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Choroidal neo-vascular membrane in a paediatric optic disc pit: A case report

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ABSTRACT

Purpose: To report a paediatric patient with unilateral optic nerve pit found to have choroidal neo-vascular membrane in the same eye.
Observation: An 8-year old girl known to have a unilateral optic nerve pit presented with exotropia of the right eye. On examination, her vision was 20/40; sub-retinal haemorrhage was noted on clinical fundus exam and a sub retinal vascular net was confirmed on optical coherence tomography angiography.
Conclusion and importance: The unusual finding of a choroidal neo-vascular membrane in a known optic nerve pit case represents a new finding of a possible complication. Patients with optic nerve pit may be considered as candidates for OCT-angiography for further diagnosis and proper treatment leading to improved vision, care and prognosis.

1Introduction

Optic nerve pit was first described in 1882 by Wiethe.[1] It presents as a hypo-pigmented, yellow or whitish, oval or round, excavated colobomatous defect of the optic nerve head. The pit is usually found within the inferior temporal portion or central portion of the optic nerve head, sometimes covered with a grey veil of tissue and an emerging cilio-retinal vessel from the optic nerve head margin. Most are unilateral, asymptomatic, and congenital. Optic pit consists of herniation of dysplastic retina into a collagen-lined pocket extending posteriorly often into a subarachnoid space through a defect in the lamina cribrosa.[2–4] Optic pits may lead to serous macular detachment in the second and third decades of life and can be associated with poor prognosis. The macular retinal thickening and detachment typically extend from the optic pit in an oval shape toward the fovea. OCT imaging classically reveals macular intraretinal fluid as well as sub retinal fluid. Various successful treatments have been reported, involving pars plana vitrectomy with or without internal limiting membrane peeling, endo-laser and gas-bubble tamponade placement.[5]

Choroidal neovascularization is an abnormal growth of vessels from the choroidal vasculature into the neurosensory retina through Bruch's membrane. Historically, CNVM treatment included submacular surgery, laser photocoagulation, photodynamic therapy with verteporfin, transpupillary thermotherapy and finally anti-vascular endothelial growth factor (anti-VEGF) agents became the standard of care treatment of choice.[6–12]

2. Case presentation

An 8-year old girl presented to the clinic with a 14 days history of exotropia of the right eye. Her ophthalmic history is positive of having an optic nerve pit in the right eye. On examination, her best corrected visual acuity was 20/40 in right eye and 20/20 in left eye. Her refractive error was +0.75D Sphere OD and +1.25D sphere OS. The pupils were normal with no relative afferent pupillary defect. She had constant exotropia of the right eye with full extraocular muscles movements. The anterior segment was within normal limit bilaterally. Dilated fundus exam of the right eye revealed a grey subretinal lesion in the maculopapular bundle area and with adjacent sub-retinal haemorrhage and sub-retinal fluid. The fundus exam of the left eye was normal. Optical coherence tomography showed the presence of sub-retinal fluid with some subretinal hyper-reflective material, consistent with subretinal haemorrhage (Fig. 1). OCT Angiography was performed and confirmed the presence of a sub-retinal vascular net in the maculopapular bundle area (Fig. 4). After a lengthy discussion with the parents, this patient received Aflibercept injection in the right eye, which was done on her

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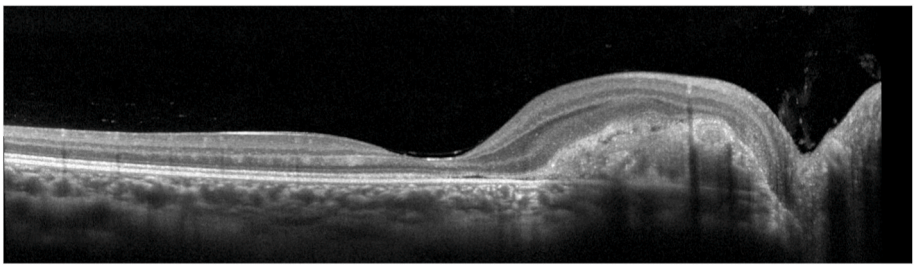

Fig. 1. OCT at presentation showing sub-retinal fluid and sub-retinal hyper-reflective material.

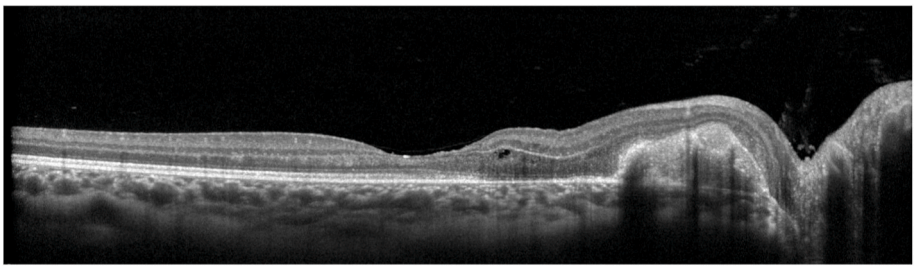

Fig. 2. OCT 3-weeks post Anti-VEGF injection showing resolution of the sub-retinal fluid.

