FUNDAL LESIONS IN SARCOIDOSIS

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INvolvement of the eye occurs in approximately one-quarter of patients with sarcoidosis. In various large series the figure varies from 28% to 50% (Table 1). The commonest type of involvement is anterior uveitis, occurring in from 23% to 90% of patients with ocular involvement in these series. Not surprisingly, posterior uveitis is less frequently discernible because it is hidden by inflammation of the anterior segment of the eye. However, in recent years the introduction of corticosteroids has changed the natural history of ocular sarcoidosis because these drugs readily control acute iridocyclitis. Thus when disease of the anterior segment is rapidly controlled it is possible to see the fundus oculi at a much earlier stage when sarcoid lesions are still present. We feel that this will become increasingly evident in the future. In a series of 450 patients with sarcoidosis attending the Royal Northern Hospital Sarcoidosis Clinic, 16 have been observed with fundal lesions (Table 2). Apart from the eye lesions they do not differ significantly in any parameter.

Choroiditis
In our series, 11 patients exhibited choroiditis. In posterior uveitis the primary lesion is always in the choroid. There are two distinct forms of sarcol choroiditis. One is a non-specific type which cannot be distinguished ophthalmoscopically from any other form of posterior uveitis (Crick, 1961, Green and Kennedy, 1957). In this form the abnormal findings on ophthalmoscopy are usually restricted to several well-defined whitish nodules, juxtapapillary or peripheral in distribution; if the disease is active the outlines of these nodules may be obscured by inflammatory exudate from the surrounding tissues (Dressier and Wagner, 1941, Roberts and Neison, 1945, Gifford and Crowther, 1949, Morax, 1956, Bruntse, 1958 and Hudelo, 1962).

In the specific type of choroiditis, thought to occur in sarcoidosis only, yellowish-white patches of irregular shape and size are distributed along the retinal vessels, usually the veins. These lesions were first described by Walsh (1939). Franceschetti and Babel in 1949 used the term chorido-retinitis "en tache de bougie" to describe this appearance which has also been described by several other writers (Levitt, 1941, Lewis, 1950, Mackensen, 1952, Morax, 1956, Gould and Kaufman, 1961 and Hudelo, 1962). These nodules nearly always constrict the retinal vessels along which they are distributed and cause perivasculitis.

TABLE 1
MANIFESTATIONS OF OCULAR SARCOIDOSIS IN VARIOUS SERIES

| Authors       | Year | No. of patients | No. | %       | No. | %       | No. | %       | No. | %       | No. | %       | No. | %       | No. | %       | No. | %       | No. | %       | No. | %       |
|---------------|------|-----------------|-----|---------|-----|---------|-----|---------|-----|---------|-----|---------|-----|---------|-----|---------|-----|---------|-----|---------|-----|---------|-----|---------|
| Gifford and  | 1949 | 30              | 10  | 33      | 9   | 90      | 1   | 10      | 0   | 0       | 2   | 20      | 0   | 0       | 7   | 70      | 0   | 0       | 7   | 70      |
| Krause       |      |                 |     |         |     |         |     |         |     |         |     |         |     |         |     |         |     |         |     |         |
| Longcope     |      |                 |     |         |     |         |     |         |     |         |     |         |     |         |     |         |     |         |     |         |
| Freiman      | 1952 | 142             | 53  | 37      | 28  | 53      | 1   | 2       | 3   | 6       | 10  | 15      | 0   | 0       | 1   | 2       | 6   | 12      | 5   | 10      |
| Crick et al  | 1961 | 185             | 93  | 50      | 21  | 23      | 40  | 43      | 1   | 1       | 0   | 0       | 13  | 14      | 5   | 5       | 58  | 62      | 5   | 5       |
| James et al  | 1964 | 442             | 123 | 28      | 89  | 72      | 11  | 9       | 1   | 1       | 7   | 6       | 6   | 8       | 5   | 4       | 10  | 81      | 2   | 2       |
| TOTAL        |      | 799             | 279 | 35%     | 147 | 18.3%   | 53  | 6.6%    | 5   | .5%     | 19  | 2.3%    | 21  | 2.6%    | 18  | 2.2%    | 74  | 9%      | 19  | 2.3%    |
TABLE 2

