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Communications

Benign retinal vasculitis

Clinical and fluorescein angiographic study

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Central retinal vein occlusion occurs predominantly in the older age groups, and an increased incidence has been reported in association with systemic vascular disease (Paton, Rubinstein, and Smith, 1964), chronic simple glaucoma (Smith, 1955), high viscosity syndromes (Rowlands and Vaizey, 1938), and malignant disease (Ellis, Hamer, Hunt, Lever, Lever, Peart, and Walker, 1964). Central retinal vein thrombosis under the age of 40 years is infrequent (Vannas and Raitta, 1966), and both clinical (Lyle and Wybar, 1961) and pathological studies (Ballantyne and Michaelson, 1947) have suggested an inflammatory aetiology. The appearances in both groups are similar, but the visual loss and retinal changes are less severe in younger patients.

The first description (Lyle and Wybar, 1961), comprised six patients whose ages ranged from 14 to 49 years, who had minimal visual symptoms in spite of a florid retinopathy. The process was unilateral in five cases, and after resolution of the retinopathy mild sheathing of the veins and increased vascularity at the disc was seen in three cases. The sixth patient had cells in the anterior chamber in both eyes with a central vein occlusion on one side and a peripheral haemorrhagic retinopathy on the other. Lyle and Wybar postulated that the condition was mainly inflammatory, since the peripheral changes were similar to those of Eales's disease, and suggested the term retinal vasculitis. A further report of five cases (Lonn and Hoyt, 1966) substantiated the benign unilateral course of the condition and stressed the absence of inflammatory signs. Cogan (1969) divided his series into three groups according to the degree of severity and contributed both pathological and aetiological evidence on several cases. This paper presents a clinical and fluorescein angiographic study of nine cases of the syndrome of unilateral central retinal vasculitis occurring in patients under the age of 40 years. Patients with peripheral involvement and vitreous haemorrhages (Eales's disease) were excluded.

Case reports

CASE 1, A 21-YEAR-OLD WOMAN

This patient complained in February, 1967, of black spots in front of the right eye and blurring of vision for 3 days. She gave a history of intermittent frontal headaches for 9 months and was admitted to the National Hospital with a diagnosis of unilateral disc oedema.

Examination

The visual acuity was 6/24 in the right eye, and 6/6 in the left. The ocular media were clear and the left fundus normal. The right fundus had disc oedema with grossly dilated veins and scattered

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retinal haemorrhages extending to the equator. Perimetry revealed enlargement of the blind spot and a small central scotoma.

X ray examination of the skull and optic foramina was normal. The blood count and erythrocyte sedimentation rate were normal, and serological tests for syphilis gave negative results.

Fluorescein angiography 4 days after the onset of symptoms showed normal arterial filling but some delay in venous filling. Residual studies showed leakage from the region of the disc and in relation to the larger retinal veins.

Diagnosis

Retinal vasculitis.

Subsequent course

The patient was treated with 40 mg. prednisolone daily and this was reduced over a 2-month period without significant improvement. The disc oedema and retinal haemorrhages resolved over the next 18 months, so that when she was seen in November, 1969, the visual acuity was 6/5 and the fundus appeared to be normal apart from minimal sheathing of the larger retinal veins. Spontaneous venous pulsation was present in both eyes.

Fluorescein angiography showed a normal dye transit with no leakage in the residual photographs.

Summary

A healthy young woman had mild visual loss from retinal vasculitis. Complete resolution occurred over an 18 month period leaving residua of sheathing of the retinal veins.

CASE 2, A 35-YEAR-OLD MAN

This patient complained of black spots and blurring of vision in the right eye in November, 1969. These symptoms lasted for 3 days and then subsided, but 3 days later the right vision again became blurred. A diagnosis of unilateral disc oedema was made and the patient was admitted to the National Hospital for investigation in January, 1968. He had no significant past history.

Examination

The visual acuity was 6/9 in the right eye and 6/6 in the left. The ocular media were clear and the left fundus was normal. In the right eye there was disc oedema with dilatation of the veins and numerous haemorrhages and cotton wool spots. Perimetry showed enlargement of the blind spot.

X ray examination of the skull and optic foramina was normal. Serological tests for syphilis gave negative results.

Fluorescein angiography 2 months after the onset of symptoms showed normal dye transit and residual studies demonstrated leakage around the disc and larger veins.

Diagnosis

Retinal vasculitis.

Subsequent course

No treatment was given. The visual acuity returned to 6/6 within 6 months and this was associated with complete resolution of the abnormal fundus appearances. Repeat fluorescein studies showed normal transit without any leakage in the residual studies.

Summary

A healthy young man developed retinal vasculitis with mild visual loss and complete resolution occurred over a 6-month period.

