

Optic disc vasculitis

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Lyle and Wybar (1961) reported seven cases of a condition which they designated "retinal vasculitis". Lonn and Hoyt (1966) described a group of five similar cases under the title of "papillophlebitis". Eleven identical cases were described by Cogan (1968) with a diagnosis of "mild retinal and papillary vasculitis". Recently nine more cases have been reported as "benign retinal vasculitis" (Hart, Sanders, and Miller, 1971). The main features of the condition described by these authors under four different titles are:

- (1) One eye only was involved in all cases.
- (2) The patients were usually healthy young adults.
- (3) The only symptom was generally a vague foginess of vision in one eye.
- (4) The visual acuity was usually normal or insignificantly affected and was almost always restored to normal in due course.
- (5) There was marked oedema of the optic disc (ODO).
- (6) There was marked dilatation and tortuosity of the retinal veins.
- (7) Variable amounts of retinal haemorrhage were observed on and around the optic disc (OD).
- (8) Corticosteroid therapy was not effective, and recovery was not affected by treatment (Lonn and Hoyt, 1966; Cogan, 1968; Hart and others, 1971).
- (9) The condition had a benign though protracted course (6 to 18 months—Lonn and Hoyt, 1966; months or years—Cogan, 1968; several up to 12 months—Hart and others, 1971) without serious complications.
- (10) Sequelae were perivenous sheathing of the large retinal veins and dilated vessels on the OD.

In addition to these definite cases described in the literature, reports of identical cases have been included erroneously among cases of central retinal vein (CRV) occlusion or unilateral ODO (Klien, 1944; Roper-Hall, 1958; Huber, 1961; and others).

I have studied in detail eight cases which generally conformed very well to the above description, but also revealed interesting additional features. The present study suggests that these cases fall into two types in spite of their having some common features.

Type I

ODO is the dominant feature which clinically and fluorescein angiographically is similar to ODO of other aetiologies, *e.g.* intracranial hypertension, except that it is almost always unilateral. It responds satisfactorily to adequate systemic steroids, and is a benign condition.

Type II

Clinically and fluorescein angiographically it resembles CRV occlusion. It responds comparatively less favourably to systemic steroids. This is usually a benign condition but the outcome depends upon the site and extent of the CRV occlusion, as discussed in the text.

The primary objects of the present communication are (*a*) to stress the existence of such a condition, (*b*) to describe the signs and symptoms, (*c*) to show the beneficial effect of adequate steroid therapy in shortening the course of the disease and preventing complications, and (*d*) to discuss its pathogenesis.

Observations

The Table (pp. 654 and 655) summarizes the findings in the eight cases of optic disc vasculitis (ODV) in the present study. These have been analysed separately for Type I and Type II, so as to determine the similarities and differences between the two categories. Intravenous fluorescein angiography was performed in all cases on first examination and in half as a follow-up procedure as well.

Treatment*

TYPE I

(*a*) Three patients were treated with systemic corticosteroids with an initial dose of 80 mg. prednisolone (or equivalent dosage of other steroids) per day. Subjectively and objectively they showed a dramatic response. Within a week, sometimes within 48 hours, the haziness of vision cleared and the ODO was significantly reduced (Compare Fig. 3*a, b* pre-treatment with Fig. 3*e, f* after treatment for one week). Once the response was observed, the dose was gradually reduced. In 3 to 6 weeks the disc was perfectly normal and the patients had no complaint. The steroids were then tailed off, although one patient required a maintenance dose for about 6 months because rapid reduction or stoppage produced subjective symptoms. There was no residual effect.

(*b*) No steroids were given to one patient because of the reluctance of the doctor in charge. It was more than 4 months before the visual acuity returned to normal. After about 8 months the ODO cleared and some pallor of the disc was seen.

TYPE II

(*a*) Three patients in this group had systemic corticosteroids in an initial dose of 40 mg. prednisolone daily. The response was in no way as satisfactory as in patients of type I treated with 80 mg. daily.

(*i*) One of these three cases was treated with regular control of the dose according to his response. The fundus returned to normal within 4 months. Subjective improvement was seen within a week.

(*ii*) The other two patients were treated rather haphazardly with steroids. Since the response was not so dramatic with the low dosage, the ophthalmologists treating them tended to reduce steroids or stop them early. A rapid reduction of the dose to 20 mg. or less in about a week resulted in a deterioration of the visual acuity and fundus appearances, whereas increasing the dose to a higher level produced an improvement. One of these patients returned to normal after 7 months, while the other, when last seen after 16 months, had a slight permanent deterioration of vision, with some sheathing of the retinal veins.

* See Addendum, p. 670

Table Summary of findings in eight cases of optic disc vasculitis

Type	I	II
No. of patients	4	4
Age (yrs)	30-35	26½, 34, 44, 55
Sex	2 males, 2 females	3 males, 1 female
Visual acuity	As good as, or one line less than, the fellow normal eye, except in one with 3 lines	As good as, or one line less than, the fellow normal eye
Presenting symptoms	Vague blurring of vision	Vague blurring of vision
Duration of symptoms (wks)	1-2	Few
Fundus	<p><i>A. Optic disc oedema:</i></p> Severity Distribution Unilateral Bilateral Central depression in OD Hyperaemia Microaneurysms	Mild to moderate (Figs 4, 5, 6) OD and PPR In four In none Present in all Present, may be more than in Type I On OD, PPR, and posterior part of fundus (Figs 4b, 5c)
	<p><i>B. Retinal veins:</i></p> Engorgement and tortuosity Sheathing Retino-ciliary veins	Very marked (Figs 4, 5, 6) Absent Present in 2 (Fig. 5)
	<p><i>C. Retinal arterioles:</i></p> Periarterial exudative sheathing on OD and PPR	Patchy sheathing in one faint white sheathing along main retinal arteriole in another
	<p><i>D. White exudates</i> (?cotton wool spots):</p> Present Site Shapes Sizes	In three On surface of OD and PPRs usually along arterioles Irregular patches Small patches
	<p><i>E. Haemorrhages:</i></p> Number Shape Size	Few to many Flame-shaped and some deep Variable Small patches

Microaneurysms	Absent	Present in almost all
G. White deposits (<i>Hard exudates</i>)	Fine punctate, in PPR and nasal to fovea in one (Fig. 1)	A patch supero-temporal to macula in eye with macular oedema
Fluorescence fundus angiography	A. During transit of dye: (1) Distribution of microaneurysms and dilated capillaries (2) Retinal veins (3) Circulation time (i) Arterial phase (ii) Venous phase (4) Retino-ciliary veins	Large number, thickly scattered in area of OD, macular region, and most of posterior fundus (Figs 4b, 5c) Markedly distended (Figs 4, 5)
Other ocular abnormality	(1) In area of distribution of microaneurysms (Figs 2c, 3c, d) (2) No perivenous fluorescence along main retinal veins outside PPR (Figs 3c, d) (3) No macular abnormality Few cells in posterior vitreous in one	Normal Much more prolonged than in Type I Seen in 2 (Fig. 5)
Visual fields	Enlarged in all	(1) In area of distribution of microaneurysms (Figs 4d, 5d) (2) Marked perivenous fluorescence along main retinal veins, more marked in their concavities (Figs 4c, d, 5d) (3) Evidence of macular oedema in one eye Few cells in anterior chamber and vitreous with thickened retro-ocular optic nerve in one Less enlarged than in Type I
Scotomata	(1) In centro-caecal area — one (2) Superior nasal sector — one (3) Temporal to fixation — one	Small scotomata corresponding to the retinal haemorrhages
Associated systemic diseases	(1) Maxillary sinusitis — one (2) Childhood history of TB — one (3) 13-week pregnancy in eighth para with bilateral ODV — one	(1) Penicillin allergy — one (2) Skin depigmentation — one
Follow-up period (yrs)	1-3	1-3
Recurrences	Nil	Nil
Sequelae	None	(1) Macular oedema with loss of central vision, venous sheathing — one (2) Deterioration of VA from 6/6 to 6/9-6/12 with venous sheathing — one (3) Retino-ciliary veins on OD — two