| Clinical, Radiological and Immunological Features of 16 Patients with Fundal Sarcoïdosis |
|------------------------------------|-----------------------------------|
| Feature                             | No. | %       |
| Male                                | 7   | 44      |
| Female                              | 9   | 56      |
| 21-30                               | 7   | 44      |
| 31-40                               | 5   | 31      |
| Age                                 | 41-50 | 3 | 19     |
|                                     | 51-60 | 1 | 6      |
| Bilateral fundal involvement        | 8   | 50      |
| Unilateral fundal involvement       | 8   | 50      |
| Chest X-ray abnormalities           | 10  | 63      |
| Enlarged lymph nodes                | 8   | 50      |
| Positive Kveim test                 | 11/11 | 100     |
| Negative Mantoux test               | 7/12 | 58      |

Papilledema

There were 3 patients with papilledema. Two also had cranial nerve palsies when they presented in a neurological department and had burr-holes made in the course of the investigation of their raised intra-cranial pressure before a diagnosis of sarcoidosis was considered. In one case the papilledema was confined to the left eye but there was a right 6th nerve palsy.

Meningeal or encephalitic sarcoidosis may produce raised intra-cranial pressure. Papilledema is seen, with a swollen hyperaemic disc with blurred margins, distension of the veins and hemorrhages and oedema extending to the macula (Roberts and Neilson, 1945, Colover, 1948, Fine and Flocks, 1953 and Ross, 1955).

Spread of sarcoid tissue from the meninges or the third ventricle may involve the optic nerve or chiasma. Lesions in the nerve head may present as papillitis (Morax, 1956) or as a pseudo-tumour (Walsh, 1939, Laval, 1952, Alajouanine, 1952, Green, 1957). Meitke (1958) described a pseudo-tumour producing 20 dioptries of swelling of the disc.

Optic Atrophy

In sarcoidosis, optic atrophy is of the consecutive type following papilledema (Walsh, 1939, Klein, Calvert, Joseph and Smith, 1955).

Optic nerve lesions are always associated with visual disturbances. In the presence of papilledema produced by raised intra-cranial pressure there may be blurring of vision with enlargement of the blind spot and constriction of the peripheral fields. In papillitis the blurring of vision will be transient and there may be central scotomata. Field loss of varying types depending on the site of the lesion will be caused by involvement of the optic nerve. In optic atrophy and pseudo-tumour the visual loss may be severe.

Course and Treatment

Fundal sarcoidosis has been insufficiently recognised and followed for the natural history of this form of the disease to be clear. Untreated choroiditis may heal within weeks or remain active for months; in either case scar tissue with surrounding pigmentation persists. Juxta-papillary and peripheral lesions heal with little or no residual visual disturbance, but macular and perimacular lesions leave permanent impairment of vision.

Preretinal and vitreous hemorrhages usually absorb when the perivasculitis settles though should the hemorrhages occur repeatedly or be
prolonged they may organise and retinitis proliferans may result.

Papilloedema and papillitis may resolve spontaneously within a few weeks or months; two in this group cleared up with, and one did well without steroids.

Today corticosteroids should be given to all patients with fundal sarcoidosis. Systemic treatment is essential for topical applications do not influence fundal lesions. Choroiditis and perivasculitis usually respond rapidly to treatment and Alajouanine and others (1952) report the disappearance of pseudo-tumour and optic disc swelling. Scar tissue, retinitis proliferans and optic atrophy are unaffected by treatment.

All but two of the patients with choroiditis received steroids and none of those treated was left with any visual defect. Of the two who had sarcoidosis before steroid treatment was available one had an eye enucleated and the other recovered.

Summary

Since corticosteroids have accelerated resolution of inflammation of the anterior segment of the eye, abnormalities of the fundus oculi are being more readily recognised in sarcoidosis.

In a series of 450 patients with sarcoidosis, 16 had fundal lesions comprising choroiditis (11 patients) papilloedema (3) and perivasculitis retinae (2).

The majority received systemic corticosteroids. Treated patients had no residual complications. In one of three untreated patients, enucleation of the eye was necessary.

REFERENCES


MEITKE, H. (1958): A Rare Ocular Manifestation of Sarcoid, ibid, 133, 891.


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