CASE 3 A 31-YEAR-OLD MAN

This patient complained of blurring of vision and blank areas in his visual field for 3 days when first seen in October, 1969. There were no relevant past illnesses.

Examination

The visual acuity was 6/9 in the right eye and 6/6 in the left. The ocular media were clear and the left fundus normal. The right fundus showed disc oedema, the retinal veins were grossly engorged, and there were cotton wool spots and numerous haemorrhages. The retinal periphery was normal. Spontaneous venous pulsation was present in the left eye but not in the right.

Perimetry showed enlargement of the blind spot and two small paracentral scotomas.

X rays of the skull and optic foramina were normal. The erythrocyte sedimentation rate and blood count were normal. Serological tests for syphilis gave negative results.

Ophthalmodynamometric studies showed arterial pressures of 110/65 in the right eye and 110/60 in the left.

Fluorescein angiography 5 days after the onset of symptoms showed a rapid arterial transit with slow filling of the veins. Residual studies showed massive leakage of dye in relation to the disc and larger veins.

Diagnosis

Retinal vasculitis.

Subsequent course

The patient was treated with 30 mg. Prednisolone daily. This therapy was stopped after 6 weeks as no significant improvement had occurred. The abnormal fundus appearances resolved over a 2-month period, leaving mild pigmentary abnormalities in the macular region. The visual acuity improved to 6/6 though the patient complained of micropsia. Spontaneous venous pulsation was now present in both eyes. The fluorescein angiogram was repeated and a normal transit was recorded with minimal leakage in the region of the disc.

Summary

A healthy young man developed retinal vasculitis with mild visual impairment. Complete resolution occurred within 2 months, though the symptoms of micropsia persisted and some pigmentary changes were noted at the macula.

CASE 4 A 31-YEAR-OLD MAN

This patient woke up one morning with blurring of vision and a shimmering sensation in the temporal half of the left visual field in December, 1969. He also complained of mild frontal headaches.

Examination

10 days later the visual acuity was 6/6 in each eye. The ocular media were clear and the right fundus normal. The left fundus showed disc oedema, dilated tortuous veins, cotton wool spots, and haemorrhages extending to the equator. Ophthalmodynamometry showed arterial pressures of 105/60 and 110/65 in the right and left eyes respectively. Venous pulsation was present spontaneously in the left eye and was elicited at a pressure of 50 mm.Hg in the right. Perimetry demonstrated enlargement of the blind spot.

X ray examination of the skull and optic foramina was normal. No haematological abnormality was detected. Serological tests for syphilis gave negative results.

Fluorescein studies 14 days after the onset of symptoms showed rapid passage of dye through the retinal vessels but with profuse leakage in the later stages from the major veins and in relation to the disc (Fig. 1*a*, overleaf).

Diagnosis

Retinal vasculitis.

Subsequent course

3 weeks after the first onset of symptoms the vision in the left eye deteriorated to 6/36. The disc swelling had increased, the veins were more dilated and tortuous, and the number of haemorrhages and exudates had increased. Fluorescein studies showed a slow transit, dilatation of the arterioles, venules, and capillaries, and marked leakage of dye in all areas.

The patient was admitted for investigation in March, 1970, when the visual acuity remained at 6/36. The gross retinal changes had partly resolved and fluorescein studies showed a normal retinal transit. The capillaries were dilated with microaneurysm formation and extensive leakage of dye (Fig. 1*b*). The macular region showed the appearances of cystoid maculopathy.

Further resolution had occurred when he was seen in May, 1970, and the vision had improved to 6/6, though he still complained of micropsia. There were fine scattered retinal haemorrhages but no exudates. Fluorescein studies showed a normal flow pattern; the paramacular capillaries were dilated but no leakage was observed in the late photographs. Complete resolution had occurred when this patient was last examined in February, 1971.

Summary

A healthy 31-year-old man developed sudden mild visual impairment and retinal vasculitis was diagnosed. Severe visual impairment developed after 3 weeks and was associated with increased capillary permeability in the macular region. Spontaneous resolution occurred over a 6-month-period.

CASE 5 A 21-YEAR-OLD MAN

This patient first became aware of bright spots in the temporal field of the right eye in December, 1968, and was referred for fluorescein studies by Mr. Geoffrey Davies of King's College Hospital in February, 1969. He had suffered no relevant previous illnesses.

Examination

The visual acuity was 6/9 in the right eye and 6/5 in the left. The ocular media were clear and the left fundus normal. The right fundus had disc oedema, the retinal veins were grossly dilated, and there were numerous cotton wool spots and haemorrhages extending to the periphery (Fig. 2*a*, overleaf). The retinal periphery appeared normal. Spontaneous venous pulsation was present in the left eye but absent in the right.

Perimetry showed enlargement of the blind spot.