(b) One case of this type was not treated with steroids. He developed marked macular oedema which reduced the visual acuity to counting fingers (Fig. 6) with a permanent central scotoma. In addition, he developed swelling of the retro-ocular part of the optic nerve with axial proptosis of 2 mm. and was suspected of having an optic nerve tumour; 2 years after the onset, the fundus and optic nerve lesions resolved completely, leaving a permanent central scotoma and sheathing of the retinal veins.

Discussion

In the literature so far, 32 cases of this disease have been described. This does not, however, represent the true incidence of the condition, since the vast majority of such cases are erroneously diagnosed either as unilateral ODO of obscure aetiology (Type I cases) or as incipient CRV occlusion (Type II cases), and there is an equal likelihood that a considerable number of these patients never seek a medical opinion because the visual symptoms are comparatively slight. Since the condition is usually benign and self-limiting, the anxiety of ophthalmologist, neurologist, or physician is usually relieved by the eventual spontaneous recovery of the patient and such cases are ultimately labelled as "interesting" or "obscure".

CLINICAL PICTURE OF OPTIC DISC VASCULITIS (ODV)

The following is a description of this disease based on my eight cases and on a study of the 32 cases reported in the literature:

(1) *Laterality*

All the cases reported in the four previous reports were unilateral. In the present series one case of Type I was bilateral – more marked in one eye than the other (Fig. 3a, b). When unilateral the lesion had no special predilection for either side. Munro (1971) saw a Type II patient who developed a possible recurrence in the other eye 2 years later. Hart and others (1971) reported one patient, in whom after 9 months' interval mild ODO and some retinal haemorrhages were seen in the second eye; however, that patient had malignant hypertension.

(2) *Age*

The condition is usually seen in young adults. The age groups were 35 ± 12 years in Lyle and Wybar (1961), 41 ± 17 years in Lonn and Hoyt (1966), 31 ± 9 years in Cogan (1968), 28 ± 6 in Hart and others (1971), and 36 ± 9 years in the present series. Type I patients of the present series were slightly younger than Type II patients* (Table); statistical analysis revealed no significant difference in the age incidence between the two types or in the ages of the cases described by the authors.

(3) *Sex*

Lyle and Wybar (1961) found that the condition occurred more often in men than women (5 males, 2 females). The findings of Hart and others (1971) were similar (8 males, 1 female). Lonn and Hoyt (1966) had three males and two females. According to Cogan (1968), the disease was equally distributed between the sexes. In the present series, there were three females and five males. A further splitting of the present series into the two types showed that in Type I the sexes were equally distributed while in Type II there were three males and one female. As far as can be judged from the previous

* This can also occur in a much older age group (See Addendum p. 670)

reports, most of the cases of Lyle and Wybar (1961), Lonn and Hoyt (1966), and Hart and others (1971) fit into Type II and those of Cogan (1968) into Type I. This could explain the differences in sex distribution reported by Lyle and Wybar (1961) and Cogan (1968). However, the number of cases is too small to draw any definite conclusion.

(4) *Presenting symptoms*

The presenting symptom is usually an intermittent vague blurring or haziness of vision for a few weeks, which may be worsened by stooping or by hot baths. It may occur for some time after the patient wakes in the morning*. There may also be black spots, flickering or shimmering of light, etc. In one patient there was also a dull ache behind the eyeball. In only one case of the present series was there a definite sudden reduction in visual acuity. There was no other ocular complaint.

(5) *Visual acuity*

The visual acuity in the affected eye is either normal or no worse than 6/9 except for the occasional case in which marked deterioration may occur. The latter is usually seen when there is an associated macular lesion, *e.g.* haemorrhage or oedema.

(6) *Ophthalmoscopic examination of the fundus*

Type I (Figs 1, 2, 3)

Fundus appearances in this group are identical to those seen in patients with ODO of any aetiology, *e.g.* intracranial hypertension. The main finding is the marked ODO in the affected eye with a perfectly normal fundus in the fellow eye, except for the rare case (Fig. 3) with bilateral involvement. The disc is hyperaemic. The central depression in the disc is still present, contrary to the opinion held by most ophthalmologists and neurologists. The retinal veins are moderately dilated and engorged. The retinal arteries show no abnormality except for a variable amount of sheathing of some arterioles with white exudates on the OD and peripapillary region (PPR) (Fig. 3a). Small irregular patchy white exudates (indistinguishable from cotton-wool spots) lying on the surface of the nerve fibres of the OD and PPR are seen in all cases, in addition to the periarterial

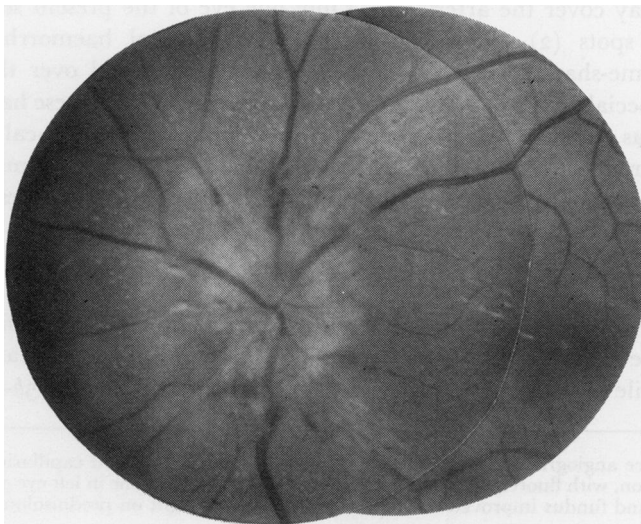


FIG. 1 *Left fundus in a 35-year-old man, one week after noticing a greenish-grey patch in outer part of his field. Visual acuity normal. Markedly enlarged blind spot. Noticed distinct improvement within 48 hours of starting prednisolone 80 mg. daily; after 1 week much less ODO with no symptoms*

* Particularly in Type II

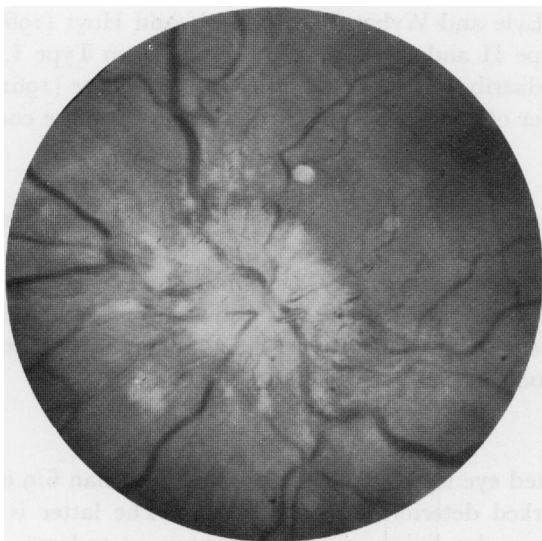


FIG. 2 (a) *Left fundus in a 30-year-old woman, 3 weeks after onset of mistiness. Visual acuity 6/9. Enlarged blind spot with small centrocaecal scotoma. Some cells in posterior vitreous*

sheathing. There is either no retinal haemorrhage or only occasional tiny flame-shaped haemorrhages on the swollen disc or at its edges. In one eye fine punctate white deposits ("hard exudates") were seen in the PPR and nasal to the fovea 1 week after the onset of symptoms (Fig. 1).

