Ocular injury in the battered baby syndrome

Report of two cases

ALAN MUSHIN AND GWYN MORGAN

Departments of Clinical Ophthalmology and Pathology, Institute of Ophthalmology, University of London

In the differential diagnosis of obscure skeletal and soft tissue lesions in children it is important to consider the possibility of physical abuse. The association of chronic subdural haematoma with multiple fractures in the long bones of infants was first reported by Caffey (1946), but he failed to confirm the traumatic origin he suspected. Silverman (1953) also described skeletal abnormalities in infants due to trauma, but was hesitant to blame the custodians of the child. The problem was high-lighted by the study of Adelson (1961) of 46 child murders; he emphasized the need for complete autopsies of all children "found dead" or who presented bizarre clinical pictures not explained by known syndromes. Kempe, Silverman, Steele, Droegemueller, and Silver (1962) seem to have been the first to recognize the full implications of the problem and coined the term "battered child syndrome" – now usually called the "battered baby syndrome".

At the present time in America deaths from all types of accidents account for nearly twice as many childhood deaths as cancer, and Fontana (1964) suggested that many of these accidental deaths may be due to undetected or unreported parental abuse.

Kiffney (1964) was the first to describe ocular injuries in this syndrome – retinal detachment with multiple skull fractures and bilateral subretinal haemorrhages. One eye was enucleated and provided the first opportunity for histological examination, but there were no special features apart from the detachment. Gilkes and Mann (1967) reported extreme degrees and persistence of retinal haemorrhages, with venous engorgement, snowy exudates and papilloedema, which reminded them of the fundus picture described by Purtscher (see Duke-Elder, 1954). Maroteaux, Fessard, Aron, and Lamy, (1967) and Maroteaux and Lamy (1967) described atrophic lesions in the retinal periphery in cases of the battered baby syndrome.

The purpose of this paper is to present the clinical and pathological features of two eyes which were removed from children with this syndrome.

Case 1

CLINICAL HISTORY

A male infant aged 8 months who was cared for during the day by a "baby-minder" while both parents were at work, was brought to hospital with a bulging anterior fontanelle and bilateral retinal haemorrhages. He became drowsy and was found to have bilateral subdural haematomas.

These were drained and after a stormy course the child recovered. The haemorrhages in the right eye cleared, but in the left eye vitreous haemorrhage occurred. He was not brought for follow-up and was next seen 6 months later with a painful and swollen left eye.

Examination showed that the left eye was larger than the right (vision normal), and it was red, with corneal oedema and raised tension, aqueous flare, and a small hyphaema. The lens was pushed forward and subluxated below. Ectropion uveae and iris atrophy were present, and the vitreous was filled with blood which obscured the fundus. The eye, being blind and painful, was enucleated.

PATHOLOGY

Macroscopical examination

There was hyphaema, an eccentric pupil, and total retinal detachment with subretinal sanguineous exudate. No neoplasm was seen (Fig. 1).

FIG. 1 Photograph of sectioned left eye (Case 1), showing a hyphaema, retinal detachment, and sanguineous subretinal exudate. The iris shows ectropion, and the ciliary processes are drawn toward the lens by fibroblastic tissue

Microscopical examination

The cornea showed limbal infiltration and superficial vascularization. Peripheral anterior synechiae occluded the filtration angle on both sides, and the anterior chamber contained haemorrhagic exudate. The iris showed pigment scattering, rubeosis, and ectropion, and on one side was fibrosed and contracted thus distorting the pupil. There was extreme pigmentary disturbance in the ciliary processes which were covered with fibroblastic tissue and drawn towards the lens. The latter was subluxated and cataractous and adhered posteriorly, through a vascularized hyaline membrane, to the totally detached retina which was disorganised and necrotic, showing lipid macrophages (Fig. 2) and also neovascularization into the vitreous (Fig. 3). Lipoidal macrophages and cholesterol crystals were present in the abundant subretinal fluid (Fig. 4). The optic nerve was engorged and gliosed, while the choroid and sclera were normal.