X ray examination of the skull and optic foramina was normal. No haematological abnormality was detected. Serological tests for syphilis gave negative results.

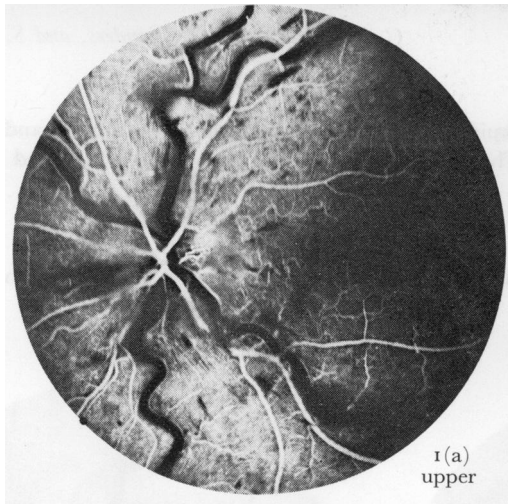
Fluorescein angiography 2 months after the onset of symptoms revealed a relatively normal flow through the arteries and veins, but leakage of dye was seen in the residual studies in the region of the disc and in relation to the larger veins.

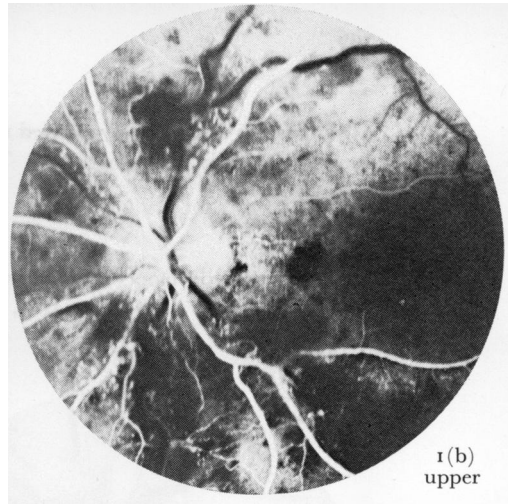
Diagnosis

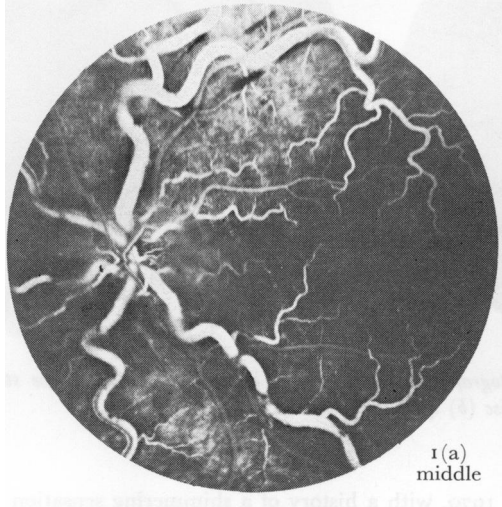
Retinal vasculitis.

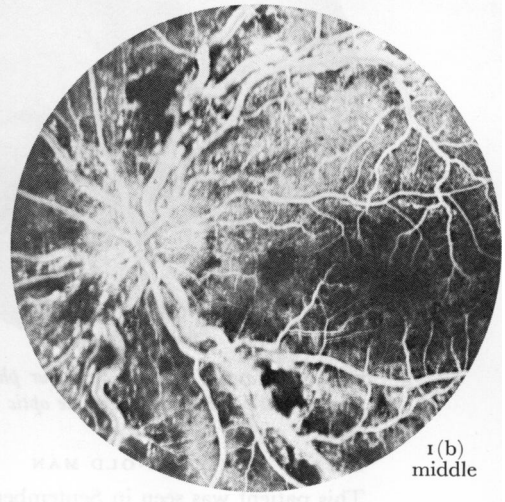
Subsequent courase

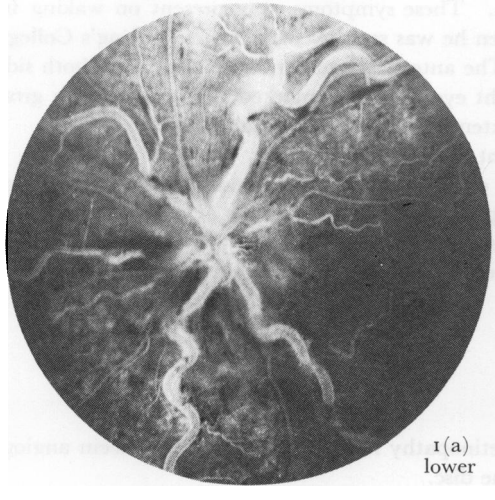
The patient was seen again in January, 1970, when the vision in the right eye had improved to 6/6. The haemorrhagic retinopathy had resolved, though the veins, which were of normal calibre, showed mild sheathing. Numerous shunt vessels were nowapparent in relation to the disc (Fig. 2*b*, overleaf). Spontaneous venous pulsation was present in both eyes.











