BILATERAL ENDOPHTHALMITIS ASSOCIATED WITH SICKLE-CELL HAEMOGLOBIN C DISEASE*

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SICKLE-CELL haemoglobin C disease is the result of the combination of two abnormal haemoglobins, Hb S and Hb C, and is found almost exclusively in the Negro race. It is characterized by recurrent infarctive episodes, which vary from the mild and asymptomatic to the severe and fatal. The infarction of an organ or tissue is due to the mechanical obstruction of the small capillaries by distorted inflexible red cells. The organs mainly affected are the spleen, the bones, the kidneys, and the lungs, but any organ may be involved and the eyes are no exception.

About 31 cases with ocular complications in sickle-cell haemoglobin C disease have so far been described by different authors, including Edington and Sarkies (1952), Smith and Conley (1954), Hannon (1956), and Munro and Walker (1960). Lieb, Geeraets, and Guerry (1959) described four stages in the retinopathy in sickle-cell anaemia:

1. Tortuosity and dilatation of the vessels with small peripheral ischaemic areas;
2. Areas of stasis, micro-aneurysms, and early new vessel formation;
3. All forms of retinal haemorrhages and marked new vessel formation;
4. Retinitis proliferans, vitreous haemorrhages, venous thrombosis, and papilloedema.

Munro and Walker (1960) stated that vitreous haemorrhages, uveitis, and cataract may be additional complications of the disease. Kennedy and Cope (1957) described peripheral retinal gliosis and recurrent vitreous haemorrhages in one case, which progressed finally to uveitis and hypotony.

As far as we know, no case of sickle-cell haemoglobin C disease has been associated with bilateral endophthalmitis.

Case Report

A male Jamaican aged 35 years, who came to England 7 years ago, was admitted to Whipps Cross Hospital on September 1, 1962, with loss of vision in both eyes for 6 days.

6 days before admission he had gone to work feeling quite well, when at lunch time he felt cold and shivery and developed pains in both elbows and knees, and in the lower half of the left side of the chest. That same evening he noticed blurring of vision in the right eye; next morning his left eye was also affected and by the evening he was virtually blind.

Past History.—Scars on the lower part of both legs were due to chronic ulceration in childhood. He had had mild intermittent joint pains for as long as he could remember but they never necessitated his staying home from work.

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692
ENDOPHTHALMITIS AND SICKLE-CELL HAEMOGLOBIN C

In 1958 he was admitted to the Whittington Hospital with pleurisy, and a pleural biopsy revealed the presence of purulent granulomatous tissue. Blood investigation showed that he was suffering from sickle-cell haemoglobin C disease. The chest lesions were considered to be pulmonary infarctions due to his disease.

Family History.—Nothing relevant.

Ophthalmological Examination.—The visual acuity was perception of light in the right eye and hand movements in the left eye. Both eyes presented the same picture with congestion, chemosis, and oedema of the corneal endothelium. The anterior chambers were deep and full of a heavy plastic exudate. Both pupils were small, fixed, and plugged with exudate, which obscured the fundus and vitreous. The globes were soft and tender. Mydriatics produced full dilatation of the left pupil and partial dilatation of the right, which was bound in the lower half by posterior synechiae. The entire retina was covered and obscured by a thick exudate and the fundus appeared as a grey reflex.

Laboratory Findings

Blood Count: Polymorph leucocytosis 8,000–19,000/c.mm.; Hb 72–88 per cent.; erythrocyte sedimentation rate 12–46 mm./hour.

Blood Film: Some target cells and sickling demonstrated under reduced oxygen tension.

Hb Electrophoresis: Abnormal Hb S and Hb C.

Blood Cultures: Taken on September 4 and 28, 1962, both sterile.

Conjunctival Swabs: No organisms grown.

Agglutination Tests: Brucellosis, toxoplasmosis, Wassermann, Kahn, and gonococcal complement-fixation; all negative.

Virus Investigation: Negative

Anti-Streptolysin Titre: 50 units/ml.

Urine: Normal

Mantoux Test: Negative 1:100

Chest X Ray: Increased broncho-vascular markings.

Treatment.—He was given local mydriatics, antibiotics, corticosteroids, and systemic steroids, and treatment on antituberculous lines.

Progress.—After one week the right eye showed signs of increasing irritation and a raised ocular tension. The left eye was quiet. 4 weeks later the irritation of the right eye subsided. Visual acuity was still reduced to perception of light in both eyes. The exudates in both anterior chambers had absorbed leaving a heavy flare, numerous cells, and large keratic precipitates. The massive exudate in the right vitreous had new vessels growing in from the periphery and the anterior surface had advanced to lie hard up against the posterior surface of the lens. A similar sequence of events now began in the left eye with increased irritation, raised intra-ocular pressure, and neovascularization of the vitreous exudate. Three months after admission both eyes had become quiet and painless, cataracts had formed, hypotony had developed, and the perception of light was lost.

Discussion

The aetiology in this case is not clear. The clinical picture suggested an infection with an organism of low virulence or a profound uveitis. Sickle-cell haemoglobin C disease is known to produce uveitis, but Prof. E. S. Perkins (personal communication) had not seen a case of uveitis that so rapidly resulted in loss of light perception.
Neither had he seen a case of sickle-cell haemoglobin C disease with such a profound uveitis. If this uveitis was caused by infarction of the ciliary and choroidal vessels it is surprising that there was no evidence of cerebral infarction.

We are unable to offer a definite aetiology but suggest that there was an initial uveitis with a superimposed metastatic infection. The uveitis may have been due to bilateral infarction of ciliary vessels similar to the infarctive lesions of the pleura.

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REFERENCES