DIAGNOSIS

The picture is that of post-concussional injury. Before the history of trauma was known the histological diagnosis of Coats's disease was made.

FIG. 2 Portion of the detached retina, showing disorganization, lipid macrophages, and necrosis (Case 1). Haematoxylin and eosin. $\times 90$

FIG. 3 Neovascular tissue extending into the vitreous from the detached retina (Case 1). Haematoxylin and eosin. $\times 90$

FIG. 4 Subretinal fluid, showing cholesterol clefts and lipid macrophages ("ghost cells") (Case 1). Haematoxylin and eosin. $\times 90$

Case 2

CLINICAL HISTORY

A male infant $3\frac{1}{2}$ months old was strangled with a blanket by his father. The baby fainted and had convulsions, and the parents spent the rest of the night shaking the child to revive him. Next day he was admitted to hospital moribund with extensive bruising and bilateral intraocular haemorrhages. His condition deteriorated and he died 24 hours after strangulation. *Post-mortem* examination showed a large subdural haemorrhage, pharyngeal bruising, and scattered haemorrhages. The left eye was examined at the Institute.

PATHOLOGY

Macroscopical examination

The eye was opened horizontally. The retina was *in situ* (Fig. 5) and showed extensive haemorrhages.

FIG. 5 Photograph of sectioned left eye (Case 2), showing retina *in situ*

Microscopical examination

The only abnormalities to be found in the globe were sited in the posterior segment, where there were extensive intraretinal, subhyaloid, and small vitreous haemorrhages (Figs 6 and 7). Some sections showed myelin extrusion at the disc.

FIG. 6 Haemorrhage in inner layers of retina (Case 2). Haematoxylin and eosin. $\times 50$

FIG. 7 Haemorrhage in subhyaloid space (Case 2). Haematoxylin and eosin. $\times 90$

DIAGNOSIS

Intraocular haemorrhages due to strangulation.

Comment

These two cases illustrate the fact that the "battered baby syndrome" must be borne in mind in the differential diagnosis of ocular conditions in childhood, not only by the clinician, but perhaps more especially by the ocular pathologist, who might well be the first to recognize that the lesions are traumatic in origin. In the absence of a history of injury, as in the majority of cases, he may be trapped into making a misdiagnosis. Trauma should especially be considered in pseudoglioma, Coats's disease, lens dislocation, and all forms of old or recent intraocular haemorrhage. Case 1 was of particular interest because an erroneous histological diagnosis of Coats's disease was made in the absence of any history of trauma. The histological features of this eye simulated very closely those seen in Coats's disease, and the presence of cholesterol crystals and lipoidal macrophages in the sero-sanguineous exudate can probably be explained by chronic or recurrent haemorrhages or exudation. There seems little doubt that other cases which were previously diagnosed as Coats's disease may in fact be examples of the "battered baby syndrome".

We are grateful to Prof. Norman Ashton for his advice during the preparation of this report.

References

- ADELSON, L. (1961) *New Engl. J. Med.*, **264**, 1345
 CAFFEY, J. (1946) *Amer. J. Roentgenol.*, **56**, 163
 DUKE ELDER, S. (1954) "Text-book of Ophthalmology", vol. 6, p. 6369. Kimpton, London
 FONTANA, V. J. (1964) "The Maltreated Child". Thomas, Springfield, Ill.
 GILKES, M. J., and MANN, T. P. (1967) *Lancet*, **2**, 468
 KEMPE, C. H., SILVERMAN, F. N., STEELE, B. F., DROEGEMUELLER, W., and SILVER, H. K. (1962) *J. Amer. med. Ass.*, **181**, 17
 KIFFNEY, G. T., JR. (1964) *Arch. Ophthal. (Chicago)*, **72**, 231
 MAROTEAUX, P., FESSARD, C., ARON, J. J., and LAMY, M. (1967) *Presse méd.*, **75**, 711
 ——— and LAMY, M. (1967) *Lancet*, **2**, 829
 PURTSCHER, O. (see Duke-Elder, 1954)
 SILVERMAN, F. N. (1953) *Amer. J. Roentgenol.*, **69**, 413