Type II (Figs 4, 5, 6)

The appearance of the fundus in this group is like that of pure CRV occlusion (Hayreh, 1971). The main feature of the fundus appearance is a marked degree of distension, engorgement, and tortuosity of the retinal veins with no sheathing. All the cases reported so far have been unilateral. There may be retino-ciliary veins (Hayreh, 1969) on the OD which are collateral channels to short-circuit the blood from the CRV to the choroidal circulation (Fig. 5a, b). The OD swelling is mild to moderate (less marked than Type I), with marked hyperaemia. The retinal arterioles are normal. Small irregular patches of white exudates (indistinguishable from cotton-wool spots), situated at the OD and PPR, may cover the arterioles. Only one eye of the present series showed classical cotton-wool spots (2) on the retina (Fig. 6). Retinal haemorrhages varied from a few small flame-shaped ones to a large number, distributed over the posterior part of the fundus, especially above, below, and temporal to the OD. These haemorrhages are not as extensive as those seen in haemorrhagic retinopathy of the so-called classical CRV occlusion, but are more numerous than those in Type I. The macula may occasionally be involved by flame-shaped haemorrhages or oedema (Fig. 6) which varies in severity. Rarely, white deposits may be seen near the macula.

(7) *Intravenous fluorescein fundus angiography*

The pattern is given in the Table and has been found to differ distinctly from Type I to Type II. In Type I cases, the pattern resembles that seen in ODO of any aetiology (Fig. 2b, c; 3c, d), while in Type II it is that of CRV occlusion (Fig. 4b-d, 5b-d).

FIG. 2 (b, c) Fluorescence angiograms of (a), showing microaneurysms and dilated capillaries on the optic disc and peripapillary region, with fluorescein leak. Retinal venous circulation time in left eye 56 sec. (in right eye 23 sec.). Symptoms and fundus improved dramatically within a fortnight on prednisolone 80 mg. daily

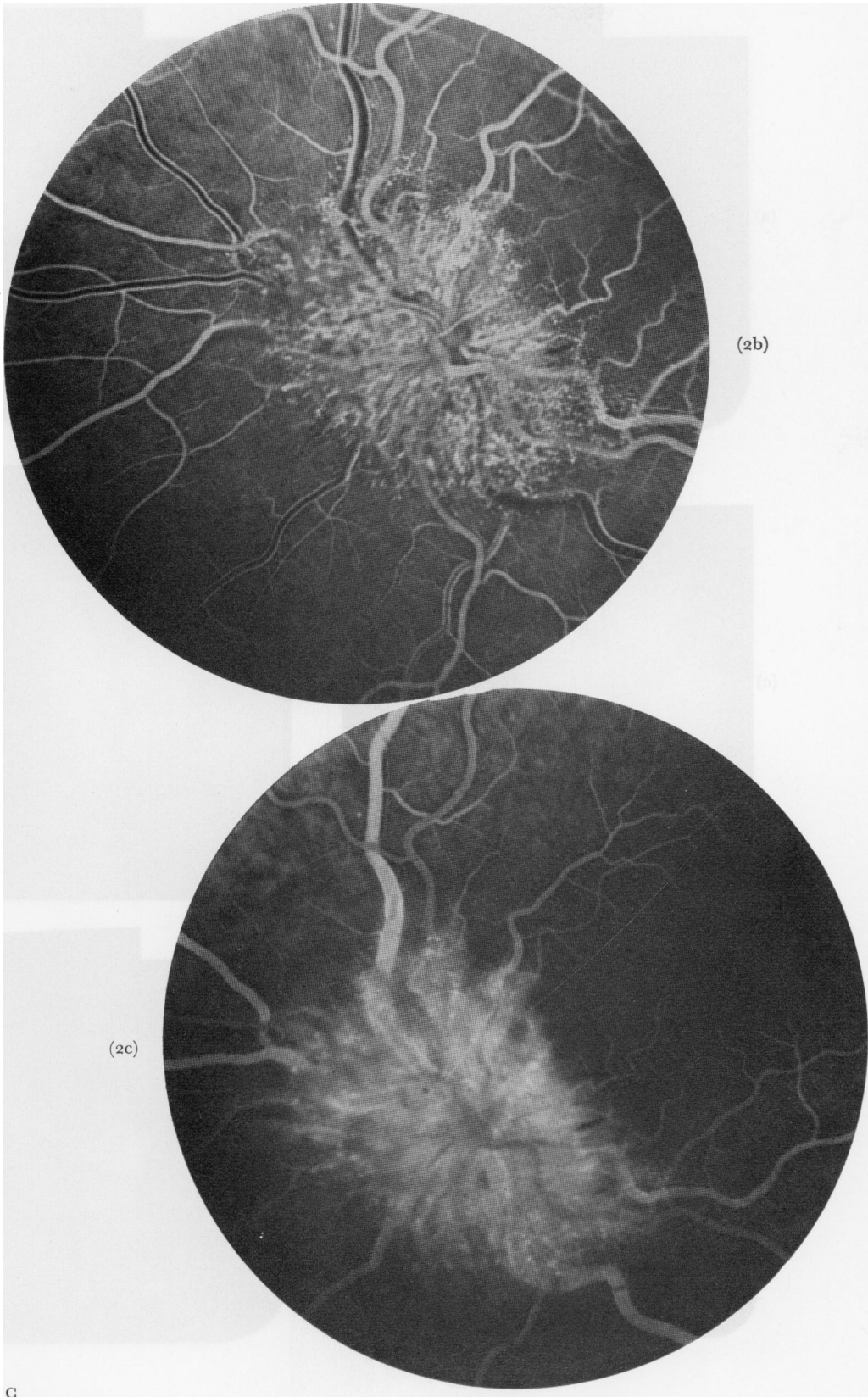
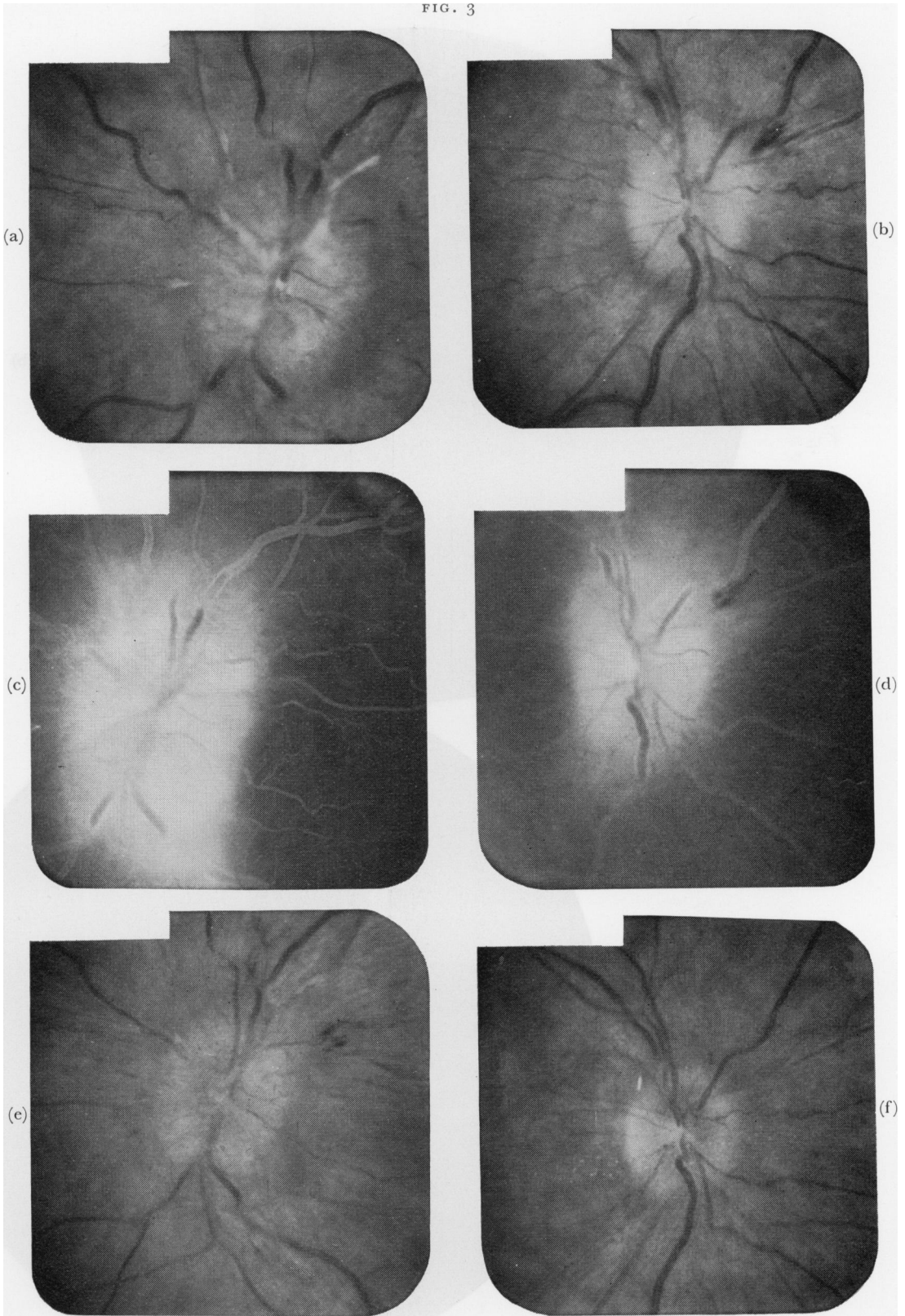