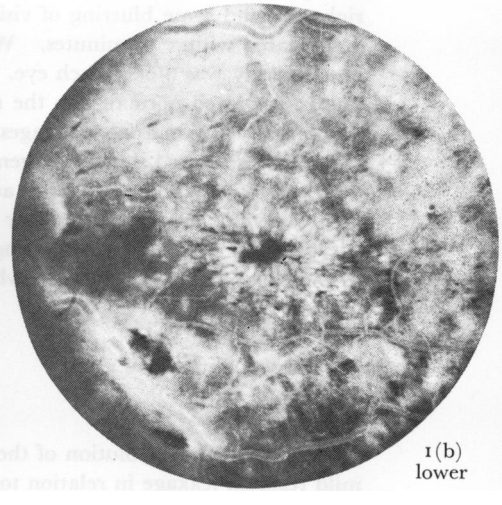


FIG. 1(*a*) *Case* 4. *Fluorescein angiograms in the arterio-venous* (*upper*), *venous* (*middle*), *and residual phases* (*lower*), *to show the stage of venous decompensation. The veins are grossly dilated and there is perivenous leakage of dye. Capillary flow is intact and the visual acuity at this stage was* 6/6

FIG. 1(*b*) *Case* 4, 3 *months later. The fluorescein angiograms are comparable and show the stage of capillary decompensation. The veins remain dilated but gross capillary dilatation is seen with microaneurysm formation. In the residual phase extensive capillary leakage has produced the appearances of cystoid maculopathy. Visual acuity was* 6/36

Fluorescein angiography showed rapid transit of dye through the retinal vessels and no leakage of dye in the residual photographs. The shunt vessels in the region of the disc filled in the venous phase.

Summary

A young man developed retinal vasculitis, and complete resolution occurred within 12 months with shunt vessels at the optic disc.

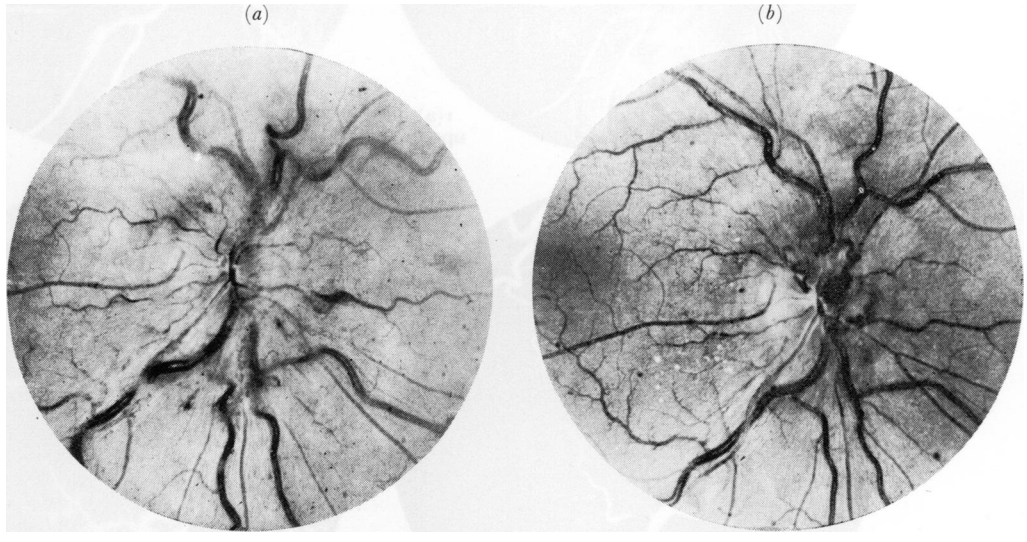


FIG. 2 *Case 5. Copies of colour photographs to show the disc appearances in the active stage (a) and the development of shunt vessels at the optic disc (b) after resolution*

CASE 6 A 20-YEAR-OLD MAN

This patient was seen in September, 1970, with a history of a shimmering sensation in front of the right eye and some blurring of vision. These symptoms were present on waking in the morning and cleared within 30 minutes. When he was seen by Mr. Davies of King's College Hospital the visual acuity was 6/6 in each eye. The anterior chambers were normal on both sides and the left fundus appeared normal. In the right eye there was marked disc oedema with gross dilatation of the retinal veins and haemorrhages extending to the equatorial region.

Perimetry showed slight enlargement of the blind spot.

X ray examination of the skull and optic foramina was normal. Routine haematological studies were normal. Serological tests for syphilis gave negative results.

