CASE REPORT

Paediatric case of peripapillary choroidal neovascularisation associated with optic disc drusen treated with aflibercept

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SUMMARY

Peripapillary choroidal neovascularisation (PPCNV) associated with optic disc drusen is a rare complication that can result in severe vision impairment in children. We report the first case of paediatric PPCNV secondary to optic disc drusen successfully treated with intravitreal aflibercept. A 6-year-old girl presented with a one week history of reduced vision in her right eye with best-corrected visual acuity of 20/500. Fundus examination revealed bilateral elevated discs with a peripapillary pigmentary lesion in the right eye. Optical coherence tomography of the right eye showed marked subfoveal fluid. Both B-scan ultrasonography and fundus autofluorescence demonstrated findings consistent with optic disc drusen. Diagnosis of PPCNV was further confirmed on fluorescein fundus angiography. The child received three intravitreal aflibercept injections with complete resolution of the subfoveal fluid. Her visual acuity improved to 20/25 with no recurrence at a 16-month follow-up. No adverse side effects were reported.

BACKGROUND

Peripapillary choroidal neovascularisation (PPCNV) associated with optic disc drusen is a rare complication that can result in severe vision impairment in children.¹ Treating this subgroup of patients can be challenging because of the paucity of the natural history data and lack of sufficient clinical trials in children.

In recent years, several case reports^{2–8} have been published illustrating the successful use of anti-vascular endothelial growth factor (anti-VEGF) agents, such as ranibizumab and bevacizumab, for the

Figure 2 FFA of the right eye showing evidence of leakage around the disc consistent with active peripapillary choroidal neovascularisation. FFA, fluorescein fundus angiography.

treatment of PPCNV secondary to optic disc drusen in children. We report the first case of successful treatment using the newer intravitreal therapy, aflibercept.

CASE PRESENTATION

A 6-year-old girl presented with a one week history of decreased visual acuity in her right eye. Her best-corrected visual acuity (BCVA) was 20/500 in the right eye and 20/20 in the left eye. There were no other associated ocular or systemic symptoms including headache, nausea or vomiting. There

Figure 1 Optos colour fundus photographs (top) showing a peripapillary pigmented lesion in the right eye with bilateral disc elevations. FAF (below) showing hyperfluorescent spot in the discs consistent with optic disc drusen. FFA, fluorescein fundus angiography.

Figure 3 OCT of the right eye showing marked intraretinal and subfoveal fluid at presentation. OCT, optical coherence tomography.

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Figure 4 OCT of the right eye showing resolution of the subfoveal fluid at 16 months follow-up. OCT, optical coherence tomography.

was no existing or past medical history. There is also no family history of any ocular problems.

Slit-lamp examination of the anterior segment was unremarkable including normal intraocular pressure. Dilated fundus examination revealed bilateral elevated discs with an ill-defined peripapillary pigmentary lesion in the right eye (figure 1). There was no associated haemorrhage, cotton wool spots or exudate in either eye. Cranial nerve and neurological assessment of the child were normal.

INVESTIGATIONS

CT head scan confirmed no mass lesion. Both B-scan ultrasonography and fundus autofluorescence demonstrated areas of hyperfluorescence in the disc consistent with optic disc drusen (figure 1). Diagnosis of active PPCNV was confirmed on the fluorescein fundus angiography (FFA), which showed evidence of leakage (figure 2). Optical coherence tomography (OCT) showed marked intraretinal and subfoveal fluid in the right eye (figure 3).

TREATMENT

Following discussion of treatment options with the parents, the child received 3 monthly injections of intravitreal aflibercept (2 mg in 0.05 mL) under general anaesthesia.

OUTCOME AND FOLLOW-UP

Her visual acuity improved to 20/200 after the first injection and stabilised at 20/25 after the third injection. The subfoveal fluid resolved with no recurrence at a 16-month follow-up (figure 4). No adverse ocular or systemic side effects were reported.

DISCUSSION

Gass⁹ reported the first case of paediatric CNV with optic disc abnormalities in a 4-year-old child with optic disc drusen.