FIG. 3



(8) *Other ocular findings*

There is usually no ocular abnormality. A few findings, suggestive of uveitis, were noticed in two cases of the present series (Table). The presence of cells in the anterior chamber of the eye has been explained by Hart and others (1971) on the grounds of ocular ischaemia. There is no evidence of ischaemia in the eyes of this group of young patients and this fact was admitted by Hart and others (1971) themselves when they wrote: "The absence of arterial disease with maintenance of a normal arterial pressure may be the main reason for the good prognosis in this group".

The thickening of the retro-ocular optic nerve in one case was presumably due to extension of vasculitis to involve the optic nerve meninges, especially the pia, because of the continuity of pial vessels of ciliary origin with those of the ciliary vessels of the disc, (Hayreh, 1969).

(9) *Visual fields*

A definite enlargement of the blind spot is seen, more marked in Type I than in Type II, and this depends upon the severity of ODO. Sometimes, small field defects may be seen in the central field (Table). The peripheral fields are normal.

(10) *Associated systemic diseases*

All the cases so far reported have shown that these patients are perfectly healthy individuals with no significant abnormality. Cogan (1968), in his eleven cases, found lupus erythematosus in one, cerebral vasculitis in one, and non-specific vasculitis of the skin in one. Lyle and Wybar (1961) found active pulmonary tuberculosis in one case. One of the patients of Hart and others (1971) had malignant hypertension. Associated systemic conditions seen in the present series are shown in the Table.

(11) *Treatment*

The opinion expressed by previous workers has been that no treatment is required in these cases because the condition benign and self-limiting. Corticosteroids or anticoagulants have, however, been tried in some cases.

(a) *Steroid therapy*

Lyle and Wybar (1961) gave prednisolone to two patients; the dosage was 25 mg. daily in one and was not stated in the second. These patients showed a slow response but a relatively quicker recovery than untreated cases. Lonn and Hoyt (1966) and Hoyt (1971) treated four cases with systemic steroids—two with 60 to 80 mg. prednisolone and two with smaller doses; none showed any response. Hart and others (1971) treated three cases with 30 to 40 mg. prednisolone but with no specific efficacy. Cogan (1968) gave steroids in three cases (no dosage mentioned) and found beneficial results in only one.

FIG. 3 (a) *Left fundus* and (b) *Right fundus* in a 31-year-old woman, during 13th week of her 8th pregnancy, 1 week after onset of hazy vision. Note periarterial sheathing on optic disc and peripapillary region in (a) and more marked ODO in (a) than (b). Visual acuity normal

(c) and (d) Fluorescence angiograms of (a) and (b) respectively during the late phases, showing fluorescein leak

(e) and (f) Fundus appearances of (a) and (b) respectively 1 week after the start of Decadron 4 mg. thrice daily; note regression of ODO and disappearance of periarterial sheathing, with no symptoms

