COMMUNICATIONS

RETINAL VASCULITIS ASSOCIATED WITH ANTERIOR UVEITIS*†

BY

J. NOLAN AND J. F. CULLEN

Department of Ophthalmology, Royal Infirmary, Edinburgh

INFLAMMATION of the anterior uvea is often accompanied by inflammatory changes in the posterior uvea, because the iris, ciliary body, and choroid are parts of a single entity. Anterior uveitis accompanied by a retinitis or retinal vasculitis presents a much rarer combination in spite of the functional dependence which the retina comes to have upon the choroid. This is no doubt because the retina and the posterior uvea are morphologically different structures.

Nonetheless, retinal vasculitis associated with an anterior uveitis does occur, and we have seen five such cases in the last 4 years. All the patients belonged to the younger age group (16 to 39 years) and, despite full investigations in every case, no definite aetiology has been established in any, although one (Case 5) had been diagnosed in the past as an example of Reiter's syndrome.

Case Reports‡

Case 1, a 24-year-old games mistress, first presented on April 17, 1962, with a mild anterior uveitis in the right eye. The corrected visual acuity was 6/18. Despite local treatment, the condition remained unchanged for 2 months, when she developed a swollen optic disc with tortuous retinal veins and superficial retinal haemorrhages (Figs 1 and 2, overleaf). Macular oedema was marked and a "pseudo-hole" was present. Oral prednisolone therapy was therefore started, 30 mg. daily in divided doses initially, reduced after a week to 20 mg. daily; 2 weeks later the dosage had been reduced to 10 mg. daily, but the retinal picture worsened, a macular star developed, and the dosage was raised again to 20 mg. prednisolone daily. The condition thereafter improved steadily, and 7 weeks after starting systemic treatment the optic disc was flat and the haemorrhages were absorbing (Fig. 3, overleaf). Prednisolone was discontinued. The visual acuity was then 6/6. The hard exudates at the macula disappeared within a year.

12 months later there was a similar inflammatory episode in the left eye, with anterior uveitis, a swollen optic disc, venous engorgement, and retinal oedema. The visual acuity was reduced to 6/18. As she was 8 months pregnant, labour was induced and after delivery of a healthy baby treatment was started with oral prednisolone, 20 mg. daily. Within one week the disc had become flat. It took 6 weeks for the veins to return to normal and 2 months for the retinal oedema to disappear and the vision to improve to 6/6.

This patient has now been followed up for over 3 years, during which time she has had one mild attack of anterior uveitis in the right eye which settled quickly on local treatment without any

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† Address for reprints: As above

‡ Note: All the patients were treated from the outset with topical corticosteroid and atropine drops.

FIG. 1.—Case 1. Right eye, showing swollen optic disc with engorgement of retinal veins and retinal oedema.

FIG. 2.—Case 1. Right eye, showing pre-retinal haemorrhages in mid-fundus.

FIG. 3.—Case 1. Right eye after 7 weeks' treatment.

FIG. 4.—Case 4. Right eye, showing neovascularization, fibrous tissue overlying the optic disc, and retinal oedema.

retinal involvement, and also (in 1963) bilateral acute glaucoma, which was treated by bilateral Scheie operations.

When last seen on June 30, 1965, the visual acuity was 6/6 in the right eye, and 6/5 in the left. The intra-ocular pressure was normal in each eye, inflammatory signs were absent, and the fundi were within normal limits.

Case 2, a 29-year-old housewife, presented on August 17, 1963, with bilateral anterior uveitis. Both fundi showed swollen optic discs and retinal oedema with "star" formations at the maculae. Visual acuity in the right eye was less than 6/60 and in the left 6/9. Treatment was started with oral prednisolone, 40 mg. daily in divided doses; 6 weeks later the fundi showed considerable improvement, the prednisolone having been reduced to 15 mg. daily. Systemic treatment was continued for a further 4 months, by which time the fundi had returned to normal except for slight stress lines at the maculae. The visual acuity had improved to 6/18 in the right eye, and 6/6 in the left.

The patient has now been followed up for 2½ years. Visual acuity has not improved further, despite inactivity of the uveitis over this period.

Case 3, a 16-year-old schoolboy, presented on August 27, 1963, with a 2 weeks' history of painful red eyes. He was found to have bilateral anterior uveitis, and there were inflammatory cells in the vitreous. The visual acuity was 6/9 in the right eye and 6/12 in the left. The fundi were normal.

One week later both eyes had developed retinal oedema with venous engorgement. Treatment was started with systemic prednisolone, 30 mg. daily in divided doses, and 2 weeks later the fundus appearances were vastly improved, the corticosteroids having been gradually reduced to 15 mg. daily; 4 weeks after starting systemic treatment, both fundi were normal except for slight stress lines at the maculae and a localized dilatation of the right supero-nasal vein. The prednisolone was discontinued. Visual acuity was 6/6 in each eye, and in the follow-up period of over 2 years no further inflammation has occurred.

