

Punctate inner choroidopathy-associated choroidal neovascular membranes during pregnancy

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Abstract

Purpose and materials Punctate inner choroidopathy (PIC), first described by Watzke *et al*¹, is a disease in young women of child-bearing age. We present three cases of PIC-associated choroidal neovascular membrane (CNVM) occurring during pregnancy, and discuss associated investigative and treatment dilemmas.

Results All three patients described showed evidence of recurrence of CNVM during their pregnancy. None underwent FFA but benefited from OCT monitoring. Different therapeutic strategies were adopted in each of our cases. Case 1, with a history of spontaneous CNVM regression, was managed conservatively. Cases 2 and 3 chose steroid treatment to their better-seeing eye. All cases remained stable postpartum.

Discussion Management of PIC-related CNVM creates diagnostic and therapeutic challenges. The problem is exacerbated as the pathology is often sequentially bilateral and sight threatening. Owing to the rarity of such cases, there is a paucity of evidence on which to base the treatment strategies. A history of pregnancy should always be elicited before investigation with FFA, and women warned of the potential for disease exacerbation with limited therapeutic options during pregnancy.

Conclusions spontaneous resolution of CNVM is common in PIC, and should be borne in mind while treating pregnant women. Peri/intraocular steroid injection represents a reasonable option for sight-threatening CNVM in the better-seeing eye.

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Keywords: punctate; inner; choroidopathy; pregnant; neovascular; membrane

Introduction

Punctate inner choroidopathy (PIC), first described by Watzke *et al*¹ is a disease in young women of child-bearing age. We present three cases of PIC-associated choroidal neovascular membrane (CNVM) occurring during pregnancy, and discuss associated investigative and treatment uncertainties.

Case 1

A myopic 33-year-old woman with known PIC, presented 17 weeks pregnant, with intermittent blurring of her right vision. She had a right recurrent extrafoveal CNVM, previously treated with oral, intravitreal steroids, and surgical membranectomy. Right visual acuity (VA) had been stable at 6/18 in the preceding 2 years.

On examination, best-corrected VA was reduced to 6/24 RE and 6/36 LE. Fundal examination revealed multiple punctate chorioretinal scars in both retinæ and a left macular scar. In the right macula, there was subretinal fluid extending from a raised pigmented lesion (Figure 1), confirmed on optical coherence tomography (OCT) (Figure 2). After discussion with the patient, we decided not to perform fundus fluorescein angiography (FFA), as there was no diagnostic dilemma. VA recovered to 6/9 RE at 3 weeks, and the subretinal fluid spontaneously resolved (Figure 2). Her vision remained stable at 4 months postpartum.

Case 2

A myopic 28-year-old woman, with known PIC, presented 9 weeks pregnant, with a reduction of

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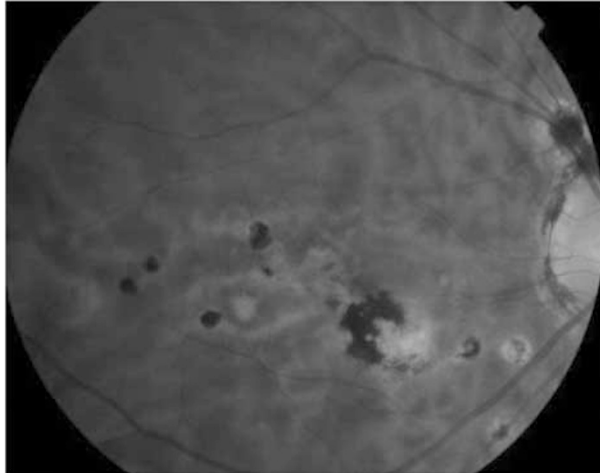


Figure 1 Colour photo of case 1 showing multiple punched-out lesions consistent with PIC and a submacular scar from previous surgery.

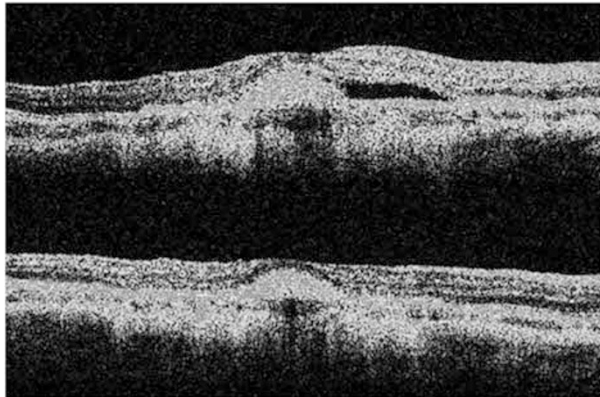


Figure 2 OCT image of case 1 showing subretinal fluid accumulation adjacent to submacular scar (above), and spontaneous resolution after 3 weeks (below).

