Placoid pigment epitheliopathy

Presenting with bilateral serous retinal detachment

A. C. BIRD* AND A. M. HAMILTON†

Moorfields Eye Hospital, City Road, London, E.C.1

Gass (1968) described a syndrome affecting adults which he termed "acute posterior multifocal placoid pigment epitheliopathy". The patients present with bilateral visual loss and the condition is characterized by the presence at the posterior pole of multiple non-elevated grey-white lesions at the level of the pigment epithelium. Fluorescein angiography shows that these small lesions occlude the choroidal fluorescence during initial transit of dye, and become hyperfluorescent during subsequent minutes. The condition resolves spontaneously within a few weeks and visual recovery is usually good though it may be incomplete. After recovery scattered areas of depigmentation with pigment clumping remain.

In this paper two patients with features of acute posterior multifocal placoid pigment epitheliopathy are described, both of whom presented with detachment of the posterior retina.

Case reports

Case 1, a 28-year-old female student, presented with progressive blurring of vision of both eyes and micropsia for 5 days.

Her finger had become infected after mild trauma 3 weeks before and she had received an intramuscular injection of penicillin followed by a 5-day oral course. She had also been given a single injection of tetanus toxoid. She was otherwise well.

Examination

On admission, the visual acuity in the right eye was reduced to finger counting at 1 m. and in the left to 6/60. No cells were seen in the aqueous humour or vitreous body. There were two small serous detachments of the retina at the right posterior pole associated with multiple pale discrete lesions at the level of the pigment epithelium. The fundus was otherwise normal. Identical changes were seen in the left eye.

Angiography

Fluorescein angiography was performed on admission. During the initial transit of dye there were multiple discrete areas in which the background fluorescence was occluded. During the following minute these areas became hyperfluorescent (Fig. 1*a*), and within 5 minutes of dye injection the serous detachments were outlined by dye in the subretinal space (Fig. 1*b*). Identical changes occurred in the left eye (Fig. 1*c*).

Laboratory investigations

The following investigations were performed and the results were normal: haemoglobin, white cell count, erythrocyte sedimentation rate, serum proteins and electrophoresis, immunoglobulins (IgG, IgA, IgE, and IgM), Paul-Bunnell (sheep's red cells), and standard serological tests for syphilis.

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Address for reprints: A. M. Hamilton, F.R.C.S., Moorfields Eye Hospital, City Road, London EC1V 2PD.

*Leverhulme Fellow, Institute of Ophthalmology, London

†Wellcome Lecturer, Institute of Ophthalmology, London

FIG. 1 Case 1. *Fluorescein angiography at first examination*

Progress

Fluorescein angiography was repeated 3 weeks after presentation at which time the serous detachments had resolved and the visual acuity was 6/18 in each eye. Slow irregular filling of the choriocapillaris (Fig. 2a-c) was demonstrated, but there was no leakage into the subretinal space. Visual recovery was complete 3 weeks later.

Result

3 months after presentation the only abnormality to be seen was patchy depigmentation at the posterior poles, and fluorescein angiography revealed only transmission defects of the pigment epithelium.

Case 2, a 21-year-old female clerk, presented with a 2-day history of progressive blurring of vision in the left eye. During the previous year she had received many courses of antibiotics for recurrent cystitis and was currently receiving penicillin. She was otherwise healthy.

Examination

Visual acuity in the right eye was 6/5 and in the left 6/24. At the left posterior pole there were multiple discrete cream-coloured lesions at the level of the pigment epithelium, and a serous detachment of the retina at the macula. The fundus was otherwise normal and no lesions were present in the right eye. No cells were seen in the aqueous humour or vitreous body of either eye.

Angiography

Fluorescein angiography was performed on admission and the pigment epithelial lesions blocked out the background choroidal fluorescence during the initial transit of dye (Fig. 3a). During the following minute these lesions became hyperfluorescent (Fig. 3b). Fluorescein then leaked into the subretinal space and the retinal detachment was still outlined by fluorescein 30 minutes after dye injection (Fig. 3c). Photographs of the right eye at this time were normal.

FIG. 2 Case 1. Fluorescein angiography of right eye 3 weeks later

- (a) 2 sec. after dye entry
- (b) 3 sec. after dye entry
- (c) 6 sec. after dye entry

Laboratory investigations

The haemoglobin, white cell count, and erythrocyte sedimentation rate were within normal limits, and the Paul-Bunnell screening test was negative.

