

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 sarcoid 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 (Dressler and Wagner, 1941, Roberts and Neilson, 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 choroido-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	Patients with ocular involvement		Anterior uveitis (iritis & iridocyclitis)		Posterior uveitis (choroiditis)		Other fundal lesions		Eyelids & lacrimal glands		Heerfordt's syndrome		Conjunctival follicles		Kerato-conjunctivitis sicca		Corneal	
			No.	%	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
Gifford and Krause	1949	30	10	33	9	90	1	10	0	0	2	20	0	0	7	70	0	0	7	70
Longcope and 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	8	6	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
SARCOIDOSIS

	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

Choroido-retinitis may also occur in the non-specific form of sarcoid choroiditis if the granulomata involve the retina. If the granulomata project into the vitreous the mass is called a "sarcoidoma" (Walsh, 1939, Goldberg and Newall, 1944 and Ozarewski and Bennett, 1952).

Peripheral choroiditis is symptomless. Involvement of the macula will cause blurring and loss of vision. Fundal lesions are painless but an associated anterior lesion may cause pain; the more severe the posterior lesion, the greater the probability that it will be associated with some anterior manifestation of the disease (Geraets, 1962).

Perivasculitis Retinae

This was observed in two of our patients. Retinal perivasculitis, usually periphlebitis, may occur without choroiditis if the sarcoid nodule forms in the vessel wall itself (Gould and Kaufman, 1961, Geraets, 1962). Marked variation in calibre of affected vessels is seen with white exudate and sheathing along their walls. If the vessel is not obstructed the condition is symptomless; should occlusion occur, perivenous exudates and hæmorrhages will form and may, if extensive, become preretinal and vitreous.

Vitreous Opacities

These were not seen in any of the patients in this series, although according to Crick they are commonly seen in ocular sarcoidosis. They are globular, greyish-white, and usually found in the lower vitreous near the retina (Landers, 1949, Roberts and Neilson, 1945, Bruntse, 1958). Their origin is not clear; Crick thinks that they are formed in the ciliary body and are carried by gravity to the lower vitreous where a retinal reaction occurs.

Papilloedema

There were 3 patients with papilloedema. 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 papilloedema was confined to the left eye but there was a right 6th nerve palsy.

Meningeal or encephalitic sarcoidosis may produce raised intra-cranial pressure. Papilloedema is seen, with a swollen hyperæmic disc with blurred margins, distension of the veins and hæmorrhages 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 papilloedema (Walsh, 1939, Klein, Calvert, Joseph and Smith, 1955).

Optic nerve lesions are always associated with visual disturbances. In the presence of papilloedema 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 hæmorrhages usually absorb when the perivasculitis settles though should the hæmorrhages 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.

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