Case 4, a 39-year-old bus driver, presented on August 6, 1965, with anterior uveitis and inflammatory cells in the vitreous in the right eye. There were no keratic precipitates and no abnormality was seen in the fundus. The visual acuity was 6/24. 3 weeks after the onset of symptoms a peculiar whitish patch was seen above the right disc, the detail of which was obscured by vitreous haze. The visual acuity had dropped to 6/36. Treatment was started with oral prednisolone 40 mg. daily, and there followed a gradual clearing of the ocular media. The corticosteroid dosage was gradually reduced to 10 mg. daily, and 3 weeks after starting treatment, it could be seen that the whitish patch was not an area of choroiditis, but a proliferation of fibrous tissue with fine new vessels in it. This neovascularization progressed and marked venous changes in the form of dilatation and tortuosity developed. After 2 months, these changes began to subside, and the systemic corticosteroids were then discontinued.

In February, 1966, the fundus appearance had improved considerably but the disc was still hyperaemic, with new vessels and fine fibrous tissue overlying it (Fig. 4), slight retinal oedema, and a faint "pseudo-hole" at the macula. The visual acuity was 6/9.

Case 5, a 24-year-old engineer, presented on September 9, 1965, with an intense anterior uveitis and hypopyon in the right eye. The visual acuity was 6/36. He had a previous history of Reiter's syndrome associated with *Shigella sonnei* dysentery, and had had at least one other severe attack of right anterior uveitis, which occurred 7 years previously and was accompanied by disc swelling, retinal venous engorgement, and retinal oedema: these changes had subsided gradually over the course of 5 months on local treatment only.

He was treated with subconjunctival Depomedrone injections and, after 5 weeks, it was possible to see the fundus, which showed a retinal vasculitis with haemorrhages on the disc and a subhyaloid haemorrhage below the macula. After 8 weeks, the anterior chamber was quiet and the fundus changes were subsiding. After 13 weeks, the eye was normal except for the absorbing subhyaloid haemorrhage and the visual acuity had improved to 6/6 partly.

Comment

In all the cases reported above, the retinal veins were markedly affected, but not the arteries, and there was disc swelling and retinal oedema. In two cases there were retinal haemorrhages and in one there was neovascularization. In no instance was there any frank choroiditis.

Every case responded to corticosteroids, given either systemically (4 cases), or by subconjunctival injection (1 case), and the major inflammatory changes in the fundi disappeared within 1 to 4 months.

Without any histological evidence it is impossible to be certain of the exact nature or cause of these fundal changes and the literature is not helpful on the subject. Perkins (1961) mentioned that retinal vasculitis occurred in 35 out of his total of 1,718 cases of uveitis, but 31 of these were fairly typical cases of Eales's

disease, and two others were examples of sickle-cell retinopathy. The remaining two cases are listed as "atypical vascular lesions", and these are the only ones in his series which might be similar to those described above, but as no further descriptive details are given, an exact comparison cannot be made.

Woods (1961) stated that the ocular changes in Behçet's and the Vogt-Koyanagi-Harada syndromes probably begin in the optic nerve, spreading to the retina and thence secondarily to the uveal tract. Early cases of Behçet's syndrome usually show a narrowing of the retinal arterial bed and evidence of a periarteritis or endarteritis, sometimes with retinal haemorrhages. None of our five patients, however, had any other evidence of these syndromes.

The explanation in our cases may be that vasodilator metabolites or toxins from the anterior uveal inflammation diffuse across the vitreous body and affect the retinal vessels. The posterior pole shows the maximum changes because it is the most vascular part of the retina (Warwick, 1963). Furthermore, the retinal oedema caused by this inflammatory exudate would spread to involve the optic nerve head, thereby embarrassing the retinal venous return and causing the venous changes which been noted.

Boyd (1961) pointed out that mild inflammation causes maximum dilatation of small vessels and capillaries in the area involved, together with the actual formation of new capillaries. There is an accompanying escape of various amounts of protein and blood cells. In a more severe case the escape of fibrinogen might result in fibrin formation and the appearance of fibrous tissue, as in our Case 4.

Other causes are less likely. Thus an underlying diffuse inflammation of the posterior uvea would cause more obvious signs of choroiditis, such as milky oedema or exudative detachment. Again, in no patient was there any evidence of a generalized disease which might have affected the anterior uvea and the retina simultaneously, such as was described by Woods (1961) in cases of infectious mononucleosis or septicaemia.

The logical form of treatment in the event of the aetiology suggested would be the administration of systemic or subconjunctival corticosteroids, and we have seen that all the cases thus treated did, in fact, respond to such therapy with return of normal or near-normal vision in each instance.

Summary

Five cases of anterior uveitis accompanied by retinal vasculitis are described. The possible pathology of the retinal changes is discussed, and the response to corticosteroid therapy emphasized.