Progress

10 days later the patient noticed distortion of vision with the right eye, though the sight of the left eye was improving. The visual acuity in the right eye was 6/6 and in the left was 6/9. Multiple cream-coloured lesions had now developed at the level of the pigment epithelium at the posterior pole of the right eye, and there were small serous detachments of the retina around the macula. Fluorescein angiography showed changes in the right eye identical to those seen in the left eye 10 days previously (Fig. 4*a,b*). Late pictures of the left eye showed no leakage of fluorescein into the subretinal space.

Result

Complete visual recovery occurred within a further 4 weeks and there was patchy depigmentation at the posterior poles. Fluorescein angiography was now normal apart from transmission defects in the pigment epithelium.

FIG. 3 *Case 2. Fluorescein angiography of left eye at first examination*

- (a) 7 sec. after dye entry
- (b) 55 sec. after dye entry
- (c) 30 min. after dye entry

FIG. 4 *Case 2. Fluorescein angiography of right eye 10 days later*

- (a) 6 sec. after dye entry (central bright patch due to mirror reflex)
- (b) 5 min. after dye entry

Discussion

Acute posterior multifocal placoid epitheliopathy appears to be a specific clinical entity occurring in young people. Absence of overlying serous retinal detachment in previously described cases (Gass, 1968; Van Buskirk and others, 1971) is remarkable in view of the intensity of the pigment epithelial lesions. Both patients described in this report presented with serous detachment of the posterior retina, though in each case the retina became flat within a short time. Cases 1 and 3 reported by Gass (1968) were said to have had macular oedema in the early stages of the disease, though the retina was flat when the patients were seen later by the author. Contrary to the impression given by previous reports, central serous detachment of the retina may be an early feature of placoid pigment epitheliopathy.

The pathological changes are unknown. Gass (1968) felt that this was a primary disease of the pigment epithelium due to a local infectious or toxic agent rather than to focal ischaemia. He interpreted the fluorescein changes as occlusion of the background choroidal fluorescence due to diseased pigment epithelium, and that this abnormal pigment epithelium later absorbed fluorescein rendering that area hyperfluorescent. Van Buskirk and others (1971) demonstrated slow irregular filling of the choroidal capillaries and gave a different interpretation of the fluorescein angiograms. They concluded that the changes could be explained on the basis of irregular filling of the choroid due to focal choroidal vasculopathy.

Our findings early in the disease clearly show that there are multiple well-defined areas of hypofluorescence during the initial transit of dye. These areas remain discrete and of a constant size for 35 to 50 sec. after dye entry, and only after that period do they become hyperfluorescent. This supports the hypothesis of Gass (1968) that there is focal occlusion of the background choroidal fluorescence probably as a result of pigment epithelial changes. In Case 1 fluorescein angiography during recovery showed irregular filling of the choroidal capillaries over a period of 6 sec., during which there was gradual contraction of the non-filling areas. These changes are identical to those described by Van Buskirk and others (1971).

It is probable that, in the acute stage of the disease, reduced blood flow in the choroidal capillaries causes focal swelling of the pigment epithelium due to ischaemia. Occlusion of the background fluorescence during the initial transit of dye is due to swelling or infiltration of the pigment epithelium, and dye subsequently leaks into the pigment epithelial cells and subretinal fluid. Irregular choroidal filling only becomes apparent when normal transparency is restored to the pigment epithelium. Ischaemia may appear unlikely in view of the good visual recovery. However, the pigment epithelial lesions are discrete and the visual prognosis may depend upon their precise location. If the pigment epithelium beneath the foveola is unaffected, visual recovery is likely to be good.

The aetiology of this condition is unknown although it has been described in a patient with erythema nodosum (Van Buskirk and others, 1971) and two reported cases had positive Mantoux tests (Gass, 1968).

Both cases reported here had taken oral penicillin either shortly before or at the time of onset of visual symptoms. It is possible that in these cases placoid epitheliopathy is a manifestation of a hypersensitivity to penicillin.

Summary

Two patients with acute multifocal posterior placoid pigment epitheliopathy had bilateral central serous detachment of the retina in the early stages of the disease. Fluorescein angiograms support the concept of focal choroidal ischaemia as a cause for this syndrome.

References

- GASS, J. D. M. (1968) *Arch. Ophthalm. (Chicago)*, **80**, 177
VAN BUSKIRK, E. M. A., LESSELL, S., and FRIEDMAN, E. (1971) *Ibid.*, **85**, 369