Ophthalmodynamometry readings showed pressures of 120/65 in each eye. The venous pressure in the right eye was 60 mm.Hg whereas in the left eye there was spontaneous venous pulsation.

Diagnosis

Retinal vasculitis.

Subsequent course

In October, 1970, resolution of the retinopathy had occurred and fluorescein angiography showed mild residual leakage in relation to the disc.

Summary

A young man was found to have retinal vasculitis; there was complete resolution in 6 weeks without any treatment and there were no residual signs.

CASE 7 A 24-YEAR-OLD MAN

This patient was referred by Mr. J. N. Ormrod of Maidstone with a history of blurring of vision in the right eye in November, 1970; 2 weeks later the visual acuity in the right eye deteriorated to 6/9, and a week later to 6/18.

Examination

The visual acuity was 6/24 in the right eye and 6/5 in the left. Several discrete areas of blurring were noted with the Amsler chart and the visual fields showed enlargement of the blind spot and a relative central scotoma. The fundus showed marked disc swelling with gross dilatation of the veins, numerous cotton wool spots, and haemorrhages extending to the periphery. The left fundus was normal. Ophthalmodynamometry showed diastolic arterial pressures of 60 mm. Hg in each eye. The venous pressure was 10 mm. Hg in the left eye and 55 mm. Hg in the right.

X ray examination of the skull and optic foramina was normal. Serological tests for syphilis gave negative results. The only haematological abnormality was elevation of the alpha 2 globulin.

Fluorescein angiography showed normal arterial filling but with a slow transit of dye to the venous side and with some dilatation of the peripapillary capillary plexus in the lower half of the disc, and of the capillaries in the macular region. Residual photographs after 10 minutes showed marked leakage of dye into the retina, but particularly around the disc and along the larger veins.

Diagnosis

Retinal vasculitis.

Subsequent course

The patient was treated with 40 mg. prednisolone daily and later with ACTH.

As there was no improvement Diamox was also administered in an attempt to improve ocular perfusion.

Summary

A young man developed retinal vasculitis and the condition resolved after 2 months.

CASE 8 A 33-YEAR-OLD MAN

This patient complained in January, 1970, of sudden severe visual loss in the right eye on waking. He also gave a history of headaches for 6 months.

Examination

The visual acuity was counting fingers in the right eye and 6/6 in the left. The media were clear, The left fundus was normal. The right fundus showed gross disc oedema with peripapillary cotton wool spots, and the engorged veins were surrounded by haemorrhages extending to the periphery.

Ophthalmodynamometry showed diastolic readings of 88 in the right eye and 90 in the left eye and, whereas there was spontaneous venous pulsation in the left eye, the venous pressure in the right eye was 80 mm.Hg. The blind spot was enlarged and a central scotoma was present.

The blood pressure was 200/120 and an electrocardiogram showed evidence of mild left ventricular hypertrophy. Radiological and haematological investigations were otherwise normal. Serological tests for syphilis gave negative results.

Fluorescein angiography 8 days after the visual deterioration showed rapid filling of the arteries, though there was some degree of attenuation and variation in calibre of the vessels. The veins were dilated and filling was delayed. Residual studies showed marked leakage of fluorescein from the region of the disc and along the large veins.

Diagnosis

Retinal vasculitis.

Subsequent course

Studies were initiated to assess the systemic hypertension.

When the patient was seen again 9 months later the visual acuity had improved to 6/12 though he still complained of micropsia. There was still marked disc oedema with dilated veins and oedema in the macular region. There was also early disc oedema in the left eye with a few retinal haemorhages in the peripapillary region.

Fluorescein angiography was repeated and the arterial appearances were found to be unchanged. The capillary pattern in the posterior pole and perimacular region was abnormal. There were areas of capillary closure, and some capillaries were dilated to form preferential channels with microaneurysms (Fig. 3). Residual leakage of fluorescein was seen at the disc and along the major veins.

