CHAPTER 13 TUMOURS OF THE EYE AND ORBIT

TUMOURS OF THE LIDS

A. Skin

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Basal cell carcinoma is most common (90 per cent) squamous cell less so, malignant melanoma rare.

Treatment: Excision, chemotherapy or radiotherapy

The problem is different from other skin areas in that the function of the lids must be maintained.Commonest malfunctions resulting from improper therapy are:

1. Exposure keratitis - corneal ulceration and sometimes loss of the eye
2. Excessive watering from eversion of the punctum.
3. Irregularities of the lid margin and lashes, producing corneal ulceration.

Small lesions may be treated successfully by any method.

Larger lesions, involving most or all of the lid are usually best treated by surgical excision and primary plastic repair.

B. Meibomian glands

This is occasionally the site of an adenocarcinoma, and therefore routine histological study of all chronic and recurrent 'chalazia' is advisable.

TUMOURS OF THE ANTERIOR SEGMENT:

(SUMMARY - SEE EARLIER SECTIONS FOR DETAILS)

A. Conjunctiva

1. Naevus - pigmented
2. Malignant melanoma
3. Squamous cell carcinoma

B. Iris

1. Naevus - pigmented
2. Malignant melanoma of iris and/or ciliary body

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TUMOURS OF THE POSTERIOR SEGMENT:

Whereas lesions of the anterior segment are usually noticed by the patient or his family while they are still quite small and before they interfere with vision, tumours of the posterior segment are usually hidden from view to the casual observer, and their presenting symptoms are usually:

1. In adults, impairment of vision.
2. In children:

    a) A white reflex in the pupil (Leucocoria)
    b) Strabismus
    Loss of acuity results in loss of fusion, therefore the eye deviates. It is important to rule out retinoblastoma in every child with a monocular squint.

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RETINOBLASTOMA

Retinoblastoma (most common malignant intraocular tumour in children - 1:20,000 live births).

- Present at birth, but usually no symptoms until age one to four years.

- About 25 per cent of cases are bilateral.

There are two types of growth:

1. Exophytic - proliferates from retina into the the choroid, optic nerve and brain.

2. Endophytic - proliferates into the vitreous, and may reach the anterior chamber.

Genetics:

1. Familial incidence - 6 per cent

2. Spontaneous mutation - 94 per cent

It is the only malignant tumour in man controlled by a dominant gene.

Treatment:

1. Enucleation if ocular involvement is extensive. This may be followed by systemic chemotherapy in advanced cases and in the presence of metastases.

2. Radiotherapy - External beam irradiation for medium or large tumours.
    - Cobalt 90 plaque (sutured to the sclera) for small to medium-sized

tumours and for those that do not respond to external irradiation.

3. Xenon arc photocoagulation for small tumours not involving the optic nerve or macula.

4. Cryotherapy is useful for the treatment of small peripheral tumours.

MALIGNANT MELANOMA

This is the most common malignant intraocular tumour in man.

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- usually occurs in adults
- malignancy varies
- commonly confused with
  a) idiopathic retinal detachment
  b) benign choroidal naevi
  c) Metastatic choroidal tumours - eg. from breast, lung.

Treatment

Treatment of a large malignant melanoma of the choroid is almost always enucleation. Some recent surgical innovations suggest that local excision is occasionally feasible. Survival following this procedure is usually good providing there is not extrabulbar extension. Small melanomas may be relatively benign and grow very slowly, and consequently may be careful observed.

TUMOURS OF THE ORBIT

Most orbital tumours produce proptosis.

When confronted with a patient with proptosis, the following should be considered.

1. Orbital tumour (see below).

2. Tumours involving the apex of the orbit, i.e. sphenoidal ridge meningoma, nasopharyngeal carcinomas.

3. Endocrine exophthalmos.

4. Pseudotumour of orbit.

5. Circulatory disturbance

   a) Carotid - cavernous sinus fistula
   b) Cavernous sinus thrombosis.

6. Orbital cellulitis.

7. Xanthomatoses e.g. Hand-Schuller-Christian disease.

8. Large eye - myopia, buphthalmos (congenital glaucoma).

9. Small orbit (cranio-stenosis).

ORBITAL TUMOURS:

- are usually unilateral
- show no inflammatory signs
- are slowly progressive
- have x-ray signs e.g. changes in bone density, calcification, enlargement of superior orbital fissure or optic canal.

<u>Types of orbital tumour</u>

A. In children:

1. Haemangioma
2. Rhabdomyosarcoma - most common primary malignant orbital tumour in children
3. Glioma of the optic nerve - can be associated with von Recklinghausen's disease
4. Retinoblastoma with orbital spread
5. Dermoid
6. Neuroblastoma - the most common metastatic orbital tumour in children.

B. In adults:

1. Haemangioma
2. Mucocoele
3. Meningioma
4. Lymphoma
5. Fibroma, osteoma, etc.
6. 2^{o} spread from tumour of nose or sinuses
7. Melanoma 2^{o} spread from choroid or lids

8. Lacrimal gland tumour.

Much emphasis has recently been placed on sophisticated techniques, which materially aid in the diagnosis of orbital lesions. These include:

1. Tomography
2. Subtraction x-ray techniques
3. Orbital venography (contrast dye injected into the frontal veins outlines the veins of the orbit. Displacement of the orbital veins (which normally have a fairly constant orientation) is good evidence of a space occupying lesion. Good for vascular lesions.
4. CT scans of the orbit.
5. Ultrasound scans.

Surgery

Excellent access to the structures of the orbit can be obtained by removing the lateral wall (Kronlein's procedure) or the roof (Naffzeiger's procedure). This latter procedure requires an intracranial approach, and is thus a much more major and serious procedure.