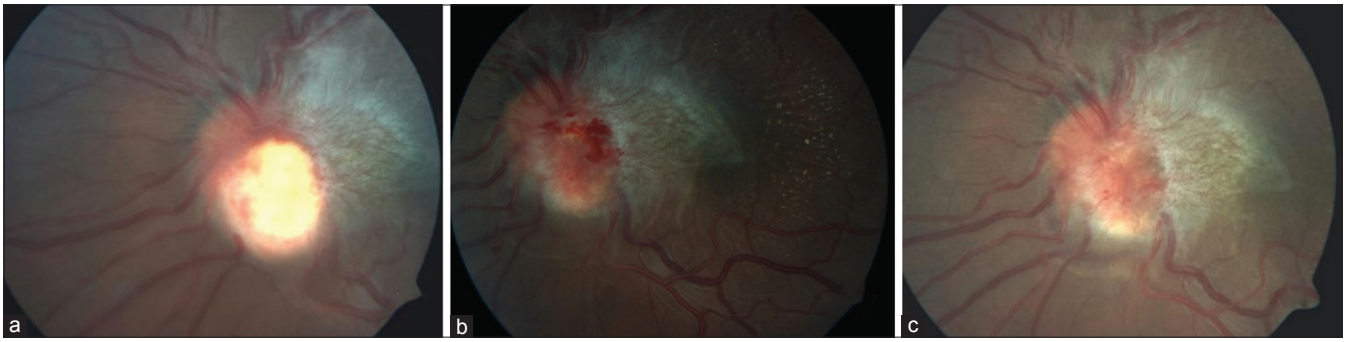

Figure 2: (a). Immediately after the first ALP session, the laser spots were seen on the tumor. (b). Following three ALP sessions, tumor appeared to have shrunk. (c). A few days after the intravitreal anti-VEGF injection, increased fibrotic component and decreased tumor size compared to baseline were noted

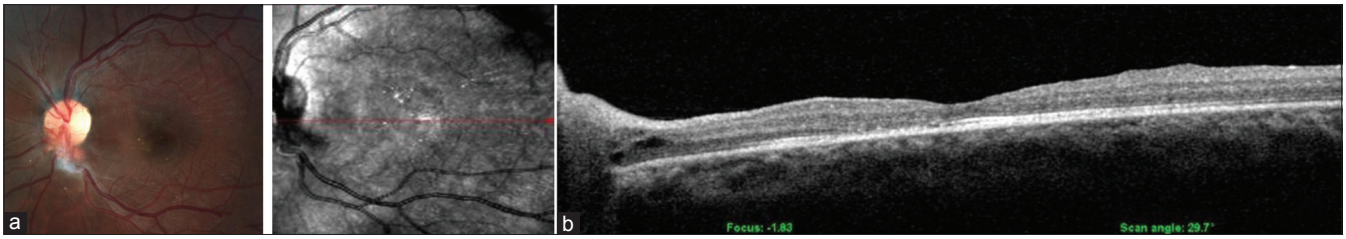

Figure 3: (a). Following the surgery, the small remnant of tumoral mass remained intact at fundus examination and did not show any growth during 7 years of follow-up. (b). Optical coherence tomography showed the loss of ellipsoid zone layer on the nasal side of the fovea. Postoperative improvement was excellent with complete visual recovery except a paracentral scotoma due to the ellipsoid zone damage

activity of the tumor was observed within our 7 years of follow-up to present.

Discussion

There are no accepted definite treatment guidelines for JRCH. Main goal of the treatment is to improve vision and minimize any hazard to the adjacent fragile structures.[1] Several treatment modalities have been suggested alone or in combination. Although favorable treatment outcomes have been reported, none of these approaches were proved to be particularly effective in the regression of JRCH.[1,2]

ALP has been reported to be effective in the treatment of small retinal capillary hemangiomas (RCHs) <3 mm in thickness, with normalization of feeder vessels and regression of exudation. But the destructive collateral damage of ALP limits its application for JRCHs.[3] In order to allocate the total energy and risk of secondary bleeding, consecutive sessions of ALP were performed with small spot sizes for inducing fibrosis and granulation formation. The stromal cells that form the neoplastic hemangioma exhibit vascular endothelial growth factor (VEGF) overexpression.[4] It is well known that elevated ocular levels of VEGF can be detected in patients with VHL disease who present with RCH.[5] In this context, anti-VEGF agents could help to stabilize the tumor and reduce the exudative and hemorrhagic activity thereby lowering the risk of intraoperative bleeding. In literature, only a few cases of JRCH have been reported who were treated with intravitreal injection of different anti-VEGF agents.[2,4,6] Herein anti-VEGF was used to decrease the vascularity and exudative changes. On contrary to our case, JRCHs were uncomplicated or associated with only subretinal hemorrhage in the aforementioned patients. Actually, the presence of thick epimacular membrane necessitated a thorough PPV in our case.

Vitreoretinal surgery has been suggested for severe cases of RCHs, but only a few cases have been reported for JRCH.[7-10]

Majji[7] reported a case of paramacular capillary hemangioma, which was successfully treated with a combination of ALP and subsequent PPV. Kreusel *et al*.[8] described a 6-year-old girl with a juxtapapillary tumor, which was associated with ERM. Both of these cases were successfully treated with PPV and removal of the membranes.[7,8]

In our case, intravitreal bevacizumab was injected following the application of ALP. Hereby, we aimed to achieve a better penetration of anti-VEGF and an enhanced effect within the tumoral tissue upon the destruction of relevant barriers with the use of laser. Indeed, owing to this approach, the tumoral mass quickly showed substantial shrinkage, which provided an easier surgical excision without significant bleeding. Fong *et al*.[9] reported a case of an inferotemporal JRCH associated with ERM and tractional detachment of the macula. This case had a successful combination therapy with intravitreal ranibizumab injection and PDT 1 week before the vitreoretinal surgery. Similarly to our case, they reported reduction of intraoperative bleeding owing to intravitreal anti-VEGF injection before vitreoretinal surgery.[9] However, the tumor could only be partially resected due to its sessile localization. More recently, minimally invasive 25-gauge vitreoretinal surgery combined with half-fluence PDT was reported as an effective and safe procedure in removing epiretinal tractional membrane and JRCH.[10] We did not prefer to use PDT before vitreoretinal surgery due to the existence of thick epimacular membrane. Following the surgery, the tumor became evidently smaller, and required no other supplementary postoperative treatment. The small remnant of tumoral mass remained intact and did not show any growth during the 7 years of follow-up. We have to acknowledge that the convenient location of the tumor enabled a combined therapeutical approach and rendered this pleasing result.

Conclusion

In summary, in cases of JCRH a combined approach with laser photocoagulation and anti-VEGF agent may lead to shrinkage of the tumor preoperatively and reduce the risk of bleeding preoperatively providing an easier surgical excision. This combination therapy may facilitate the surgical management of JRCH cases. Prospective studies with larger scale of similar cases may help to validate this foresight.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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