CNV secondary to optic disc drusen is thought to occur more frequently in the paediatric population compared with adults.¹⁰ Optic disc drusen are now recognised as a common association with CNV in children. Studies have reported between 8%–18% of children with CNV have concomitant optic disc drusen.^{11 12} The location of subretinal CNV associated with optic disc drusen is usually peripapillary and can impair vision as a result of extension of subfoveal fluid.¹³

Historically, several treatment modalities have been proposed including observation,¹⁴ laser photocoagulation,¹⁵ photodynamic therapy^{16 17} and surgery,^{18 19} all with variable outcomes. Recently, anti-VEGF agents have been increasingly used as the first line treatment for CNV in paediatric cases.¹² Several cases have been reported in the literature describing the successful use of anti-VEGF agents including intravitreal ranibizumab and bevacizumab for the treatment of CNV associated with optic disc drusen in childhood.^{2–8} We report the first case in the literature of a successful treatment using the newer intravitreal agent, aflibercept.

Table 1 illustrates the demographic characteristics and outcomes of published case reports on the use of anti-VEGF agents for the treatment of PPCNV associated with optic disc drusen in children. The majority of patients achieved excellent visual outcomes of 20/25 and better with complete resolution of subretinal fluid on OCT following anti-VEGF therapy. One patient had a recurrence of subretinal fluid at a 2-month follow-up which resolved with subsequent intravitreal injection, with no further recurrences for 5 years. The number of injections varied from one to three injections. Most patients responded well to a single intravitreal treatment and the number of injections required to stabilise the disease was lower compared with adults.²⁰ Young patients are thought to have a more favourable prognosis because of superior retinal pigment epithelium pump function compared to adults.¹³

In view of the poor BCVA at presentation and significant intraretinal and subretinal fluid threatening vision, we decided to commence the patient on intravitreal aflibercept. Aflibercept is shown to have a greater affinity for VEGF and a longer half-life compared with ranibizumab.²¹ Improvement in BCVA was observed in our patient after the first injection and her BCVA stabilised after three injections. Patients with poor BCVA at presentation tend to require more than one injection to achieve maximal visual improvements.

Anti-VEGF therapy seems to be well tolerated in children for the treatment of CNV with no reported adverse effects at 1-year

Table 1 Demographic characteristics and outcomes of published case reports on the use of anti-VEGF agents for the treatment of PPCNV associated with optic disc drusen in children

Author (year)	Gender	Age, years	Anti-VEGF agent	Number of Injections	Baseline BCVA	Final BCVA	Recurrence	Follow-up duration (months)	Successful treatment	Complications
Gan <i>et al</i> (2018)	F	6	Aflibercept	3	20/500	20/25	No	16	Yes	No
Benouaich <i>et al</i> ² (2016)*	F	9	Ranibizumab	2	20/200	20/25	At 2 months	60	Yes	No
Benouaich <i>et al</i> ² (2016)*	F	9	Ranibizumab	1	20/25	20/20	No	48	Yes	No
Alkin <i>et al</i> ³ (2014)	F	13	Ranibizumab	1	20/80	20/25	No	9	Yes	No
Baillif <i>et al</i> ⁴ (2013)	M	5	Ranibizumab	1	20/60	20/25	No	12	Yes	No
Jedrzejczak-Stróznik <i>et al</i> ⁵ (2013)	F	13	Ranibizumab	1	20/50	20/20	No	30	Yes	No
Delas <i>et al</i> ⁶ (2012)	F	12	Ranibizumab	2	HM	20/60	No	12	Yes	No
Gregory-Evans <i>et al</i> ⁸ (2009)	M	11	Ranibizumab	3	20/100	20/20	No	6	Yes	No

*Bilateral case of PPCNV associated with optic disc drusen.

anti-VEGF, antivascular endothelial growth factor; BCVA, best-corrected visual acuity; PPCNV, peripapillary choroidal neovascularisation.

follow-up based on recent studies.^{12 20} Similarly, there were no ocular or systemic complications reported in all cases including our patient for a follow-up period between 6 months and 5 years.

Learning points

- ▶ Intravitreal aflibercept is effective in treating peripapillary choroidal neovascularisation (PPCNV) associated with optic disc drusen and is well tolerated in children.
- ▶ Excellent anatomical and visual outcomes can be achieved with anti-vascular endothelial growth factor (anti-VEGF) therapy in children with symptomatic PPCNV associated with optic disc drusen. Recurrence of the intraretinal and subfoveal fluid is uncommon.
- ▶ Patients with poor best-corrected visual acuity at presentation tend to require more than one intravitreal anti-VEGF injection to achieve maximal visual improvements.

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