Retinoblastoma - management

DESCRIPTION, SIGNS, PREVALENCE, SIGNIFICANCE

See Retinoblastoma and leukocoria – assessment (*Optician*, May 30).

MANAGEMENT

Imaging investigations

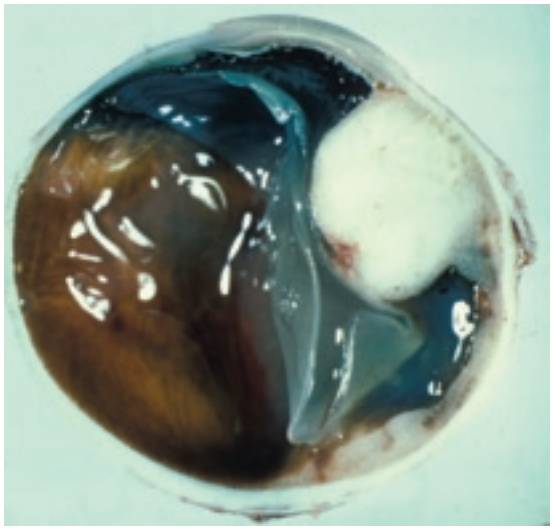
Various imaging tests (including B-scan ultrasound, computerised tomography, magnetic resonance imaging and fluorescein angiography) are used to exclude differential diagnoses, detect calcification, measure tumour dimensions and delineate extraocular extension or metastases. They are also required to visualise lesions obscured by vitreous opacification or retinal detachment.

Pathology

Further tests are required when the diagnosis is equivocal. For example, enzyme-linked immunosorbent assay (Elisa) is useful in the diagnosis of ocular toxocariasis. Anterior chamber paracentesis and/or fine-needle aspiration biopsy are occasionally performed, but carry the risk of facilitating dissemination of retinoblastoma. In some cases of advanced or metastatic retinoblastoma, further investigations including lumbar puncture, bone marrow aspiration and tumour markers are indicated.

Laser and incisional surgery

The management of retinoblastoma is complex. In many cases, radioactive plaques are sutured to the sclera adjacent to the tumour (brachytherapy). Tumours less than 3mm in greatest diameter, without vitreous seeding or optic nerve involvement, may be amenable to laser photocoagulation or transpupillary thermotherapy. Advanced tumours require enucleation, often with adjuvant chemotherapy and/or external beam radiotherapy. Risks of radiotherapy include retinal necrosis and radiation-induced tumours. Metastatic disease is treated with high-dose chemotherapy.



Bisected globe with multiple white tumour deposits

Prognosis

The overall mortality from retinoblastoma is 10 per cent. After enucleation, optic nerve involvement beyond the point of surgical transection entails 65 per cent mortality. Other adverse prognostic features include choroidal invasion, large tumour dimensions and poor cellular differentiation. Metastasis is rare. Other non-ocular primary tumours (such as, osteogenic sarcoma or pinealoblastoma) will occur in 10 per cent of patients.

Genetics

Genetic counselling is necessary, and genetic tests are often conducted. For unaffected parents with one affected child, there is a 5 per cent risk that a subsequent child will be affected. Patients with sporadic, unilateral retinoblastoma transmit the disease to approximately 10 per cent of subsequent children. Those with germline mutations have a 50 per cent chance of transmitting the mutation to each child.

The full series of these articles is 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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