

# Ocular Oncology

**Qualifications : MBBS, MS/MD/DNB/DO/DOMS**

**Observership – One to three months duration.**

#### **Basic Level Goals:**

Year 1 equivalent: trainee ophthalmologist, any grade, not expecting to specialize in ocular oncology.

##### **A. Cognitive Skills**

1. Describe the basic categorization of common conjunctival and intraocular tumors.\*\*
2. Describe the clinical features of the major types of ocular tumor.\*\*
3. Describe the symptoms and clinical manifestations indicating the presence of an ocular tumor (eg, leukocoria, sentinel vessels).\*\*
4. Describe the differential diagnosis of the major tumors.\*\*
5. Describe the examinations and tests by which ocular tumors are diagnosed.\*\*
6. Describe the systemic features of ocular tumors and how these features are detected.\*\*
7. Describe the basic management principles of ocular tumors.\*\*
8. Describe the epidemiology of the more common tumors (eg, melanoma).\*\*
9. Describe the methods, risks, and benefits of tumor biopsy.\*\*

#### **Standard Level Goals:**

Year 2 equivalent: senior general ophthalmologist who may need to diagnose and refer patients with an ocular tumor and collaborate with an ocular oncologist in long-term aftercare.

##### **A. Cognitive Skills**

1. Describe the classification of ocular tumors (ie, conjunctival and intraocular).\*\*
2. Describe the clinical features of ocular tumors and their secondary effects.\*\*
3. List the differential diagnosis for each of the ocular tumors.\*\*
4. Describe diagnostic techniques for ocular tumors (eg, examination under anesthesia for pediatric

tumors, imaging, biopsy, laboratory tests, oncology referral).\*\*

5. Describe indications (eg, biopsy for lymphoma) and contraindications (eg, biopsy for retinoblastoma) for the various diagnostic techniques.\*\*
6. Describe the management options for ocular tumors with indications and contraindications for each form of management.\*\*
7. Describe the complications of ocular therapy and their management.\*\*
8. Describe basic histopathology of tumors, including immunohistochemistry.\*\*
9. Describe the prognosis of the different types of ocular tumor.\*\*
10. Describe the epidemiology of the more common tumors (eg, melanoma).\*\*
11. Describe the methods, risks, and benefits of tumor biopsy.\*\*

#### **Advanced Level Goals:**

##### **A. Cognitive Skills**

1. Describe the applied surgical anatomy, histology, and physiology of the eye and ocular adnexa with relevance to ocular oncology.
2. List the most common conjunctival and intraocular tumors.\*\*
3. Describe relevant pathological conditions, such as:
  - a. Nonneoplastic tumors (eg, hamartomas)\*\*
  - b. Neoplastic tumors\*\*
    - i. Benign (eg, nevus, hemangioma)
    - ii. Malignant (eg, melanoma, carcinoma, metastasis)
  - c. Traumatic lesions (eg, implantation cysts, hemorrhages)\*\*
  - d. Degenerative lesions (eg, disciforms, sclerochoroidal calcification)\*\*
  - e. Idiopathic disease (eg, juvenile xanthogranuloma, vasoproliferative tumor)\*\*
  - f. Paraneoplastic disease (eg, Bilateral diffuse uveal melanocytic proliferation)\*\*
  - g. Iatrogenic disease (eg, radiation-induced disease)\*\*
4. Describe relevant pathological techniques (eg, fixation, histology, immunohistochemistry).
5. Describe relevant genetic abnormalities and techniques:\*\*

