A case of frosted branch angiitis

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SUMMARY We report a case of frosted branch angiitis in a 16-year-old-girl. She noted a sudden and severe visual disturbance in both eyes, without other systemic symptoms. Diffuse retinal oedema and unusual sheathing of retinal veins were characteristic in both fundi. Fluorescein angiography showed no occlusion of the sheathed retinal veins, but some paravenous extravasation of dye was found in the late phase. With high doses of systemic corticosteroids her fundi and visual acuity improved greatly, though the vessels continued to show severe narrowing. At three months the pattern visually evoked cortical potentials were found to be normal, while flash electroretinograms were absent. No systemic abnormality has been found to explain the aetiology of this condition.

In 1976 Ito et al.¹ reported a case of a 6-year-old-boy who suffered from a bilateral acute uveitis with severe sheathing along all of the retinal vessels. They named this condition 'frosted branch angiitis'. Since this publication five similar cases have been reported in Japan, in Japanese.²⁴ The disease occurs in children and has an acute onset with bilateral visual disturbance. It has been reported to respond well to steroid therapy. We report here the case of a girl who conformed quite well to the above description. The clinical features of previous cases of frosted branch angiitis are summarised and compared with those of similar retinal diseases.

Case report

A 16-year-old girl was first seen on 11 April 1985 with a one-day history of sudden redness and blurred vision in both eyes. Her visual acuity was counting fingers at 50 cm in both eyes. Ciliary injection and a cellular reaction was seen in the anterior chamber. The intraocular pressures were within normal limits. The
vitreous was slightly hazy and the retina was oedematous, with scattered nerve fibre haemorrhages. Severe sheathing along all the veins was found throughout the retina in both eyes except in areas surrounding the optic discs; the sheathing was more severe peripherally (Fig. 1). Dilatation or tortuosity of the veins was not seen. The optic discs were almost normal.

A visual field test showed bilateral enlargement of the blind spot and general deterioration of sensitivity (Fig. 2).

Fundus fluorescein angiography demonstrated that the retinal veins were patent, but extravasation and staining of veins were obvious in the late stages (Fig. 3). Because the patient complained of dizziness and malaise after the injection of fluorescein, fundus angiography was not repeated.
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No systemic abnormality was found. Red and white cell counts, haemoglobin, plasma proteins, urea, and electrolytes were normal. Chest x-rays were also normal. There was no serological evidence of syphilis, toxoplasmosis, or rheumatoid arthritis. A skin test for tuberculosis was negative. Serum complement levels for herpes simplex virus, herpes zoster virus, and cytomegalovirus were within normal limits.

CLINICAL COURSE
The patient was admitted to hospital immediately, and treatment was started with topical cycloplegics and 0.1% betamethasone, and with systemic betamethasone (3 mg/day). This treatment was continued for three consecutive days, but there was no improvement, and the visual acuity fell to counting fingers at 30 cm in both eyes. On the fourth day the patient was therefore switched to 20 mg/day of dexamethasone (Fig. 4). The retinal oedema, sheathing along the veins, and cellular floaters in the anterior chamber improved by the next day. The sheathing almost disappeared on the 10th day, but sheathed veins and adjacent retinal haemorrhages remained in the peripheral areas, and hard exudates similar to the macular star figure were seen in both posterior poles. At this stage irregularities in calibre of the veins and narrowing of the arteries appeared (Fig. 5).

After 20 days of treatment the cells in the anterior chamber had almost disappeared and the visual acuity

Fig. 4 Steroid therapy and time course of visual acuity.
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Fig. 5  Right eye, 22 April 1985. Left, temporal periphery. Diffuse hard exudates and haemorrhages along the veins. Arteries are very narrow. Right, posterior pole. Hard exudates like a macular star figure are seen.

had slightly improved (Fig. 4). By the 40th day her acuity had improved to 6/12 in both eyes, but attenuation of retinal veins continued to be seen. She was discharged on the 41st day. Dexamethasone was reduced gradually and completed on the 45th day. The total dosage of dexamethasone was 141.5 mg.

Three months after the patient’s initial presentation faint hard exudates in the macular areas and abnormal reflexes around the maculae were still present. In the mid and far periphery the vessels were so narrow that they were difficult to recognise, and even in the posterior pole the arteries were severely attenuated. Some blot haemorrhages were present in the mid periphery. Coalescence of small, sharply demarcated, atrophic lesions was seen in the far peripheral areas, especially in the temporal periphery (Fig. 6). Her visual acuity improved to 6/9 five months after onset.

CHANGES OF VISUAL FIELDS
The bilateral central scotoma that appeared six days after initial presentation disappeared in about two weeks. However, generalised constriction of visual fields progressed. Her visual field then enlarged slowly, but the improvement almost stopped near the end of the second month after initial presentation. The patient was left with a general constriction and slight deterioration of central sensitivity. Visual field constriction was more severe on the nasal side, which corresponded well with the atrophic appearance of the temporal peripheral retina.