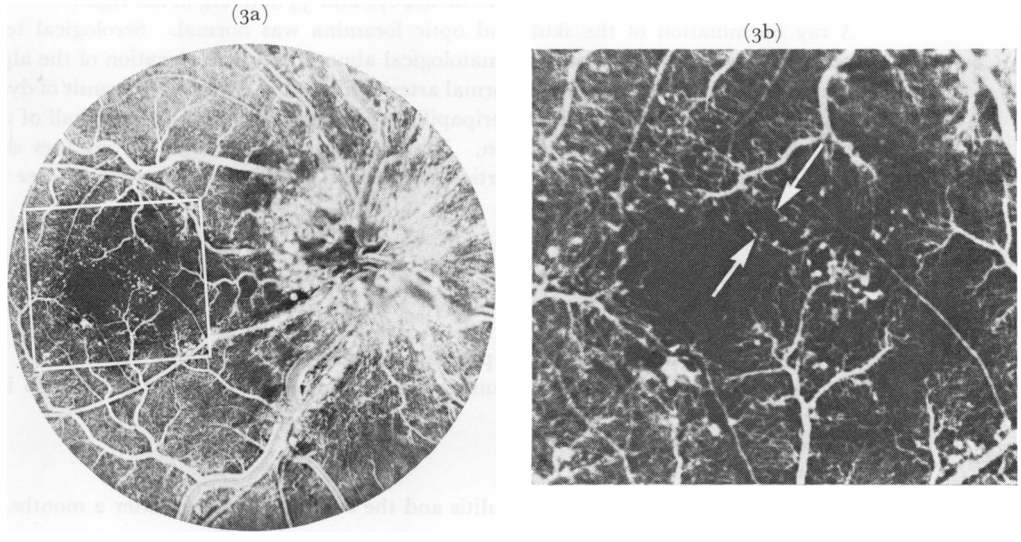


FIG. 3(*a*) *Case* 8. *Arterio-venous fluorescein angiogram* 8 *months after onset in this patient with hypertension. The arteries are attenuated, the capillaries show areas of closure with microaneurysm formation, and the veins are distended. Marked leakage of dye was visible in the residual photographs. The visual acuity was reduced to* 6/36.

FIG. 3(*b*) *Insert of Fig.* 3(*a*) *enlarged to show capillary dilatation with microaneurysm formation, areas of closure, and development of arterio venous-preferential channels* (*arrows*).

Summary

A young man presented sudden loss of vision in one eye and the clinical appearances of a retinal vasculitis. Systemic examination showed that he was suffering from malignant hypertension.

CASE 9 A 37-YEAR-OLD MAN

This patient referred by Mr. Peter Wright of King's College Hospital in January, 1971, with a week's history of sudden loss of temporal vision in the right eye. He complained of a dark shadow on the temporal side. There were no relevant previous illnesses though he had been kicked in the right eye some years previously.

Examination

The visual acuity was 6/9 in the right eye, and 6/6 in the left, with a small myopic correction. The blind spot on the right side was enlarged and objects were slightly dimmer on the temporal side.

Ophthalmodynamometry showed arterial pressures of 90/40 in each eye. The venous pressure was 40 mm.Hg in the right eye, whereas spontaneous venous pulsation was present in the left.

The left fundus was normal. The right disc was swollen, and the veins were grossly dilated with numerous haemorrhages and a few cotton wool spots in the posterior pole extending to the periphery.

X ray examination of the skull and optic foramina was normal. Haematological tests and serological tests for syphilis gave negative results.

Fluorescein angiograms showed normal arterial flow, with delayed venous filling and perivenous leakage in the late phases.

Diagnosis

Retinal vasculitis.

Subsequent course

The patient was seen again 6 weeks later, when the retinopathy had settled apart from a few resolving haemorrhages and small shunt vessels were visible at the optic disc. The visual acuity had improved to 6/5, Ishihara testing was normal, the visual field was normal, and venous pulsation was visible in the right eye.

Summary

A 37-year-old man experienced the sudden onset of retinal vasculitis. This resolved over a 6-week period and shunt vessels were visible at the optic disc.

Results

VISUAL SYMPTOMATOLOGY

Presenting visual complaints ranged from ill-defined blurring of vision to more specific complaints such as multiple small scotomata, black spots, and 'shimmering' sensations. These were often associated with good visual acuity and in one patient the symptoms were present on waking in the morning but subsided after 30 minutes. In three further patients the visual symptoms occurred suddenly. Severe visual loss, when present, was correlated with permeability defects in the capillaries in the macular region on fluorescein angiography. Micropsia was reported in three cases after resolution of the retinopathy.

OCULAR SIGNS

Ocular examination indicated more extensive involvement than the symptoms would suggest and consequently four cases were immediately referred to neurologists for further investigation of unilateral papilloedema. The similarity of the physical signs in our group, however, facilitated diagnosis, since all the patients had marked disc oedema, peripapillary cotton wool spots, and grossly dilated tortuous veins with perivenous haemorrhages extending to the periphery. Slit-lamp examination showed no activity in the anterior chamber in any case.

OPHTHALMODYNAMOMETRY

Readings were performed in six cases and the arterial pressures were equal in both eyes. Spontaneous venous pulsation was present in the non-affected eyes, but absent in the involved eyes, in all cases.

Measurements in five cases showed grossly elevated venous pressure to figures which were approaching the diastolic arterial pressure (Table I).

