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Extraocular Malignancy

1. GROSS DESCRIPTION

Specimen

— fine needle aspirate/excision biopsy/exenteration.

- size (cm) and weight (g).
- extraocular tumours can present as an obvious nodule or plaque (eyelid, conjunctiva), swelling (lacrimal apparatus), proptosis or exophthalmos (orbital). Tissue diagnosis and treatment of eyelid and conjunctiva lesions usually involves primary local excision sufficient to remove the tumour but minimized to preserve function. Excision is by wedge (eyelid) or strip (conjunctiva) resection and these oculoplastic procedures may sometimes require intraoperative frozen section assessment of surgical margins. Exenteration is reserved for those tumours showing more extensive local spread demonstrated by CT or MRI scans. Pleomorphic adenoma of the lacrimal gland is usually amenable to local excision via a lateral orbitotomy but like adenoid cystic carcinoma may require more extensive surgery. Initial investigation of orbital tumours dictates planned management with avoidance of surgical excision of irresectable primary malignancies and metastatic tumours. Tissue diagnosis may be obtained by FNA or punch biopsy but often the latter is insufficient and a formal, deep, open biopsy via a lateral orbitotomy is required. Clinical assessment should consider various non-surgical possibilities, e.g. metastatic carcinoma, malignant lymphoma, thyrotoxicosis and Wegener's granulomatosis. Surgery is geared towards excision of localized primary tumours, e.g. cavernous haemangioma, pleomorphic adenoma, but exenteration, involving removal of the eye, surrounding orbital contents, eyelids, nasolacrimal apparatus ± orbital bone, may be required. The commonest indications for exenteration are malignant tumours of the eyelid such as basal cell, squamous cell or sebaceous carcinomas.

Tumour

Site
— ocular adnexae: eyelid
conjunctiva
lacrimal apparatus.
— orbit/retro-orbital tissues.

Size
— length × width × depth (cm) or maximum dimension (cm).

Appearance
— exophytic/verrucous/sessile/ulcerated/fleshy/infiltrative/pigmented.

Edge
— circumscribed/irregular.

2. HISTOLOGICAL TYPE

Adnexae
— basal cell carcinoma: most common, >80%.
— squamous cell carcinoma: 5–10%.
— sebaceous carcinoma: epithelioma/carcinoma.
— Merkel cell carcinoma: NSE/chromogranin/synaptophysin/cytokeratin (CAM5.2, CK20) positive; aggressive.
— malignant melanoma: primary (de novo or origin in eyelid/conjunctional naevus, or acquired conjunctival melanosis) or secondary.
— lymphoma: low-grade MALToma with indolent behaviour.
— metastatic carcinoma: breast, gut, lung.
— lacrimal gland tumours: e.g. pleomorphic adenoma, adenoid cystic carcinoma and other salivary type neoplasms.

Orbit: children
— embryonal (less commonly alveolar) rhabdomyosarcoma, Burkitt's lymphoma.

Orbit: adults
— haemangioma, neurilemmoma, lipoma, nodular fascitis, Langerhans cell histiocytosis.
— inflammatory pseudotumour: 20–40 years of sudden onset and painful but potentially steroid responsive.
— lymphoma (MALToma): the presence of lymphoid tissue in the orbit (not usual) is suspicious of neoplasia and up to 50% are part of systemic disease.
— haemangiopericytoma.
— fibro-/osteo-/chondro-/liposarcoma, malignant fibrous histiocytoma, alveolar soft part sarcoma, malignant teratoma are all rare, but