ELECTROPHYSIOLOGICAL AND PSYCHOPHYSICAL TESTING
Almost no electroretinogram (ERG) response could be elicited on the fifth day after the patient’s initial presentation. Only a slight recovery of the response

Fig. 6  Right eye, 22 July 1985. Left, temporal periphery. Sharply demarcated atrophic lesions and narrowing of vessels like threads. Right, posterior pole. Hard exudates have almost disappeared. Narrowing of vessels, particularly arteries, is seen.
was found on the 54th day, and no further improvement was observed even on about the 100th day. On the other hand pattern visually evoked cortical potentials (pattern VECPs) using check size of 18-9 minutes, contrast of 80%, reversing at a rate of 3 or 10 per second, performed three months after onset were normal both in peak latencies and in amplitudes (Fig. 7). Colour vision of both eyes was normal with the Ishihara test plates and Farnsworth panel D-15, but the left eye showed a mild red-green deficiency with standard pseudoisochromatic plates.

Discussion

The main features of frosted branch angiitis as originally reported by Ito et al. 1 can be summarised as follows: (1) bilateral acute visual disturbance in a healthy child. (2) The whole retina is swollen and there is severe sheathing of the retinal vessels, so that they look like frosted branches of a tree, especially at the periphery. (3) Fundus fluorescein angiography shows almost a normal pattern in the first stage, but leakage of the dye from vessels is seen in later stages; in particular, the sheathed vessels show no sign of occlusion or stasis. (4) Visual fields show a marked concentric constriction during the acute stages of the disease but improve with the reduction of the swelling and inflammation. (5) ERGs are markedly abnormal. (6) No systemic abnormality is found. (7) High doses of systemic corticosteroids are effective in treating the disorder, and the visual acuity recovers to almost a normal level. (8) The disease has not recurred.

The features of the other cases of frosted branch angiitis previously reported are summarised in Table 1.

In all cases both eyes were affected, and the visual acuity fell rapidly within a week to less than 6/30.

The fundus findings in these cases were similar and can be summarised as follows: (1) The optic discs were almost always normal, though slight oedema or hyperaemia was seen in some cases. (2) In the acute stage the whole retina was oedematous and opacified. Intense sheathing of retinal vessels, especially veins, was seen throughout the fundi, but was severer in the peripheral areas, showing the appearance of frosted branches of a tree. These inflammatory changes gradually became normal by absorption of retinal oedema and improvement in the sheathing. (3) Geographic atrophic lesions remained at the periphery in most cases. Hard exudates were often seen in the maculae or elsewhere during the absorption of the retinal oedema.

In the present case intense narrowing of vessels remained after subsidence of inflammation in addition to the above mentioned peripheral atrophic lesions. As the retinæ were atrophied by the severe inflammation, it is doubtful whether the vascular narrowing will return to normal.

From the above summary of reported cases, including ours, the pathological condition of frosted branch angiitis can be defined as follows. It is an acute panuveitis with severe vasculitis that occurs in the whole retina. Sakanishi et al. 2 suggested that the atrophic lesions that remained at the periphery are a consequence of poor blood perfusion of the peripheral retina. The peripheral areas are therefore more susceptible to damage. The present patient had briefly a central scotoma similar to optic neuritis, but the cause is likely to be macular oedema rather than direct damage to the optic nerve. This explanation is supported by the exudates that appeared later in the macular area. Diminished ERGs recorded three months after onset suggest that substantial damage occurred to the retina as a whole. The normal pattern
VECPs found at the same time, however, indicate that the visual pathway from macula to occipital lobe returned to a normal condition.

No aetiological abnormality has yet been identified. However, systemic steroid was reported to be effective. Although the natural course of frosted branch angiitis is unknown, anti-inflammatory drugs are essential to minimise retinal inflammatory damage.

The differential diagnosis includes four diseases with sheathing of retinal vessels:

1. Acute retinal necrosis (Kirisawa type uveitis and bilateral acute retinal necrosis). Sheathing is also seen, but is mainly restricted to arteries. Fluorescein angiography shows occlusion of arteries and capillary beds. This disease may occur monocularly, and in bilateral cases the day of onset in each eye is usually different. In addition children are rarely affected. The prognosis is extremely poor in contrast to that of frosted branch angiitis.

2. Peripheral uveitis. This disease has been considered to be not a single entity but a syndrome from various causes. Though sheathing of veins may be seen in peripheral areas, subjective signs are commonly mild.

3. Retinitis caused by cytomegalovirus or herpes simplex virus. These viral infections tend to affect patients who are immunosuppressed. Pathological changes of the fundus are not so diffuse as in frosted branch angiitis. Titres against these viruses have always been negative in frosted branch angiitis.

4. Eales' disease. Although vessels may be sheathed in peripheral areas, fluorescein angiography shows obliteration of retinal capillaries and vessels.

Central visual acuity is retained unless vitreous haemorrhages cover the posterior pole.

Frosted branch angiitis seems to be a newly recognised disease, and further studies will be necessary to understand its causation.

References

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