- a. Germinal and somatic mutations relevant to oncology (eg, retinoblastoma)\*\*
- b. Important genetic techniques (eg, fluorescence in situ hybridization)\*\*
6. Describe the relevance of staging tumors (eg, TNM [Tumor, lymph Nodes, Metastasis] Classification of Malignant Tumors).
7. Describe the etiology of ocular tumors, such as:
  - a. Environmental factors (eg, conjunctival squamous cell carcinoma)
  - b. Genetic factors (eg, retinoblastoma)
  - c. Syndromes (eg, von Hippel-Lindau disease)
  - d. Malformations (eg, choroidal osteoma)
8. Describe the pathogenesis of ocular tumors (ie, how tumors cause harm):\*\*
  - a. Ocular effects (eg, neovascular glaucoma)\*\*
  - Systemic effects (eg, metastatic disease)\*\*
9. Describe the epidemiology of the more common ocular tumors (eg, melanoma).\*\*
10. Describe the principles of examination techniques:
  - a. Inspection
  - b. Transillumination
  - c. Color photography
  - d. Optical coherence tomography
  - e. Autofluorescence
  - f. Angiography (indocyanine green and fluorescein)
  - g. Ultrasonography
  - h. Magnetic resonance imaging
  - i. Computerized tomography
  - j. Positron emission tomography
  - k. Biopsy
    - i. Aspiration
    - ii. Incisional
    - iii. Excisional
    - iv. Impression cytology
- l. Systemic investigation according to ocular tumor diagnosis
  - i. History
  - ii. Clinical examination
  - iii. Hematology and biochemistry
  - iv. Radiography
  - v. Ultrasonography
  - vi. Computerized tomography
  - vii. Magnetic resonance imaging
  - viii. Genetic testing
11. Describe the clinical features of each tumor type:\*\*
  - a. Inspection/color photography\*\*
  - b. Investigational (ie, angiography, echography)\*\*
12. List the differential diagnosis of each tumor, and describe the investigational approach for each condition.\*
13. Describe how the following therapeutic modalities and their effects are relevant to ocular tumors:\*\*
  - a. Radiotherapy (eg, brachytherapy, external beam radiotherapy, proton beam)\*\*
  - b. Chemotherapy (eg, topical, intraocular, systemic)\*\*
  - c. Phototherapy (eg, photocoagulation, photodynamic therapy)\*\*
  - d. Cryotherapy (eg, liquid nitrogen, carbon dioxide)\*\*
  - e. Surgical resection (eg, local resection, enucleation)\*\*
14. Describe how statistics can be applied to ocular oncology (eg, survival analysis).
15. Describe the methods, risks, and benefits of tumor biopsy and how these can be avoided (eg, biopsy of retinoblastoma, incisional biopsy of conjunctival tumor).\*\*

### Very Advanced Level Goals: Subspecialist

Subspecialist equivalent: senior ophthalmologist responsible for ocular oncology, either part-time or full-time, who receives ocular oncology patient referrals.

#### A. Cognitive Skills

1. Describe the applied surgical anatomy, histology, and embryology of the eye and ocular adnexa with relevance to ocular oncology.
2. Describe the applied physiology of the eye and adnexa with relevance to ocular oncology.
3. Describe the applied pathology of the following:\*\*
  - a. Ocular tumors and pseudotumors\*\*
    - i. Congenital/developmental
- 1.1. Conjunctiva
- a. Dermoid
  - b. Dermolipoma
  - c. Choristoma (simple and complex)