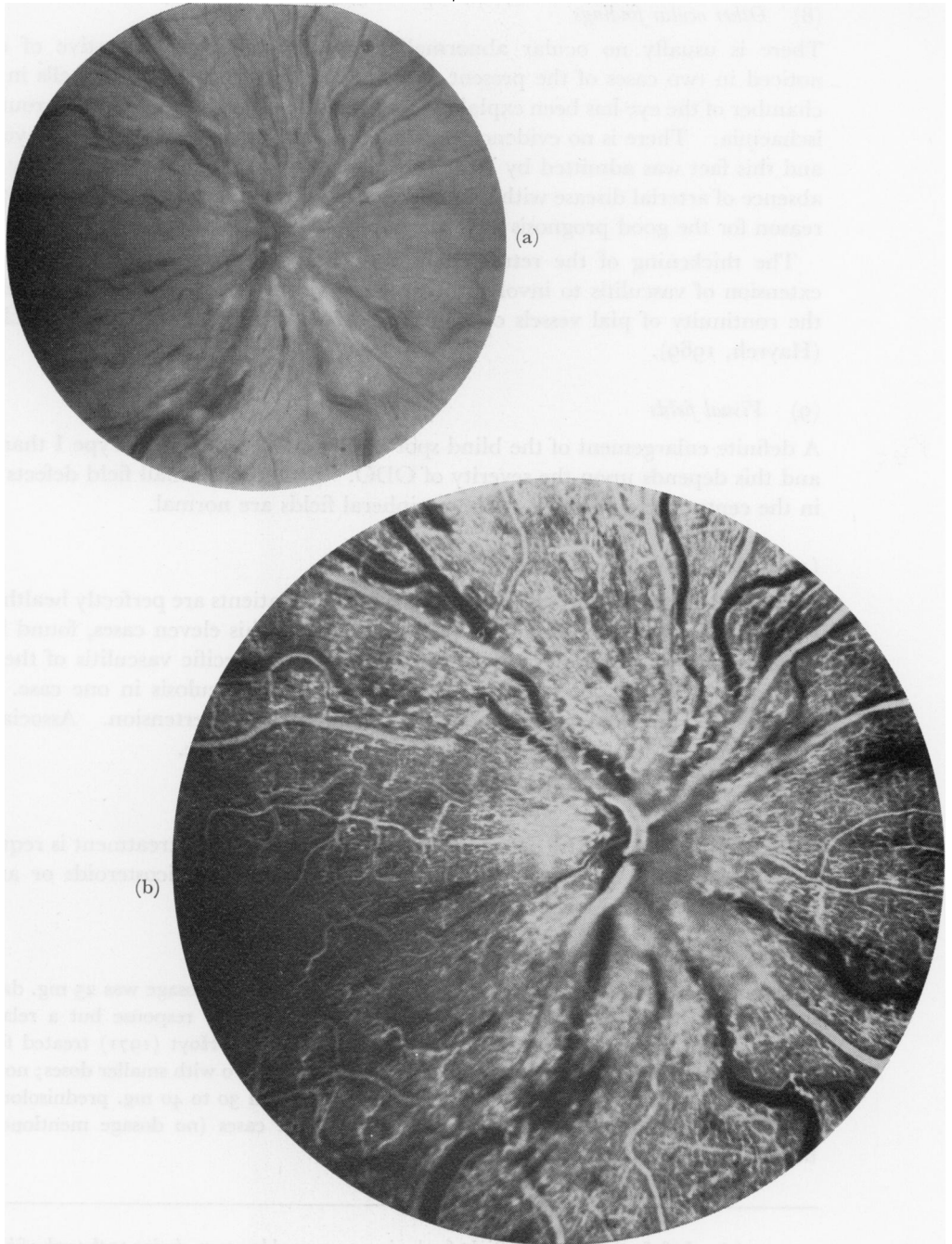
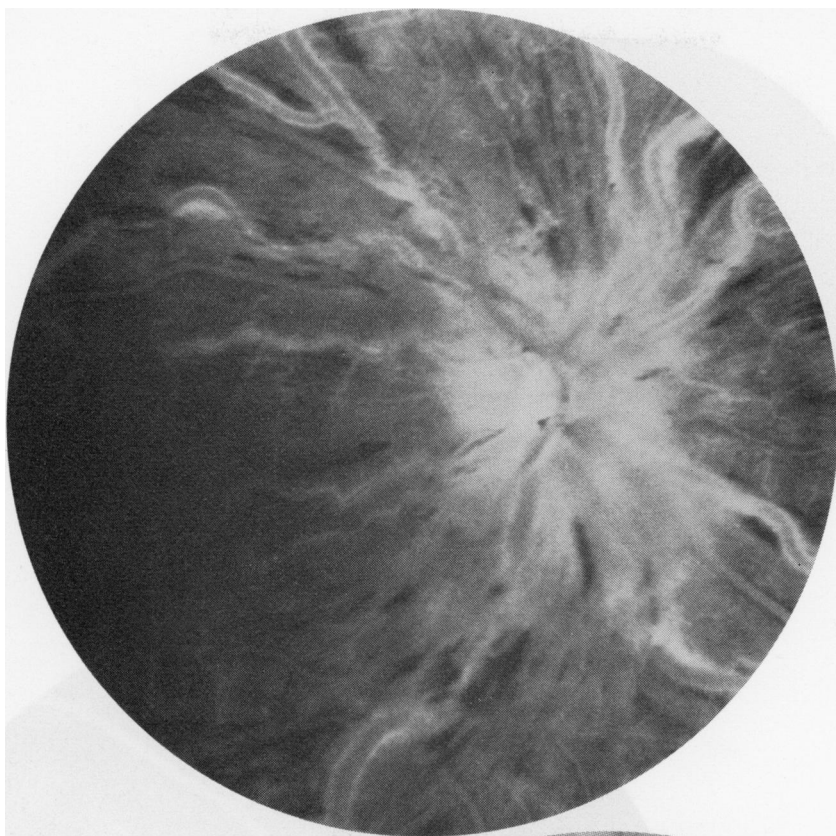
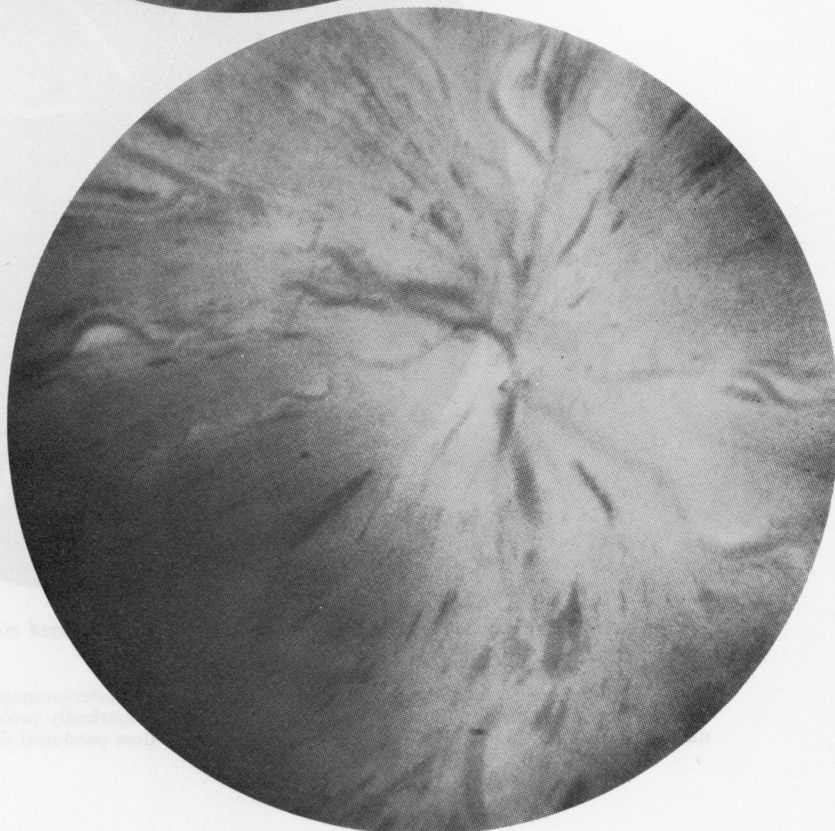


FIG. 4 (a) *Right fundus of a 34-year-old woman complaining of blurred vision and flickering light every morning for about an hour for one week. Visual acuity 6/6*

(b, c, d) *Fluorescence angiograms of (a) during late arterial (b), post-venous (c), and late (d) phases. Retinal venous circulation time in right eye 39 sec. as compared to 11 sec. in left eye. One week after (a) the visual acuity dropped to 6/18 which improved to 6/9 in 3 days with prednisolone 40 mg. daily. After this a reduction of steroids produced deterioration and an increase in dose an improvement*