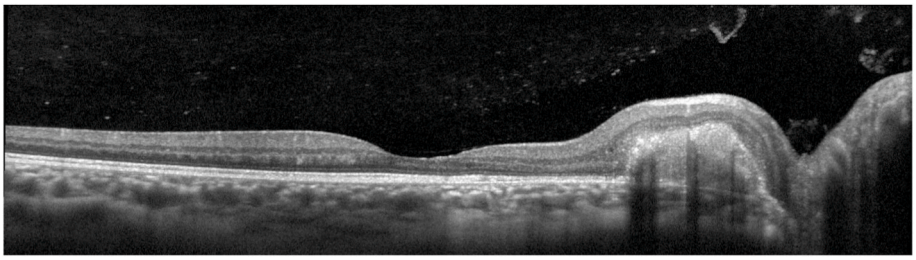

Fig. 3. OCT 67-weeks post Anti-VEGF injection showing stable treatment response.

Fig. 4. OCT Angiography at presentation showing a sub-retinal vascular net in the maculo-papular bundle area.

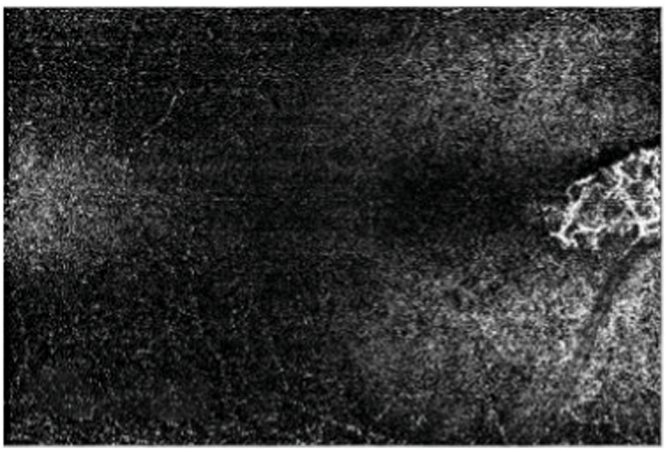

Fig. 5. OCT Angiography 3-weeks post Anti-VEGF injection showing reduction of size of the sub-retinal vascular net.

first visit.

On the second visit, three weeks after the injection, her best corrected visual acuity was the same 20/40 right eye and 20/20 left eye. Intra-ocular pressure measured with I-Care was 23 in the right eye and 17 in the left eye. OCT showed the resolution of sub-retinal fluid (Fig. 2). OCT angiography showed reduction of the size of the sub-retinal vascular net by about 50% (Fig. 5). The intermittent exotropia was much less frequent as noted by both parents and patient.

On the third visit, five weeks after the injection, her best corrected visual acuity improved to 20/30 in right eye and 20/20 in left eye. Anterior segment unremarkable. Posterior segment shows resolved sub-retinal fluid and sub-retinal haemorrhage. OCT shows mild IRF with no significant changes. OCT Angiography shows the sub-retinal vascular net to be unchanged from second visit.

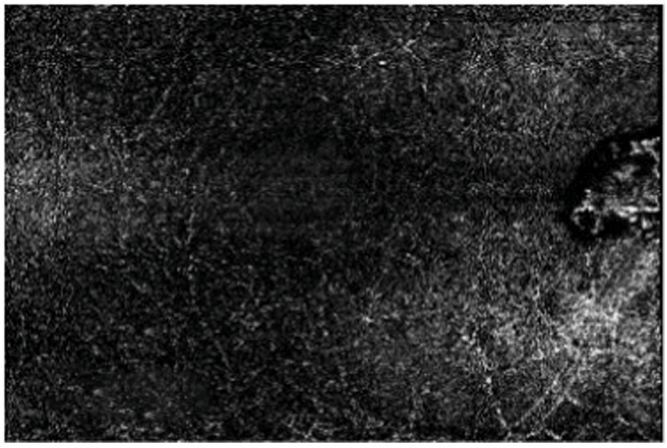

Fig. 6. OCT Angiography 67-weeks post Anti-VEGF injection showing stable treatment response.

On the fourth visit, eight weeks after the injection, her best corrected visual acuity remains unchanged from the third visit. Anterior segment unremarkable and a similar examination of the posterior segment. OCT IRF has improved. OCTA shows same size in the sub-retinal vascular net.

The patient has been followed over a period of 15 months with regular OPD visits. The OCT (Fig. 3) and OCTA (Fig. 6) at 15 months post-injection show similar stable results. The patient did not require any further intervention and there was no recurrence of the exotropia and her vision remained stable.