- 2.1. Uvea
  - a. Lisch nodules
  - b. Stromal iris cyst
  - c. Lacrimal gland choristoma
- 3.1. Retina
  - a. Multiple congenital hypertrophy of the retinal pigment epithelium (CHRPE)
  - b. Astrocytic hamartoma
  - c. Hemangioblastoma
  - d. Cavernous angioma
  - e. Dominant exudative vitreoretinopathy
  - f. Norrie disease
  - g. Incontinentia pigmenti
  - h. Solitary CHRPE
  - i. Grouped pigmentation
  - j. Arteriovenous malformation (racemose angioma)
  - k. Posterior primary hyperplastic vitreous (PPHV)
  - l. Glioneuroma
- ii. Inflammatory (infectious, noninfectious)
  - I.1. Conjunctiva
    - a. Granuloma (eg, syphilis, sarcoid)
- 2.1. Uvea
  - a. Granuloma (eg, tuberculosis)
  - b. Uveal effusion
  - c. Posterior scleritis
- 3.1. Retina
  - a. Granuloma (eg, toxocara)
- iii. Neoplastic
  - I.1. Benign
    - a. Conjunctiva
      - i. Nevus
      - ii. Papilloma
      - iii. Oncocytoma
      - iv. Primary acquired melanosis
      - v. Reactive lymphoid hyperplasia
      - vi. Other
    - b. Uvea
      - i. Nevus/melanocytoma
      - ii. Hemangioma
      - iii. Osteoma
      - iv. Neurilemmoma
      - v. Neurofibroma
      - vi. Leiomyoma
      - vii. Mesectodermal leiomyoma
      - viii. Reactive lymphoid hyperplasia
      - ix. Bilateral diffuse uveal melanocytic proliferation
      - x. Other rare conditions
  - c. Retina
    - i. Retinoma/retinocytoma
    - ii. Adenoma
    - iii. Fuchs adenoma
    - iv. Benign medulloepithelioma
    - v. Other
- 2.1. Malignant
  - a. Conjunctiva
    - i. Melanoma
    - ii. Squamous cell carcinoma
    - iii. Sebaceous carcinoma
    - iv. Kaposi sarcoma
    - v. Lymphoma
    - vi. Extraocular tumor spread
    - vii. Metastasis
    - viii. Other
  - b. Uvea
    - i. Melanoma
    - ii. Lymphoma
    - iii. Intraocular tumor spread from conjunctiva
    - iv. Systemic lymphoma
    - v. Systemic leukemia
    - vi. Metastasis
    - vii. Other
  - c. Retina
    - i. Retinoblastoma
    - ii. Adenocarcinoma
    - iii. Malignant medulloepithelioma
    - iv. Lymphoma
    - v. Leukemia
    - vi. Metastasis
    - vii. Other
    - iv. Traumatic
- I.1. Conjunctiva
  - a. Implantation cyst
  - b. Foreign body granuloma
  - c. Pyogenic granuloma
- 2.1. Uvea
  - a. Implantation cyst
  - b. Choroidal hemorrhage
  - c. Miotic cyst
- 3.1. Retina
  - a. Retinopathy of prematurity
  - b. Retinal detachment
  - c. Massive reactive gliosis
  - v. Degenerative

- 1.1. Conjunctiva
    - a. Lacrimal retention cyst
  - 2.1. Uvea
    - a. Disciform lesion
    - b. Sclerochoroidal calcification
    - c. Vortex vein ampulla
  - 3.1. Retina
    - a. Vasoproliferative tumor
  
