Acute presumed histoplasmosis of the optic nerve head

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The classic fundus findings of presumed histoplasmosis are well known and include peripheral chorio-retinal scars, haemorrhagic disciform disease of the macula, and peripapillary chorio-retinal scarring (Woods and Wahlen, 1960; Walma and Schlaegel, 1964). The development of new peripheral chorio-retinal scars and disciform macular lesions has been observed clinically. However, to the best of our knowledge the development of the typical diffuse peripapillary atrophy has not been described. A case is reported in this paper in which the development of peripapillary atrophy and peripheral chorio-retinal lesions were documented in the normal right eye of a patient with typical presumed ocular histoplasmosis lesions in the left eye.

Case report
A 38-year-old man was referred to Letterman Army Medical Center with sudden loss of vision in the right inferior temporal field. The visual loss had been preceded by a moderately severe right frontal headache of several hours' duration. The patient had spent his early childhood in the Ohio Valley area.

Except for the ocular findings, the physical examination was unremarkable. The best visual acuity was 20/25 right eye, 20/20 left eye. Tension by applanation was 16 mm Hg in both eyes. The pupillary responses and slit-lamp examination were normal. There was a right lower temporal field loss by confrontation and Goldmann perimetry (Fig. 1). Fundus examination of the right eye revealed a...
segmental nodular swelling at the nasal side of the optic disc (Fig. 2). The disc margins were blurred with overlying hyperaemia and small splinter haemorrhages. There was adjacent retinal oedema and mild engorgement of the retinal vessels. Examination with the Hruby lens revealed a few vitreous cells overlying the nodule. The macula was normal. Extensive indirect ophthalmoscopic examination revealed a normal peripheral retina. Examination of the fundus of the left eye revealed extensive, diffuse, peripapillary atrophy and scarring (Fig. 3). In addition, there were several equatorial, chorio-retinal, punched-out scars typical of presumed ocular histoplasmosis (Fig. 4). Visual field and macular examination of the left eye was normal. Fluorescein angiography revealed leakage of dye from the right disc in the arterio-venous phase (Fig. 5) and late staining (Fig. 6).

**FIG. 2** Acute nodular swelling of right optic disc at the nasal side with overlying haemorrhage

**FIG. 3** Diffuse peripapillary atrophy of left optic disc

**FIG. 4** One of several peripheral, chorio-retinal, punched-out scars seen in left eye

**FIG. 5** Fluorescein angiography of right disc, revealing leakage of dye in arterio-venous phase
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The patient was given one subtenon's injection of 40 mg of triamcinolone acetonide (Kenalog®) and followed closely by dilated fundus examinations and fundus photography.

Three weeks after the initial examination the disc swelling had decreased by 50 per cent and the haemorrhages had begun to resolve (Fig. 7). Eight weeks later there was little or no disc swelling, but a definite peripapillary atrophy was noted (Fig. 8). By 4 months there was marked peripapillary chorio-retinal scarring of the

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**Fig. 6** Fluorescein angiography of right disc showing late staining

All laboratory tests were normal except a histoplasmin intradermal skin test which was positive, with a 14 × 12 mm area of induration at 48 hours. Tine test for tuberculosis and coccidiosis intradermal skin test were negative.

**Fig. 7** Partial resolution of right optic disc swelling 3 weeks after initial examination

**Fig. 8** Complete resolution of right optic disc swelling at 8 weeks

**Fig. 9** Marked peripapillary atrophy of right optic disc 4 months after initial examination
right disc (Fig. 9) similar to that noted initially of the left disc. At 8 weeks, two faint equatorial punched-out scars were noted which became more distinctive over the next few weeks (Fig. 10).

At 8 months no new findings were noted except for early posterior subcapsular cataracts of which the right one was worse. Visual acuity was 20/20 in both eyes, but there was a persistence of the field loss in the right eye.

Discussion

No acceptable aetiological relationship between *Histoplasmosis capsulatum* and the characteristic findings of presumed ocular histoplasmosis has so far been established. The diagnosis of presumed ocular histoplasmosis is based mainly on the relationship between characteristic chorio-retinal findings and a positive histoplasmin skin test (Woods and Wahlen, 1960; Walma and Schlaegel, 1964). Reid, Scherer, Herbut, and Irving (1942) described one case of ocular involvement in the fatal form of human histoplasmosis, but unfortunately the eyes were not obtained at necropsy. Klintworth, Hollingsworth, Lusman, and Bradford (1973) recently described a fatal human case of disseminated histoplasmosis in which they identified *Histoplasma capsulatum* in choroidal lesions. Although the clinical findings were not typical of presumed ocular histoplasmosis, it nevertheless was the first time that the fungi has been identified in a human choroid. Numerous investigators have reproduced the peripheral chorio-retinal lesions in a variety of experimental animals; however, most of these studies involved direct intraocular injection of the organisms. Naturally acquired systemic histoplasmosis seems to be secondary to the inhalation of spores with subsequent haematogenous spread to susceptible organs such as the eye. Smith, O'Connor, Halde, Scalarone, and Easterbrook (1973) produced the choroidal lesions in rabbits by intra-carotid injection of the histoplasma organisms. The development of diffuse peripapillary atrophy was not one of the sequelae noted.

Peripapillary atrophy was first reported by Schlaegel and Kenney (1966), who noted it in 85 per cent of their cases. It is thought to be primarily due to a chorioiditis. The presentation of our patient with a typical nodular papillitis with overlying vitreous cells does not substantiate this theory yet the final clinical appearance of the disc was identical to that typically seen. In certain cases the peripapillary atrophy associated with presumed ocular histoplasmosis may be secondary to a retinitis rather than to a chorioiditis. Since the development of the peripapillary atrophy has not been described previously, this case may be helpful in determining the natural history of the disease. It suggests that, when examining eyes histopathogenically for histoplasmin organisms, a more careful search of the retina may be indicated.

Summary

A 38-year-old man from the Ohio Valley area presented to the Ophthalmology Clinic at a west coast hospital (USA) because of sudden loss of the right inferior temporal visual field after severe right frontal headache of several hours' duration. During the following months, diffuse peripapillary atrophy and peripheral punched-out lesions developed. When he had been seen initially, the fundus of the left eye had also revealed extensive, diffuse, peripapillary atrophy and scarring. All laboratory tests were normal except a histoplasmin intradermal skin test. The clinical appearance of both eyes 8 months after the acute episode in the right eye supports the diagnosis of presumed ocular histoplasmosis.

References