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Intraocular Malignancy

1. GROSS DESCRIPTION

Specimen

— fine needle aspirate/local resection/evisceration/enucleation/exenteration.
— weight (g).
— anteroposterior, horizontal and vertical dimensions (cm).
  - length of optic nerve (mm)
  - ocular malignancy usually presents as an alteration in visual acuity and is rarely aspirated or biopsied due to potential tumour seeding. Unless small and anteroequatorial when a local resection may be considered, treatment is usually by enucleation (removal of the globe with a short piece of optic nerve) or exenteration if significant extraocular spread is present. Exenteration is more usually reserved for extraocular malignancy, e.g. of eyelids and orbital contents.

Tumour

Site

— bulbar conjunctiva/sclera/cornea: malignant melanoma, lymphoma, squamous carcinoma.
— iris/ciliary body/choroid: uveal melanoma.
— retina/optic nerve: retinoblastoma.
— anterior chamber/posterior chamber: posterior or equatorial—superior, inferior, lateral. Posterior lesions are better prognosis than equatorial as they interfere with visual acuity and present earlier, whereas the latter can attain a larger size with potential for involvement of scleral and Schlemm's canal vessels.

Size

— length × width × depth (mm) but in particular maximum tumour dimension (mm).

Appearance

— nodular/plaque/diffuse/multicentric/pigmented/non-pigmented/haemorrhage/necrosis/calcification.

Edge
— circumscribed/irregular.

2. HISTOLOGICAL TYPE

Malignant melanoma
— 80% in the choroid and the commonest intraocular malignancy in adults. It elevates and detaches the overlying retina.

Retinoblastoma
— <3 years of age and 40% familial, of which 90% are bilateral; retinoblastoma suppressor gene (Rb) 13q14 deletion.

Metastatic carcinoma
— breast, lung, gastrointestinal tract (stomach).
— 10% incidence at autopsy in carcinomatosis.
— posterior choroid is the commonest site.

Leukaemia/lymphoma
— 50% of leukaemia patients at autopsy (infiltration and/or haemorrhage).
— lymphoma is usually secondary to extraocular disease.

Rare
— medulloepithelioma, glioma, meningioma of optic nerve.

3. DIFFERENTIATION

Malignant melanoma
— spindle cell better prognosis
  • slender cells, indistinct nucleolus, longitudinal fold in the nuclear membrane (spindle cell type A)
  • nucleolar enlargement (spindle cell type B) is an adverse prognostic sign.
— epithelioid cell worse prognosis.
— mixed (50%).
— S100, HMB-45, melan-A positive, ± CAM5.2.

Retinoblastoma
— small round cells with basophilic nuclei and scant cytoplasm, calcification, mitoses and Homer Wright rosettes.
— well differentiated: fleurettes and Flexner-Wintersteiner rosettes.
— poorly differentiated: vascular pseudo-palisading necrosis, mitoses, apoptosis, absence of rosettes.
— S100, NSE, synaptophysin, GFAP positive, high Ki-67 index.

4. EXTENT OF LOCAL TUMOUR SPREAD
Border: pushing/infiltrative.
Lymphocytic reaction: prominent/sparse.