  - vi. Idiopathic
    - 1.1. Conjunctiva
      - a. Lymphangiectatic cyst
    - 2.1. Uvea
      - a. Juvenile xanthogranuloma
    - 3.1. Retina
      - a. Coats disease
      - b. Combined hamartoma of retina and retinal pigment epithelium
      - c. Iris cyst
      - d. Ciliary epithelial cyst
  - vii. Paraneoplastic disease
    - 1.1. Bilateral diffuse uveal melanocytic proliferation
    - 2.1. Carcinoma-associated retinopathy
    - 3.1. Melanoma-associated retinopathy
    - 4.1. Other
4. Describe the following pathological conditions:\*\*
- a. Non-neoplastic tumors\*\*
    - i. Hamartoma
    - ii. Choristoma
    - iii. Granuloma
    - iv. Cyst
    - v. Hyperplasia
    - vi. Metaplasia
  - b. Neoplastic tumors\*\*
    - i. Benign
    - ii. Malignant
      - 1.1. Proliferation
      - 2.1. Invasion
      - 9.1. Seeding
      - 4.1. Metastasis
    - iii. Iatrogenic disease
      - 1.1. Radiation
      - 2.1. Pharmacology
      - 3.1. Surgery
      - 4.1. Phototherapy
- 5. Describe relevant pathological techniques, such as:
    - a. Fixatives\*\*
    - b. Frozen sections
    - c. Histology
    - d. Immunohistochemistry
    - e. Flow cytometry
    - f. Other
  - 6. Describe the following genetic abnormalities and techniques:
    - a. Germinal mutations relevant to oncology\*\*
    - b. Somatic mutations in tumors\*\*
    - c. Genetic techniques
      - i. Karyotyping
      - ii. Polymerase chain reaction
      - iii. Fluorescence in situ hybridization
      - iv. Multiplex ligation-dependent probe amplification
      - v. Gene expression profiling
      - vi. Comparative genomic hybridization
      - vii. Other
  - 7. Describe the relevant staging and grading systems for ocular tumors (with ability to use appropriate methods as necessary, using appropriate references sources):
    - a. TNM Classification of Malignant Tumors cancer staging system
      - i. Uveal melanoma
      - ii. Retinoblastoma
      - iii. Conjunctival melanoma
      - iv. Conjunctival carcinoma
      - v. Ocular adnexal lymphoma
    - b. International retinoblastoma staging system
    - c. Reese-Ellsworth staging system for retinoblastoma
    - d. Other staging systems (eg, Collaborative Ocular Melanoma Study)
  - 8. Describe the etiology of ocular tumors:
    - a. Environmental factors
    - b. Genetic factors
    - c. Syndromes
    - d. Malformations
    - e. Other
  - 9. Describe the pathogenesis of ocular tumors:\*\*
- a. Secondary effects of uveal melanoma\*\*
  - b. Secondary effects of retinoblastoma\*\*
  - c. Secondary effects of other tumors (eg, conjunctival tumors)\*\*

10. Describe the epidemiology of ocular tumors:
- Principles of epidemiology
11. Describe the principles of examination techniques:\*\*
- Inspection\*\***
    - Slit-lamp examination
    - Gonioscopy and 3-mirror examination
    - Ophthalmoscopy
  - Transillumination\*\***
    - Transpupillary Transscleral
  - Color photography\*\***
    - Standard ocular photography
    - Specialized cameras (eg, RetCam, Optos)
    - Autofluorescence photography
  - Angiography\*\***
    - Fluorescein angiography
    - Indocyanine green angiography
  - Ultrasonography\*\***
    - A-scan ultrasonography
    - B-scan ultrasonography  
(including high frequency)
    - Doppler ultrasonography
  - Magnetic resonance imaging\*\***
  - Computerized tomography\*\***
  - Positron emission tomography\*\***
    - Biopsy\*\***
    - Aspiration
    - Incisional
    - Excisional
    - Impression cytology
    - Systemic investigation according to ocular tumor diagnosis\*\*
    - History
    - Clinical examination
    - Hematology and biochemistry
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    - Ultrasonography
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    - Genetic testing
12. Describe the clinical features of each tumor type:\*\*
- Inspection/color photography\*\*
  - Investigational (ie, angiography, echography)\*\*
13. List the differential diagnosis of each tumor and describe the investigational approach for each condition.\*\*
14. Describe how the following therapeutic modalities and their effects are relevant to ocular tumors:\*\*
- Radiotherapy\*\***
    - Radiation
      - Radioactive sources (eg, iodine, ruthenium)
    - Types of radiation (eg, gamma, beta, proton)
  - Biological effects**
    - Chemotherapy\*\***
    - Phototherapy\*\***
    - Cryotherapy\*\***
    - Surgical resection\*\***
15. Describe how the following statistics can be applied to ocular oncology:
- Statistical correlations
    - Univariate
    - Multivariate
  - Survival statistics
    - Kaplan-Meier analysis
    - Cox analysis
    - Neural networks
    - Accelerated failure time
  - Bias
  - Power calculations
  - Other relevant statistical methods