Table *Ophthalmodynamometry readings in five cases*

Case no.	*Vessels*	*Eye*	
		Normal	*Involved*
5	Arterial	105/60	110/60
	Venous	Spontaneous pulsation	50
6	Arterial	120/65	125/65
	Venous	Spontaneous pulsation	60
7	Arterial	90/60	90/65
	Venous	Spontaneous pulsation	55
8	Arterial	Diastolic 88	Diastolic 90
	Venous	Spontaneous pulsation	80
9	Arterial	90/40	90/40
	Venous	Spontaneous pulsation	38

COURSE

Complete resolution occurred in eight cases, and the duration before recovery ranged from 6 weeks to over a year. Residual signs included sheathing of the veins, macular pigmentary changes, and the development of shunt vessels around the disc in two cases. Resolution of the retinopathy was incomplete in one patient (Case 8) in whom a systemic disease was detected.

FLUORESCEIN RESULTS

Rapid fluorescein fundus photographs were performed, using the modified Zeiss camera, after the injection of 5 ml. 20 per cent fluorescein. Arterial filling was slow in the cases seen within the first few days of onset, but normal in all cases seen in the later stages. One case with systemic disease had irregularity of the arteries with impaired flow. Venous filling was delayed during the active phase, but in repeat studies, performed after resolution, the venous transit was normal. Abnormalities in the paramacular capillaries were noted in four patients whose visual acuity was diminished to 6/18 or less. The capillary changes consisted of dilatation, closure with microaneurysm formation, and the development of preferential shunt channels. These changes were associated with increased permeability to fluorescein. Residual photographs taken 10 minutes after injection showed a massive leakage of dye from the region of the disc and along the larger veins.

Discussion

AETIOLOGY OF CENTRAL RETINAL VASCULITIS

The appearances of retinal vasculitis and central retinal vein occlusion are similar, both clinically (Klien and Olwin, 1956) and on fluorescein angiography. Pathological studies of severely involved eyes with thrombotic glaucoma have shown evidence of arterial disease, primary venous disease (Klien and Olwin, 1956), and inflammatory disease (Ballantyne and Michaelson, 1937). However, no pathological studies have been made of the benign form of retinal vasculitis.

The evidence for an inflammatory aetiology is tenuous, as only one case in previous reports (Lyle and Wybar, 1968; Lonn and Hoyt 1966) and none in the present series had cells in the anterior chamber. Furthermore, it is well recognized that the finding of cells in the anterior chamber is frequent in ischaemic ocular disease (Knox, 1965; Sanders and Hoyt, 1970). Cellular cuffing of the retinal veins distal to the site of occlusion has

recently been described (Kohner, Dollery, Shakib, Henkind, Paterson, de Oliveira, and Bulpitt, 1970) after experimental branch vein thrombosis. Similarly, inflammatory cell sheathing has been observed in Eales's disease (Ashton, 1962) and in inflammatory central retinal vein thrombosis (Klien, 1960). In these cases the cuffing consisted mainly of round cells, whereas in the experimental vein occlusions the cells were predominantly polymorphonuclear leucocytes. The relatively short duration of the occlusion in the experimental group before histology may be of significance. The finding of haematological abnormalities in Eales's disease (Ashton, 1962) and the similar retinal findings in sickle cell haemoglobin disease may indicate a non-inflammatory aetiology. Central retinal vasculitis, although it affects predominantly young adult males, has no other feature in common with Eales's disease. The condidion is unilateral, peripheral retinal vessels are not involved. vitreous haemorrhages are absent, and resolution without recurrence is the rule.

Experimentally, the site of central retinal vein occlusion determines the severity of the resulting retinopathy, being limited to venous dilatation when the occlusion is outside the nerve (Hayreh, 1965) but producing a gross haemorrhagic retinopathy when at or close to the nerve head (Fujino, Curtin, and Norton, 1969). Occlusion of the central retinal vein distal to the anastomoses with the choroidal and pial circulation extensively reduced retinal perfusion so that fluorescein studies revealed defective arterial transit, whereas occlusions proximal to the collaterals produced minimal changes. Thus the site of occlusion may be important in determining the pattern and prognosis in central retinal vein occlusion and central retinal vasculitis. An anatomical variation in the emergence of the central retinal vein from the optic nerve sheath has been postulated in relation to central retinal vein occlusions in adults (Harms, 1905), and this has not been excluded in the mechanism of vasculitis.

PATHOPHYSIOLOGY

Controversy exists as to the role of arterial insufficiency in retinal vein thrombosis. It has been thought to be important on clinical (Paton, Rubinstein, and Smith, 1964), pathological (Klien and Olwin, 1956), and experimental evidence (Hayreh, 1965). Ophthalmodynamometry (OD) recordings in a series of 73 cases with central retinal vein occlusion showed reduction of the systolic pressure in 30 per cent. (Smith, 1964).