(4c)



(4d)

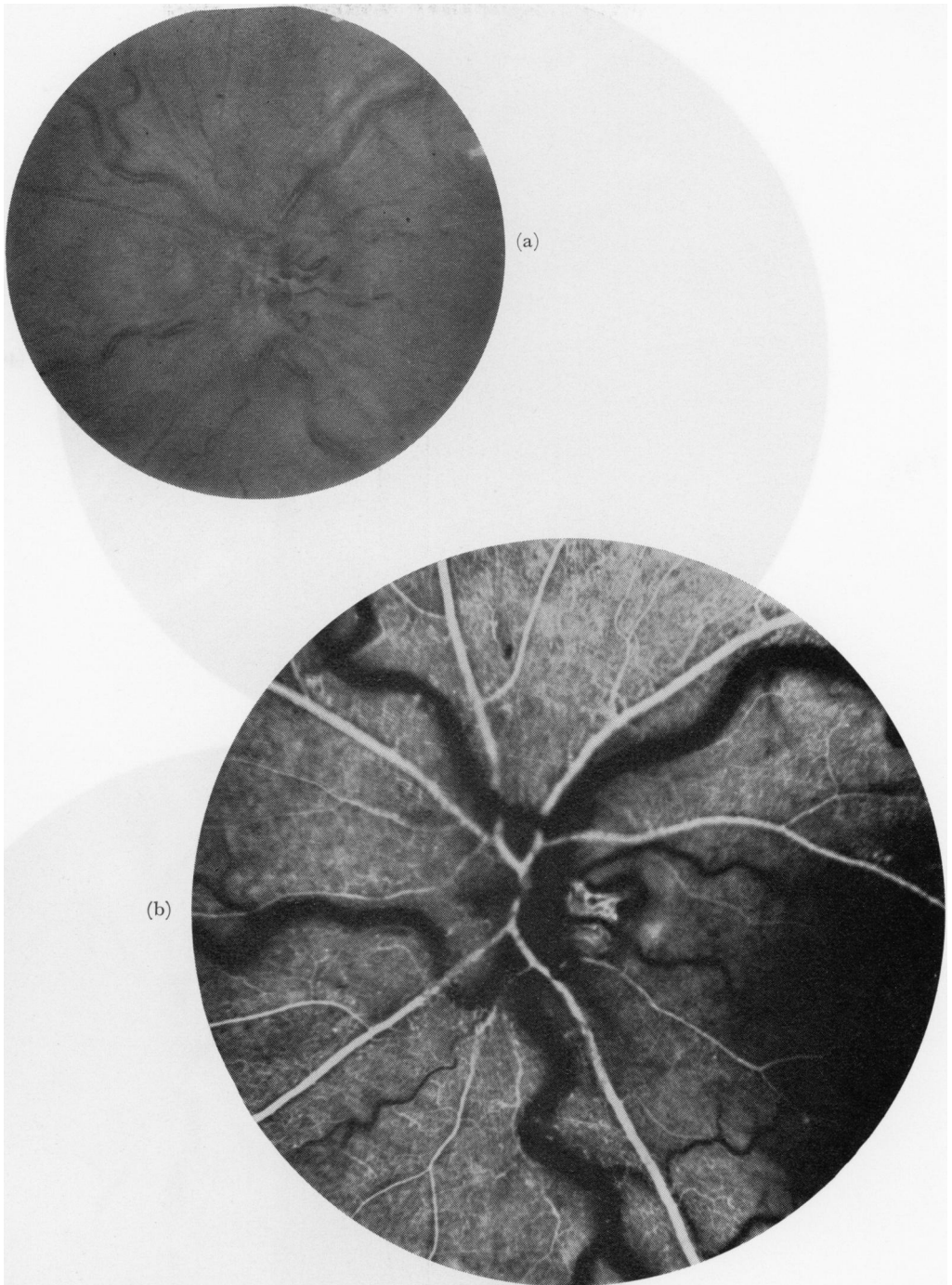
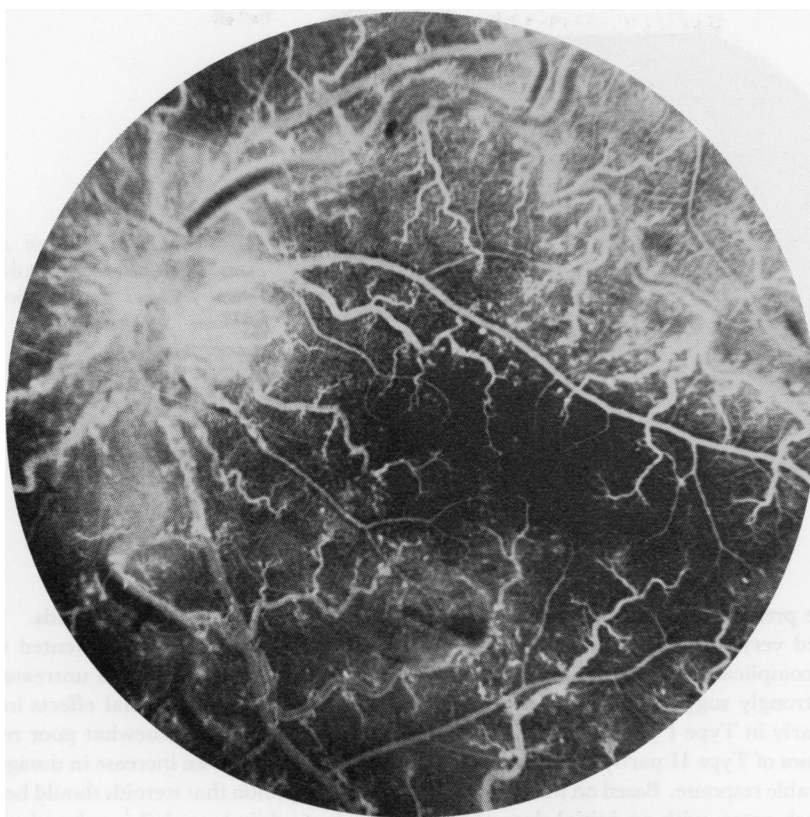
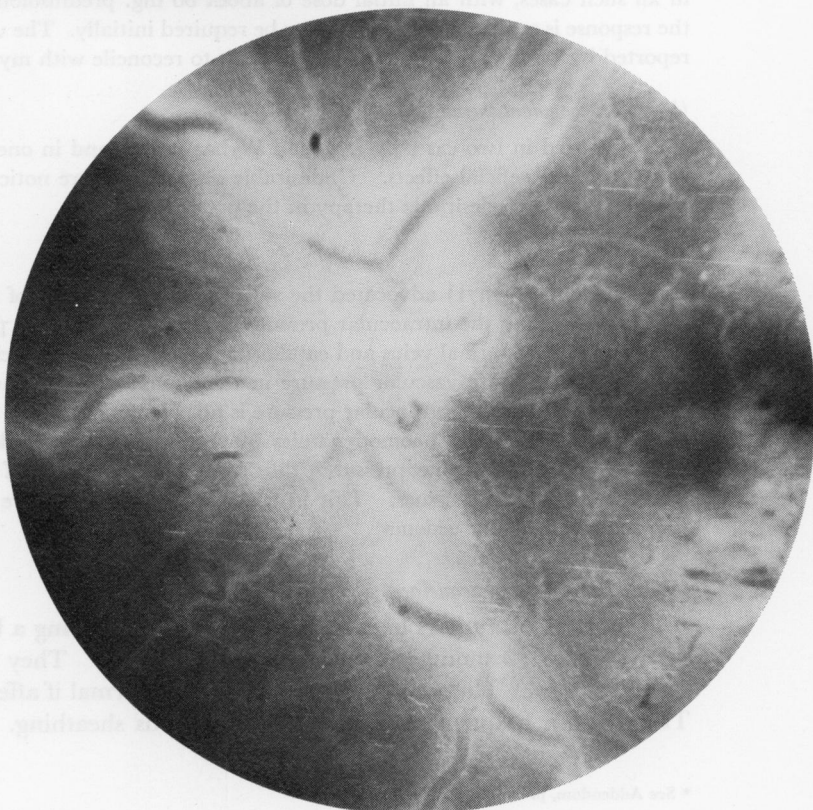


