TRANSIENT VESSEL WALL SHEATHING IN ACUTE RETINAL VEIN OCCLUSIONS

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SUMMARY

Three cases are reported which had features similar to, and evolved in a pattern consistent with central retinal vein occlusions and a fourth case is reported which behaved as a hemispheric vein occlusion. However, they differed from classic retinal vein occlusions by having prominent sheathing of the retinal venous vasculature at presentation, which in all four cases resolved within three weeks. There was no evidence for any of these cases having an inflammatory vasculitis. The significance of this transient sheathing is uncertain.

Three cases are reported of a condition that behaved like ischaemic central retinal vein occlusion (CRVO) and a fourth with a similar appearance, but with the features of a hemispheric vein occlusion. They all differed from classic retinal vein occlusions by their striking fundal appearance at presentation, with sheathing of the walls of the involved retinal venous system. Sheathing of retinal vessels is a recognised late phenomenon in both central and branch retinal vein occlusions, but is not a recognised feature at presentation in the absence of inflammation.

Such a fundal appearance at presentation suggests a differential diagnosis of retinal vasculitis, reticulum cell sarcoma, frosted branch angiitis or papillophlebitis. However, the sheathing resolved within three weeks and otherwise the features and clinical course were similar to retinal vein occlusions.

PATIENTS

Case One

A 23-year-old man presented to casualty with a one-day history of blurring of vision in his left eye. Two months previously he had had a vesicular rash of uncertain cause that affected his back, arm and legs. This had cleared within a week without obvious sequelae. Two weeks

before presentation he had noted a blind spot in the temporal field of his left eye, which had lasted for one day and had resolved.

The vision was 6/5 in the right eye and 6/60 in the left eye with a afferent pupillary defect. There were cells+ in the anterior chamber and fine keratic precipitates were noted. There were no cells in the vitreous. The intraocular pressures were normal in both eyes. The dramatic findings were on fundal examination (see Fig. 1a). All the retinal veins showed sheathing with the tributary venules terminating in retinal haemorrhages. The optic disc was swollen and there was pronounced cystoid macular oedema. There were also widespread scattered retinal haemorrhages. A few cells only were noted in the anterior vitreous.

Fluorescein angiography at this stage showed diffuse venous leakage and macular oedema (Fig. 1b).

Full assessment was performed by a general physician and no evidence of a systemic disorder, particularly Behçet's disease or sarcoidosis, could be found on history, full physical examination or investigation, which included full blood count, biochemistry profile, autoantibody screen (including ANA and ANCA), immunology, virology (including herpes viruses), SACE, VDRL, chest radiograph and urine analysis. A CT scan of his sinuses and orbits showed normal enhancement and a B-scan showed no abnormalities in either the retina or the optic nerve.

He was started on prednisolone at 60 mg/day and had a reducing course over ten days. The vision in his left eye improved to 6/12 and the sheathing had disappeared by one week, but the haemorrhages persisted and a star exudate formed at the macula (see Fig. 1c). By four weeks the vision had deteriorated to 6/36 and the fundal appearances were those of a classical CRVO with cotton wool spots.

By four months, iris rubeosis had developed and fluorescein angiography demonstrated severe ischaemia (Fig. 1d). He received extensive pan-retinal photocoagulation, but developed rubeotic glaucoma. His vision is currently perception of light only.

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Fig. 1a. The fundal appearance of case one at presentation, showing dilatation and sheathing of the retinal veins, widespread retinal haemorrhage and swelling of the optic nerve head.

Fig. 1c. The fundal appearance one week later, showing the resolution of the sheathing and the formation of a macular star.

Fig. 1b. The accompanying fluorescein angiogram shows continuous leakage of fluorescein from the venous system.

Fig. 1d. A fluorescein angiogram performed four months later, showing the development of severe ischaemia.

Case Two

A 32-year-old man presented with a twenty-four hour history of severe left visual loss. He was otherwise perfectly healthy. His vision was 6/5 in the right eye and 6/60 in the left. There were no signs of ocular inflammation and the intraocular pressures were normal. Fundal examination demonstrated engorgement of all the retinal veins with sheathing of the inferior retinal veins and tributaries (Fig. 2a). There was swelling of the optic disc, cystoid macular oedema and widespread scattered retinal haemorrhages. A fluorescein angiogram demonstrated leakage of the inferior retinal veins in a continuous manner (Figs. 2b).

A full assessment by a general physician found no other abnormality. A CT scan of the brain and orbits was normal.

Over the next two weeks the visual acuity remained unchanged, but the sheathing had completely vanished by

the end of the third week (Fig. 2c). The cystoid macular oedema, however, persisted and the visual acuity remained unaltered at 6/60. His condition was unchanged six months later.

Case Three

A 54-year-old man presented to the casualty department with a twelve hour history of severe visual loss in the right eye. The preceding day had been very hot and that evening he drank five pints of beer. He slept heavily face down and woke with his fist pressed against his right eye. The vision in the right eye was counting fingers and 6/5 in left eye with a right afferent pupillary defect. The anterior segment was normal with normal intraocular pressure. The fundal picture was that of a CRVO but the striking feature was the sheathing of the vein walls, which was virtually continuous. There were scattered haemorrhages, cotton wool spots, disc swelling and macular oedema.

(a)

(c)

(b)

He was a non-smoker and normotensive. Full history and examination was performed by a general physician and all investigations were normal, including a FBC, biochemical screen, autoantibodies and a chest radiograph.