3. Discussion

Paediatric CNVM is rare and secondary to a variety of etiologies. The presence of CNVM in the context of optic disc pit has not been described in the literature, therefore the choice for the modality of treatment had to be extrapolated from other studies of paediatric retinal diseases with choroidal neo-vascular membrane such as Best disease, post-inflammatory (VKH, serpiginous choroiditis, multifocal choroiditis, unclassified choroiditis, toxoplasmosis, viral retinitis), Stargart's disease, osteogenesis imperfecta, myopia, traumatic CNV, presumed ocular histoplasmosis syndrome (POHS), hamartoma on the retina and RPE, inherited retinal degeneration, reticular dystrophy, optic nerve head drusen, and enhanced S-cone syndrome.[9–15]

Despite the possibility of spontaneous regression of choroidal neo-vascular membrane in paediatric population studies have shown that the mean visual acuity in eyes treated with anti-VEGF were higher than the cases with spontaneous regression highlighting the need for early recognition and treatment.[10] It has been recognized that paediatric CNV require less injections to regress when compared to adults. The reduced number of treatments to achieve resolution of fluid and involution of CNVM may be explained by the relative health of the RPE pump in children compared with adults.[11] In the largest case series of intravitreal antiangiogenic therapy for CNV in paediatric patients, it was concluded that anti-VEGF therapy has the advantage of being the least destructive amongst other treatment modalities and is considered effective with 60% of treated eyes requiring one injection to halt or stabilize progression.[12]

A case series of 4 cases of paediatric CNVM secondary to different etiologies has shown the effectiveness of anti-VEGF treatment leading to resolution of fluid and improvement or stabilization of visual acuity.[11] The first patient of the series, an 8-year old male with choroidal rupture received pegaptinib sodium twice where is vision improved from 20/200 to 20/30 and remained stable for a follow-up period of 14 months. The second patient, a 9-year old female with congenital toxoplasmosis received 4 injections of bevacizumab over a period of 6 months and vision improved from 20/200 to 20/100 also remained stable for 14 months. The third patient, a 15-year old male with congenital hamartoma spontaneously regressed and vision improved from 20/200 to 20/80 but 4 injections of bevacizumab were given over a period of 9 months due to persistent exudate, SRF and macular edema which it helped resolve and maintain over 9-month follow-up period. The fourth patient a 14-year old male with bilateral multifocal choroiditis with pan-uveitis who had multiple surgeries in the left eye (intravitreal anti-VEGF agents were not available at the time) leaving him aphakic with counting finger vision had a drop of vision in the right eye from 20/30 to 20/70 with new SRF and subretinal haemorrhage. After receiving 5 injections of ranibizumab over 11 months vision became stable at 20/70 with no evidence of fluid on OCT.[11] Another case of choroidal neovascularization seen in a 5-year-old boy with enhanced S-cone syndrome followed-up for 7 years post treatment showed improvement of vision from 20/80 to 20/20 with complete disappearance of sub-retinal fluid after being treated with two intra-vitreal injection of Ranibizumab over a period of 2 months and remained stable over a period of 7 years with no detection of ocular or systemic adverse effects of treatment with intravitreal ranibizumab.[15] These cases have been summarized in Table 1.

In regards to our patient, she had received one injection of aflibercept and noticed an improvement of vision from 20/40 to 20/30 with the resolution of sub-retinal fluid and exotropia. Since the number of injections needed to be given is yet unknown and the patient is currently doing well, the decision of monitoring the patient for further management has been taken. The patient has been followed for a period of 15 months and there seem to be no need for further injections and we did not observe any adverse side effects to the injection given. Some of her residual vision loss is due to mild chorioretinal atrophy in the macular area due to chronic intraretinal fluid and subretinal fluid caused by the optic nerve pit itself.

4. Conclusion

We report a case indicating an association between optic disc pit and choroidal neo-vascular membrane. Our case focuses the importance of pattern recognition and actively looking for subretinal haemorrhage in any case of optic nerve pit that presents with subretinal fluid on OCT. For those cases, we strongly recommend OCTA or Intravenous fluorescein angiography.

Consent

Written informed consent was obtained for the case report. This report does not contain any personal identifying information.

Table 1
Pediatric cases in the literature treated with AntiVEGF injections for different indications.

Patient (In order of text)	Age in years	Sex	Eye	Cause	Initial VA	Final VA	Follow-up months	Anti-VEGF	Number of injections
1	8	M	OS	Choroidal rupture	20/200	20/30	14	Pegaptinib	2
2	9	F	OD	Congenital toxoplasmosis	20/200	20/100	14	Bevacizumab	4
3	15	M	OS	Congenital hamartoma	20/80	20/80	9	Bevacizumab	4
4	14	M	OD	Bilateral multifocal choroiditis	20/70	20/70	3	Ranibizumab	5
5	5	M	OD	Enhanced S-cone dystrophy	20/80	20/20	84	Ranibizumab	2

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

We declare no potential conflict of interest with respect to research, authorship and/or publication of this article.

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