FIG. 5 (a) *Left fundus of a 26½-year-old man, complaining of blurred vision in left eye for 1 week, visual acuity 6/6*

(b, c, d) *Fluorescence angiograms of (a) during arterial (b), arteriovenous (c), and late (d) phases. Note retino-ciliary veins on optic disc. Venous circulation time markedly prolonged. On prednisolone 40 mg. daily visual acuity and fundus improved but reduction in dose produced deterioration*



(5c)



(5d)

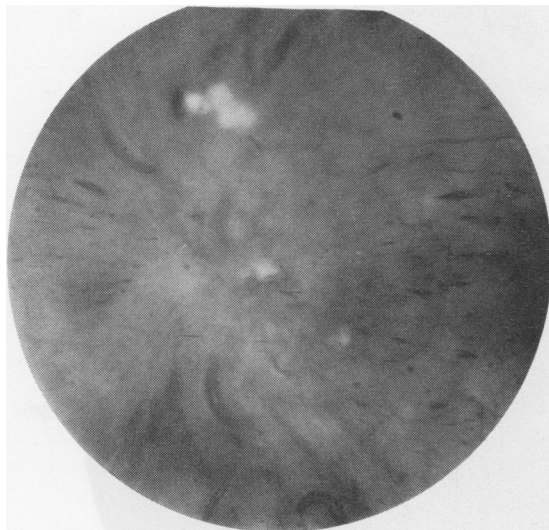


FIG. 6 *Left fundus in a 44-year-old man, 5 months after onset of defective vision. Visual acuity 1/60 with central scotoma. Note macular oedema and haemorrhages*

In the present series, six out of the eight cases were treated with systemic steroids. This not only shortened very significantly the course of the disease but also apparently prevented the possibility of any complications and perivenous sheathing, as compared with the two untreated cases. My results strongly suggest that adequate steroid therapy has definite beneficial effects in such cases*, particularly in Type I (compare Fig. 3*a, b* and 3*e, f*). I attribute a somewhat poor response in two of my cases of Type II partly to inadequate steroid therapy because an increase in dosage did produce a favourable response. Based on these findings, I am of the opinion that steroids should be administered in all such cases, with an initial dose of about 80 mg. prednisolone daily, reduced gradually once the response is seen. A higher dose may be required initially. The unsatisfactory response to steroids reported by Lonn and Hoyt (1966) is difficult to reconcile with my findings.

(b) *Anti-coagulant therapy*

This was tried in two cases by Lyle and Wybar (1961) and in one case by Lonn and Hoyt (1966) without any beneficial effects. Undesirable side-effects were noticed in some cases. This was not considered to be a desirable therapy in the present series.

(c) *Diamox*

Hart and others (1971) advocated the systemic administration of Diamox in these cases, with the idea that lowering the intraocular pressure could improve ocular perfusion. I feel that, if the rise in pressure in the retinal veins and capillaries is secondary to a block in the CRV behind the lamina cribrosa and the intravascular pressure in the retinal vessels is much higher than the intraocular pressure, lowering the intraocular pressure is not likely to improve the perfusion. In fact, it would further upset the tissue haemodynamics by creating a more marked disparity between the intravascular and extravascular pressures, thus favouring an outflow of fluid from the retinal capillaries into the surrounding tissues. This in turn would produce more oedema of the OD and retina (particularly macular oedema).

(12) *Progress and sequelae*

All the previous authors have described a disease running a benign but protracted course, taking up to 18 months or even longer to resolve. They observed no serious residual complications. The visual acuity returned to normal if affected during the active phase. There was a frequent occurrence of perivenous sheathing. Pigmentary changes at the

* See Addendum, p. 670

macula were seen by Hart and others (1971). In the present study, the progress depended upon the treatment. In Type I adequately treated cases showed a response within a week and resolved within 3 to 6 weeks, while an untreated case took 8 months. No residual sequelae were seen in the treated cases while the untreated case had some pallor of the OD. In Type II cases, my experience suggests that steroid therapy reduces the length of the disease and also the complications, because one untreated case lost his central vision permanently because of macular oedema and developed perivenous sheathing. Type II cases tend to develop retino-ciliary vessels on the OD; no such vessels were seen in Type I.

(13) *Recurrences*

None have been reported by previous authors (Lyle and Wybar followed two cases for 5 to 6 years; no other author mentioned the duration of follow-up) and my experience in following some of these cases for about 3 years has been similar. However, Munro (1971) has brought to my attention three cases (with most probably Type II ODV) seen by him in whom the condition recurred; a 25-year-old woman and a 43-year-old man had a recurrence in the same eye after 4½ and 10 years respectively, and his third case, a man aged 27, had a recurrence in the second eye 2½ years after the first attack.

DIFFERENTIAL DIAGNOSIS

Most of these cases are misdiagnosed partly because they so closely resemble the following conditions, and partly because ODV has not been defined as a clinical entity:

(i) *ODO due to intracranial hypertension*

The fundus appearance in ODV Type I is almost identical with that in intracranial hypertension. Where swelling of the OD is unilateral, the possibility of ODV Type I should always be kept in mind. Other features of intracranial hypertension and space-occupying lesions should help in differentiation.

(ii) *Haemorrhagic retinopathy (Central retinal vascular occlusion)*

This has erroneously been considered to be due to thrombosis of the CRV (Hayreh, 1971). There is haemorrhagic retinopathy and marked disturbance of vision.

(iii) *Venous stasis retinopathy (Pure CRV occlusion) (Hayreh, 1971)*

ODV Type II represents a variety of pure CRV occlusion in which the occlusion is secondary to retinal phlebitis and not to arteriosclerosis.

(iv) *Eales's disease*

This is typically bilateral and progressive, and involves initially the peripheral retinal veins with venous sheathing, vitreous haemorrhages, and retinal neovascularization; recurrences are the rule.

(v) *Hypertensive retinopathy*

This has a distinctive fundus appearance and is invariably accompanied by high arterial blood pressure.

(vi) *Chronic cyclitis with ocular hypotony*

In this there may be slight impairment of visual acuity or none, and fundus examination may show ODO, engorged retinal veins, and sometimes even retinal haemorrhages. Evidence of cyclitis on a detailed slit-lamp examination and ocular hypotony should differentiate this condition. It responds satisfactorily to systemic steroids.

