Ocular involvement in mycosis fungoides

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Abstract

A case of mycosis fungoides with bilateral intraocular involvement is reported. The diagnosis was established with the aid of a vitreous biopsy. Intraocular involvement, though rare, may be an early sign of extra-cutaneous disease.

(*Br J Ophthalmol* 1993; 77: 126–127)

A 61-year-old woman was referred to this unit in July 1989 with a 4 month history of visual loss, which had initially involved her right eye before it rapidly became bilateral. In June 1988 she was seen in another unit with rosacea keratitis which had, as a result of corneal scarring, reduced her visual acuity to 6/12 in both eyes.

In May 1984 (4 years before the onset of her visual symptoms) the patient had presented to a dermatologist with a itchy eczematous eruption. A presumptive diagnosis of mycosis fungoides was made, though a skin biopsy performed at that time appeared inconclusive. In 1987 a further skin biopsy demonstrated a T-cell infiltrate consistent with the diagnosis of mycosis fungoides.

On examination in July 1989 her visual acuities were hand movements in both eyes. Bilateral central stromal corneal scars were present. Numerous, large 'mutton fat' keratic precipitates, with aqueous cells and flare were observed in both eyes. The left iris was rubeotic and numerous posterior synechiae were present bilaterally. Funduscopy revealed dense vitreous opacities, numerous large creamy-white retinal infiltrates and multiple retinal haemorrhages in both eyes (Fig 1).

Numerous cutaneous papules and raised excoriated plaques were noted on the trunk and limbs. The patient was admitted for further investigations which included a left vitreous biopsy, a bone marrow trephine, and a thoraco-abdominal computed tomographic (CT) scan.

Examination of the vitreous aspirate revealed a monomorphic infiltrate of mononuclear cells with large, deeply indented, hyperconvoluted nuclei (Fig 2). Immunoperoxidase staining was

Figure 2 Photomicrograph of the vitreous aspirate. The cells have only scanty cytoplasm and contain a large, irregular, crenated nucleus (Papanicolaou stain; magnification $\times 530$).

positive for UCHL1 and negative for L26, which suggested that the cells were of T-cell lineage.

Histological sections from the skin biopsy obtained in 1987 were examined and found to contain a dense lymphocytic infiltrate throughout the dermis, extending into the epidermis and obliterating the dermoepidermal junction. The characteristic Pautrier 'microabscesses' of mycosis fungoides were present in the epidermis (Fig 3); some of these lymphocytes had large convoluted nuclei. The appearances supported the dermatological diagnosis of mycosis fungoides. Immunoperoxidase studies confirmed that the infiltrating lymphocytes were of T-cell origin (UCHL1 and MT1 antigen positive), and that B-lymphocyte markers (MB1) were negative.

The bone marrow trephine and aspirate showed increased normal erythropoiesis, but no evidence of lymphomatous infiltration. Thoracoabdominal CT scans were normal. A

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Accepted for publication
15 September 1992

Figure 1 Fundus photograph of the right eye. Retinal infiltrates and associated haemorrhages are present superior to the optic disc. The relatively poor photographic definition is due to the presence of dense vitreous infiltrates.

Figure 3 Skin biopsy 1987. There is a dense infiltrate of lymphocytes into the dermis and lower epidermis, obscuring the dermoepidermal junction. Pautrier 'microabscesses' of the lymphocytes (thick arrows) are present in the epidermis. Note the mitoses in dermal lymphocytes (thin arrows) (haematoxylin and eosin stain; magnification $\times 68$).

diagnosis of bilateral intraocular lymphoma secondary to mycosis fungoides was made. The patient was started on oral prednisolone (60 mg daily), methylprednisolone drops, and atropine to both eyes. This resulted in a marked improvement of her skin condition and a slight resolution of the vitreous opacities. A course of palliative radiotherapy was prescribed for both eyes (1500 cGy in 10 fractions to each eye) which resulted in some resolution of the retinal lesions in her right eye: the deposits in her left eye remained unchanged. The patient was reviewed in November 1989, when her visual acuities were counting fingers and perception of light respectively, and the fundal appearances were unaltered. The patient was then lost to follow up.

Comment

Mycosis fungoides is a malignant cutaneous T-cell lymphoma which usually runs a protracted course; it initially presents as a non-specific erythematous cutaneous eruption which progresses to form plaques and tumours. Eventually non-cutaneous involvement supervenes with infiltration of lymph nodes and visceral organs.¹ Ocular involvement may occur in up to a third of advanced cases and most frequently presents as tumours or infiltration of the eyelids.² Less frequent manifestations include involvement of the caruncle,³ the cornea,² the sclera,⁴ the optic disc,⁵ and optic nerve,⁶ and the orbit.⁷ Intraocular involvement has been reported only rarely.^{5,8,9} Keltner *et al*⁵ reported a single case of a 58-year-old male with a 4 year history of cutaneous mycosis fungoides. He presented with a swollen right optic disc and proceeded to develop bilateral retinal and vitreous infiltrates despite treatment. The patient died shortly after the development of the retinal lesions. There was no evidence of visceral involvement at autopsy. Postmortem histopathological examination of the globes revealed infiltration of the inner

retinal layers and vitreous with atypical lymphocytes. Recently, Erny *et al*⁹ reported a case of bilateral intraocular involvement in a 48-year-old male. He had suffered from mycosis fungoides for 30 years and died shortly after the development of the ocular symptoms from widespread disease. A postmortem examination revealed a lymphomatous infiltrate in the vitreous, and between Bruch's membrane and the retinal pigment epithelium.

Our case shares certain similarities with the above two reports: the patient had developed the disease several years before the onset of the ophthalmic manifestations; the disease involved the retina and vitreous in both eyes, and ocular involvement occurred in the absence of, or before the appearance, of widespread visceral disease. It would appear that, though intraocular infiltration is uncommon in mycosis fungoides, it may be an early manifestation of extracutaneous disease.

In this case the diagnosis was confirmed with the aid of a vitreous biopsy, which emphasises the importance of this technique in establishing the diagnosis in patients with suspected intraocular lymphoma.

We are grateful to Dr P B Gray for providing the histological sections from the 1987 skin biopsy.

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