Fluorescein angiography performed two days later showed that there was leakage of dye from the venous system in a contiguous manner and not in the patchy manner characteristic of retinal vasculitis. Areas of capillary non-perfusion were seen and pan-retinal photocoagulation was carried out. His vision remained poor due to cystoid macular oedema.

The sheathing was a transient phenomenon and had resolved by ten days, after which the fundal appearance resembled a classical CRVO.

Case Four

A 75-year-old lady presented to casualty with a two-day history of loss of vision in her right eye. The left eye had had no perception of light since childhood, which had been attributed to meningitis. She had no relevant previous medical or ocular history. Examination revealed perception of light vision in the right eye only, with a few cells in the anterior chamber and the anterior vitreous. The ocular pressure was normal. Fundal examination showed a swollen disc and veins. The venous walls showed continuous sheathing. There were widespread retinal haemorrhages and marked macular oedema. The left eye was phthisical.

Fig. 2a. The fundal appearance of case two at presentation, showing the dilatation of the retinal veins, sheathing of the inferior retinal veins and widespread retinal haemorrhages in the inferior half of the fundus. The sheathing of the inferior retinal veins is continuous and many of the sheathed venules are further outlined with haemorrhage.

Fig. 2b. The accompanying fluorescein angiogram shows continuous leakage of fluorescein from the inferior retinal veins.

Fig. 2c. This shows complete resolution of the sheathing, retinal vein dilatation and haemorrhages in case two, by the third week. There are some resolving deposits of hard exudate in the macular region.

The patient was admitted and commenced on 60 mg of prednisolone per day. She was fully examined and investigated by physicians and no evidence for a systemic vasculitis could be found, but a right carotid bruit was noted. Fundus fluorescein angiography suggested an ischaemic CRVO and pan-retinal photocoagulation was performed and steroids gradually withdrawn. The sheathing resolved over eight days. In view of the poor vision at a level that could not be accounted for by a CRVO alone, a CT scan was performed and demonstrated a large frontal meningioma extending into both orbits. She was referred to the neurosurgeons who removed the tumour, but she died post-operatively.

DISCUSSION

Four cases have been presented, which behaved like retinal vein occlusions, but in which transient retinal wall sheathing was present from the onset. The sheathing was continuous with periodic interruptions, which respected a-v crossings, or could be attributed to overlying haemorrhage. The venules often ended in an area of haemorrhage. Some of the venules had haemorrhage running parallel to their walls, outlining the sheathing and giving the vessels a red layer lying just outside, and contiguous with, the sheathing. Fluorescein angiography showed uniform leakage from the vein walls. In all cases the sheathing resolved within three weeks. All four cases had an acute onset and

presented within two days. As there was a wide distribution in age, (23–75 years) this would appear not to be a significant factor.

With the exception of the sheathing, the fundal appearances and the clinical course were consistent with a diagnosis of retinal vein occlusion. The fluorescein angiographic appearances of acute retinal vein occlusion are a delay in the retinal transit time of the dye, with mild to moderate leakage from the vein walls and with or without either macular oedema or capillary closure, which are consistent with the above cases. The differential diagnosis would include the papillophlebitis of Lyle and Wybar, retinal vasculitis, reticulum cell sarcoma and frosted branch angiitis.

Lyle and Wybar described a condition in young people which they called papillophlebitis^{1,2} and which is now considered to be the same as non-ischaemic CRVO.^{3,4} Sheathing in the acute phase was not a described feature. They suggested that it was due to a vasculitis of the central retinal vein. There is no evidence to suggest that CRVOs in young people are predominantly caused by a vasculitic process.⁵

In most cases of retinal vasculitis, there is patchy sheathing with posterior vitreous cells and this is in contrast to the findings in our series. The other recognised condition with continuous retinal vein sheathing and leakage is reticulum cell sarcoma,⁶ of which there was no evidence in any of these patients.

The appearances are reminiscent of frosted branch angiitis.⁷ Frosted branch angiitis is a bilateral, steroid-responsive condition affecting children, with preservation of good vision and without the late development of rubeosis. All these points distinguish this disorder from our cases.

The nature of the sheathing is uncertain. The classic experiments by Kohner and colleagues⁸ on experimental retinal vein occlusions have generated this striking fundal appearance in the pig. There is clear species variation, as it has not been produced in the rabbit or the monkey. However, no difference between the histological appearance of acute vein occlusions could be detected between the monkey or pig experimental models. In the pig, the sheathing resolved if the vein re-opened, suggesting a reversible process (possibly related to ischaemia of the vein wall itself). Abnormal leakage of the vessel walls to tracer material in animal experiments has been demonstrated within 90 minutes of the occlusion,⁹ which was unaccompanied by any marked morphological changes in the rhesus monkey model.¹⁰

The aetiology of retinal venous occlusions is still uncertain. There are well-recognised associations with diabetes mellitus, hypertension¹¹ and elevation of the intraocular pressure.^{12,13} In the third case described, the central retinal vein occlusion occurred after the patient had fallen asleep with his fist pressed against his right eye. It is likely that he

had experienced an acute rise in ocular pressure in the eye, which may have precipitated his occlusion. The postulated mechanism for raised ocular pressure causing retinal vein occlusion is via compression. The fourth case reported had a meningioma, which may also have caused compression.

There is no satisfactory explanation for the aetiology of the prominent retinal vein wall sheathing. Our cases evolved in a manner similar to that associated with retinal vein occlusion, but whether they should be regarded simply as variants of this, or as a separate condition, remains unclear. With awareness of the existence of this condition, further cases are likely to be collected and analysed.

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Key words: Frosted branch angiitis, retinal vasculitis, retinal vein occlusion and sheathing.

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