(vii) Optic neuritis

The presence of marked ODO involving the entire disc, together with retinal venous engorgement, no field defect, no pupillary abnormality, and no other sign suggestive of retrobulbar neuritis would exclude optic neuritis.

(viii) Unilateral oedema of the OD

This may be seen in intracranial hypertension when there is unilateral optic atrophy, high myopia, or obliteration of the sheath of the optic nerve, etc. It may also be due to orbital or ocular causes.

PATHOGENESIS

Lyle and Wybar (1961) considered this to be a variety of Eales's disease because in one of their cases they found simultaneously "retinal vasculitis" in one eye and a peripheral type of Eales's disease in the other.

Lonn and Hoyt (1966) regarded it as the result of total CRV occlusion, probably initiated by a phlebitis of retinal veins in and about the optic nerve head. According to them, ODO in this condition is a consequence of venous occlusion; I do not agree with their latter view (Hayreh, 1965, 1968, 1969).

Cogan (1968) concluded from his histopathological studies of severe forms of what he called "retinal and papillary vasculitis" that mild inflammatory processes affected the retinal veins preferentially, the retinal arteries being involved only in severe cases. He had no case of a mild variety (under consideration in this paper) available for histopathology and considered these to be usually idiopathic.

I feel that the beneficial effect of steroid therapy in these cases indicates that we are dealing with a non-specific inflammation. My previous observations have indicated that, in the various forms of ODO, the accumulation of fluid is in the loose prelaminar region of the OD (Hayreh, 1969); this region is supplied by the posterior ciliary vessels (Hayreh, 1969).

Type I The ODO would result from mild non-specific vasculitis of the ciliary vessels in the prelaminar region. I postulate the chain of events in ODV Type I to be "mild non-specific vasculitis of the ciliary vessels in the loose prelaminar region of the OD → increased capillary permeability → accumulation of fluid in loose prelaminar tissue → ODO → compression of the venous channels in the prelaminar region → more ODO".

The ODO would compress the CRV in the optic nerve head and produce secondary retinal venous dilatation and stasis. Primary CRV block cannot produce such pronounced ODO (Hayreh, 1968, 1969, 1971).

Type II The clinical picture is that of pure CRV occlusion (Hayreh, 1971), associated with ODO in young individuals without vascular sclerosis. The primary cause in all probability is phlebitis of the CRV in the region of the optic nerve head or retrolaminar region. This leads to localized thrombosis of the vein and results in a clinical picture of pure CRV occlusion not associated with an arterial ischaemia (Hayreh, 1971). The communication between the CRV and choroidal veins in the prelaminar region of the OD helps to drain away the blood from the retinal veins to the choroidal circulation. That these collaterals on compensatory enlargement form retino-ciliary vessels in these eyes supports this view. If the thrombotic process in the CRV extends to involve and block the above-mentioned collaterals as well, it would result in haemorrhagic retinopathy—

a phenomenon similar to that demonstrated experimentally by Fujino, Curtin, and Norton (1969). Such cases would start with a clinical picture of pure CRV occlusion (*i.e.* evidence of CRV occlusion with normal visual acuity) and end with central retinal vascular occlusion (*i.e.* haemorrhagic retinopathy with poor visual acuity) (Hayreh, 1971); the third case of Munro (1971) showed this. The associated ODO in these cases is most probably due to a co-existing vasculitis of the OD. Marked perivenous fluorescence of the main retinal veins, seen during the late phases of fluorescence angiography, indicates an extension of a mild degree of phlebitis along these veins. Steroids in these cases would thus help to control the phlebitis of the CRV and retinal veins as well as the vasculitis of the fine disc vessels. This in turn would not only prevent venous thrombosis and its extension into the proximal part of the CRV but also help in keeping open its tributaries in the prelaminar region of the OD and hence aid in establishing the retino-ciliary circulation. Once thrombosis of the CRV has occluded the vein, the response to steroids may be poor. Retino-ciliary collateral circulation helps to maintain some circulation till the thrombus recanalizes and re-establishes the normal venous circulation.

ODV, like retinal vasculitis and most cases of uveitis, seems to represent a non-specific endogenous vasculitis. This may be due to an allergic reaction involving a wide variety of antigens. The vessels may become sensitized through an intraocular antigenic source (*e.g.* lens proteins or other ocular tissue), or from an extraocular source (*e.g.* bacterial, viral, etc.), or possibly from the formation of complex autoantibodies (Ashton, 1962).

The present study shows that the division of ODV into Type I and Type II is justified on the basis of clinical and fluorescence fundus angiography findings, therapeutic response, and pathogenesis.

The clinical picture, the response to steroid therapy, and the duration of the disease in these cases would thus depend upon:

- (a) The severity of the ODV;
- (b) The degree of the venous occlusion;
- (c) The amount of collateral circulation established to overcome the obstruction in the CRV;
- (d) A difference in pathogenesis between Type I and Type II. This could explain the very good response by Type I cases to steroid therapy and the not so good response by Type II cases.

Summary

Eight cases of optic disc vasculitis are described. A clinical description of the disease based on these and on 32 cases reported so far in the literature is given in detail. The condition is seen in young adults of either sex, usually complaining of unilateral vague blurring of vision, with almost a normal visual acuity. The fundus may show either marked oedema of the optic disc (Type I) or signs of central retinal vein obstruction (Type II). No other ocular or systemic abnormality is detected. Adequate systemic steroid therapy has a beneficial effect although the disease is usually self-limiting. The differential diagnosis and the pathogenesis of optic disc vasculitis are discussed.

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Addendum

Since the submission of this paper for publication, I have seen more cases of ODV, of which the following two Type II cases are particularly instructive in demonstrating the efficacy of systemic steroids.

Case 1 A 64-year-old woman was seen with a 3-week history of attacks of foggy vision in the left eye for 2 to 3 hours on waking every morning. The visual acuity was 6/6-part and N₅ in the left eye, with fundus and angiographic findings of ODV Type II. After about 3 months without treatment, the visual acuity started to deteriorate; it was 6/60 5 months after the onset and unaltered for the following 2 months. At the end of this period she showed macular oedema and retino-ciliary veins, in addition to the original fundus changes. She was started on prednisolone 40 mg. daily; after 3 days the visual acuity improved from 6/60 and N₂₄ to 6/18+ and N₁₂. She ran out of tablets after 4 days and was seen 3 days later when the acuity had fallen to 6/60 again. After 5 days on a resumed dose of prednisolone 40 mg. daily, the acuity was 6/6-part and N₅, with some improvement of the optic disc and macular oedema. The subjective improvement was dramatic but the fundus is improving only slowly.

Case 2 A 61-year-old man was seen with a 7-week history of hazy vision and metamorphosia in the right eye. The visual acuity was 6/9+ in that eye with the fundus and angiographic appearance of ODV Type II, and some macular oedema. He was put on prednisolone 30 mg. daily by the ophthalmologist concerned, and the dose was gradually reduced after 4 days to 20 mg. daily in a fortnight, when the acuity was 6/5-part with very slow improvement in the optic disc and macular oedema. The dosage of prednisolone was reduced to 10 mg. daily over the following weeks, and the acuity deteriorated to 6/9-part. When he started on 40 mg. daily again, he noticed a dramatic improvement within 2 to 3 days and after 7 days the acuity was 6/4, but with no significant improvement in the fundus picture. At present, a normal visual acuity with a very slow improvement in the fundus is being maintained on a dose of about 20 mg. daily.