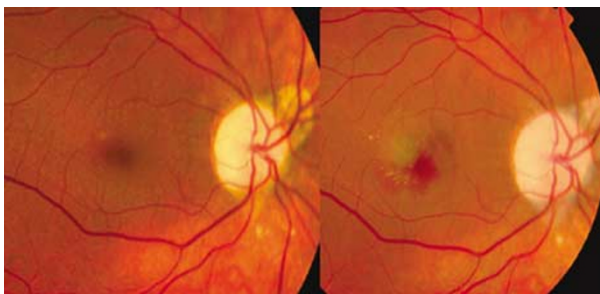


Figure 3 Right macula of case 2, 3 months before presentation (left); subfoveal retinal thickening with adjacent haemorrhage at 9 weeks gestation (right).

her better-seeing right vision of 6/9–6/24. Clinical examination (Figure 3) and OCT revealed new subfoveal fluid accumulation. After discussing all treatment options (including termination of the pregnancy, if visual

loss progressed), she decided to undergo treatment with 4 mg intravitreal triamcinolone. There was resolution of the subretinal fluid with an improved acuity to 6/9 RE at 34 weeks gestation, when she underwent an elective caesarean section. Her vision remained stable at 4 months postpartum.

Case 3

A myopic 28-year-old woman, with known PIC, presented 10 weeks pregnant, with a reduction in her right vision. VA was 6/36 RE and 1/60 LE. She had received oral steroids for bilateral CNVM in the preceding decade. Clinical examination and OCT confirmed new subfoveal fluid accumulation. All treatment options were discussed and she underwent 40 mg sub-Tenon’s triamcinolone injection. The fluid resolved on OCT, and VA remained at 6/36 RE at 2 years postpartum.

Comment

All three patients described showed evidence of recurrence of CNVM during pregnancy. None underwent FFA, but benefited from OCT monitoring. Case 1 had spontaneous resolution. However, cases 2 and 3 required steroid treatment. All cases remained stable postpartum.

Numerous ocular conditions can be exacerbated by pregnancy, which may be related to hormonal or haemodynamic change. Excess levels of endogenous steroids may alter RPE function as well as choriocapillaris’ permeability.² It is thought that excess angiogenic factors such as vascular endothelial growth factor (VEGF)³ and placental growth factor,⁴ which are important in maintaining placental growth, may contribute to the development or recurrence of CNV. However, levels of angiogenic factors have not been consistently found to be raised in maternal serum.⁵ Physiological changes during pregnancy, ie, increased blood volume and cardiac output may also exacerbate choroidal vessels or RPE damage.

Three cases of pregnancy-associated CNVM (idiopathic and presumed ocular histoplasmosis syndrome) have been reported previously.⁶ Two of these patients were treated conservatively during pregnancy, and the third received laser photocoagulation without FFA. Oral and intra-/periorbital steroid, PDT, intravitreal anti-VEGF therapies, and combinations of these measures have been used with varying success to treat PIC-induced CNVM. Therapeutic options during pregnancy, however, are limited due to theoretical teratogenic and fetotoxic effects. Interestingly, PDT administration has been reported in a patient who was

unaware that she was pregnant, with no adverse consequence.⁷

Different therapeutic strategies were adopted in each of our cases. Case 1, with a history of both spontaneous and treatment-induced CNVM regression, was managed conservatively. Cases 2 and 3 chose steroid treatment to their better-seeing eye. Corticosteroids have traditionally been used during pregnancy, eg, in asthma, when the benefits of administration outweigh the risks. Excess levels of foetal corticosteroids have been associated with foetal adrenal suppression, and a moderately increased risk of cleft lip +/- palate.⁸

Management of PIC-related CNVM creates diagnostic and therapeutic challenges. The problem is exacerbated as the pathology is often sequentially bilateral and sight threatening. Owing to the rarity of such cases, there is a paucity of evidence on which to base the treatment strategies. A history of pregnancy should always be elicited before investigation with FFA, and women warned of the potential for disease exacerbation with limited therapeutic options.

In conclusion, spontaneous resolution of CNVM is common in PIC, and should be borne in mind while treating pregnant women. Peri/intraocular steroid injection represents a reasonable option for sight-threatening CNVM in the better-seeing eye.

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