Arterial OD recordings in our series showed normal pressures which were equal in each eye. The venous pressure was elevated in all cases with absent spontaneous pulsation. Measurement by ophthalmodynamometry, when performed, showed marked elevation in all cases to levels approaching the diastolic arterial pressures. *Thus it appears that the single important factor in this group is a reduction in the arterio-venous pressure gradient (perfusion pressure) produced solely by elevation of the venous pressure.*

The absence of arterial disease with maintenance of a normal arterial pressure may be the main reason for the good prognosis in this group. This is supported by the fact that the one patient in this group with arterial disease due to malignant hypertension (Case 8) had severe visual loss with gross capillary decompensation. Similarly, in the severe cases described by Cogan (1969), there was a high association with systemic disease. The poorer prognosis in older patients is probably due to their impaired ability to form collateral channels and to the higher incidence of arterial disease. 86 per cent. were found to have associated arterial system disease (Paton and others, 1964). However, some patients in the older age group with no signs of arterial disease may have a good prognosis and may also develop collateral channels.

Fluorescein studies indicate that there are two main patterns of vascular decompensation:

(1) In the first group the most significant factor is venous decompensation. The veins which are dilated show perivenous fluorescence in the late photographs, and this is most marked in the concavity of the vein. The capillaries, though they may be somewhat dilated, are not permeable to fluorescein and there are no microaneurysms. The clinical appearance at this stage shows a moderate retinopathy with haemorrhages and cotton wool spots but with minimal retinal oedema. The visual acuity at this stage is usually only slightly reduced.

(2) The second more severe stage is that of capillary decompensation. This is characterized by dilated capillaries often with microaneurysms and extensive leakage of dye. The clinical appearances show an extensive retinopathy with retinal oedema and the visual acuity is impaired. The macular region may give the appearances of a cystic maculopathy and this has been described with central retinal vein occlusion by Gass (1968).

The capillary closure may be due in part to elevation of tissue pressure, either because of ischaemic cellular oedema of the retinal elements or because of failure of tissue fluid to be absorbed in the venous capillaries, since intraluminal pressure exceeds the plasma protein osmotic pressure (Kohner and others, 1970). The propensity of the retina for swelling and the sensitivity to ischaemia is particularly high in the macular region (Wise and Wangvivat, 1966).

Shunt vessels at the optic disc have been described in retinal vasculitis (Lyle and Wybar, 1961; Lonn and Hoyt, 1966) and in retinal vein occlusion (Vannas and Raitta, 1966). They have been recorded in chronic disc oedema (Sanders, 1969) and seen in papilloedema (personal observation). Pathological studies in central retinal vein occlusion (Klein, 1960) and in papilloedema (Paton and Holmes, 1911) have confirmed the existence of large retino-choroidal anastomoses. Development of the preferential channels was noted in two of our patients in whom a satisfactory visual result was obtained. Development of these channels would seem possible only when the occlusive process occurs within the proximal portion of the vein as it lies in the optic nerve.

CONCLUSION

A series of patients with a benign form of central vasculitis has been described, in only one of whom was an associated systemic disease detected. All were under the age of 40 years so that their vascular systems and potential for developing collateral channels was considered normal.

The symptomatology was often characteristic with a variety of complaints and in three cases the sudden onset favoured a vascular occlusive phenomenon. The visual acuity was only slightly impaired, though the retinopathy was striking. Ophthalmodynamometry readings showed normal arterial pressures with grossly elevated venous pressures. Fluorescein studies demonstrated a normal arterial filling, a slow transit of dye, and marked residual leakage in relation to the disc and larger veins. Visual loss, when it occurred, was associated with capillary decompensation and leakage of dye occurring particularly in the macular region. Resolution occurred within a period of several months with a return to normal visual acuity, but often with persistent micropsia, and in some cases fundus changes consisting of venous sheathing and pigmentary changes at the macula.

It is concluded that the main haemodynamic effect is a reduction in perfusion pressure which is due solely to the elevated venous pressure. The condition is analogous and possibly identical to retinal vein occlusion in the older age groups, though in the latter the

prognosis is poor because of the associated arterial disease and relative inability to develop proficient collateral circulation. The site of occlusion is an important factor.

The aetiology remains obscure, and probably diverse, for many different mechanisms may produce venous obstruction at any site. The systemic administration of steroids does not appear to be specifically efficacious in treatment, and methods at present employed include the systemic administration of Diamox in an attempt to improve ocular perfusion.

This series has not included those cases with vitreous haemorrhages, peripheral neovascularization, and absolute glaucoma, which represent a separate group.

Summary

A series of patients under the age of 40 years with benign central vasculitis has been studied by ophthalmodynamometry and fluorescein angiography. The main haemodynamic abnormality is a reduction in the perfusion pressure which is due to elevated venous pressure. Fluorescein studies show two phases of involvement depending on whether the veins alone or both veins and capillaries are decompensated. The visual outcome is favourable in all cases unless associated systemic disease is present.

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