Lymphoma – Primary intraocular

Reticulum cell sarcoma

DESCRIPTION

Lymphomas are lymphocytic neoplasms arising as discrete tissue masses. Primary central nervous system lymphoma (PCNSL) originates in the brain, spinal cord, leptomeninges, cerebrospinal fluid (CSF) or eyes. Primary intraocular lymphoma (PIOL) is a type of PCNSL in which only neural ocular tissue (namely, retina, vitreous or optic nerve) is involved, although more extensive CNS involvement is expected with disease progression. Alternatively, systemic lymphoma may cause metastases to the uvea.

PIOL is usually a diffuse large B-cell non-Hodgkin's lymphoma. It typically affects immunocompetent persons over age 50, or younger immunocompromised individuals. Historically, the diagnosis has often been reached after a presumptive case of uveitis has failed to respond completely to steroid therapy ('uveitis masquerade syndrome'). PIOL is usually bilateral.

SYMPTOMS

The most common symptoms are blurred vision and floaters. Other potential symptoms include photophobia, ocular pain and a red eye. Other neurological or systemic symptoms (for example, motor or sensory deficits, altered cognition, seizures, fevers or weight loss) suggest more extensive disease.

SIGNS

Anterior chamber signs are present in at least half of cases (for example, keratic precipitates, inflammatory or lymphomatous cells, or hypopyon). On fundoscopy, cells and debris often cloud the vitreous. The vitritis may have been present for some time and not responsive to corticosteroid treatment. Chorioretinal or sub-retinal pigment epithelial infiltrates often produce large, multifocal, cream to yellow-coloured diffuse patches with overlying pigmentary changes. Retinal oedema and haemorrhages, vascular sheathing or retinal detachment may exist. Thorough physical and neurological examination is required.

PREVALENCE

Very rare (less than 1/100,000). The majority of patients initially diagnosed with PIOL develop more extensive CNS disease within two to three years.

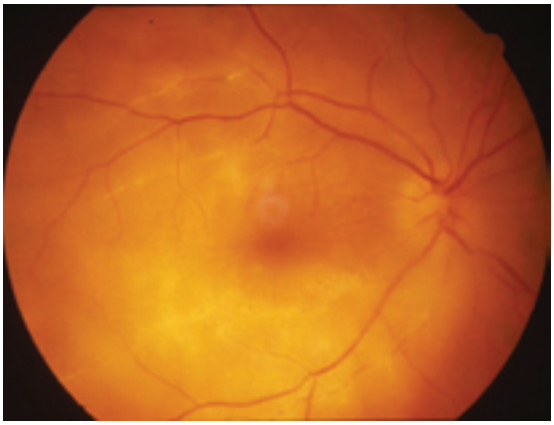

Figure 1 Retinal vascular lymphoma

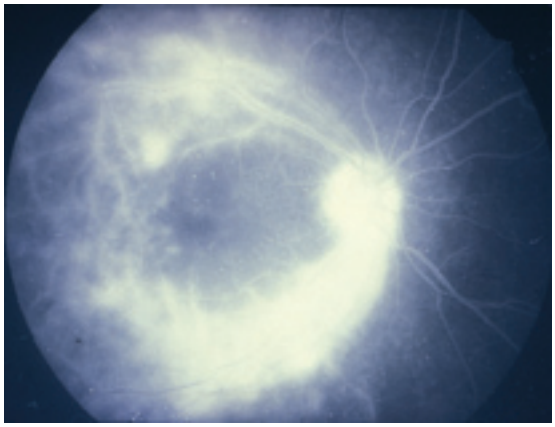

Figure 2 Fluorescein angiogram of the same patient

SIGNIFICANCE

PIOL is a malignant tumour that is lethal if untreated.

DIFFERENTIAL DIAGNOSIS

Posterior uveitis (multiple causes), Melanoma, Retinal detachment, Intraocular foreign body, Posterior scleritis, Leukaemia.

SEE ALSO

Acquired immunodeficiency syndrome (AIDS).

MANAGEMENT

Ocular tests, imaging investigations

- Computed tomography (CT) or preferably magnetic resonance imaging (MRI) of the brain are performed to delineate CNS involvement. CT imaging of other regions may also be indicated
- Ocular tests, including fluorescein angiography and ultrasound, may aid diagnosis.

Blood tests, microbiology, pathology

- Initial blood tests are performed to exclude differential diagnoses and detect evidence of systemic disease. These include complete blood examination and film, erythrocyte sedimentation rate and screening tests for vasculitis. In immunocompromised patients, viral antibody titres are often appropriate
- Lumbar puncture is performed for microscopy and cytology, biochemistry, flow cytology, immunohistochemistry, cytokine levels and detection of immunoglobulin gene rearrangements
- When the diagnosis remains unclear despite these tests, diagnostic vitrectomy or chorioretinal biopsy is performed
- Bone marrow or lymph node biopsy may be required to assess systemic disease.

Treatment

PCNSL with ocular involvement is often managed with systemic chemotherapy and radiotherapy in conjunction with ocular radiotherapy. PIOL may be treated similarly, due to the risk of subsequent CNS involvement.

Reference

Chan C, Wallace DJ. Intraocular lymphoma: Update on Diagnosis and Management, *Cancer Control*, 2004 11(5) 285-94.

The full series of these articles will be available in the book *Posterior Eye Disease and Glaucoma A-Z* by Bruce AS, O'Day J, McKay D and Swann P. £39.99. For further information click on the Bookstore at **opticianonline.net**

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