Ocular Oncology

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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.**

B. Technical/Surgical Skills

1. Perform slit-lamp and ophthalmoscopic examination of patients with an ocular tumor.**
2. Recognize an ocular tumor and refer to an ocular oncology subspecialist.**
3. Contribute to the care of patients after treatment.**

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.**

B. Technical/Surgical Skills

1. Perform naked-eye examination (eg, to recognize oculodermal melanosis).**
2. Perform palpation of cervical lymph nodes.**
3. Perform slit-lamp examination, gonioscopy, and indirect ophthalmoscopy to diagnose and localize ocular tumors.**
4. Perform transillumination for intraocular tumors.**
5. Perform B-scan ultrasonography to detect and measure intraocular tumors.**
6. Perform sequential examination to assess the tumor over time (eg, atypical nevus).**
7. Guide evaluation for systemic disease (eg, metastases, primary tumor, syndromes).**
8. Perform excision of conjunctival tumors, avoiding seeding, or refer to an ocular oncology subspecialist for such surgery if possible.**
9. Perform enucleation, obtaining long optic nerve if appropriate, or refer to a subspecialist for this surgery if necessary.**
10. Collaborate with subspecialist in the preoperative care and referral of selected patients with an ocular tumor, avoiding potential pitfalls.**
11. Provide short-term and long-term postoperative care to patients with an ocular tumor, collaborating with a subspecialist and other health care workers as appropriate.**

12. Investigate and manage ocular complications as appropriate (eg, radiation retinopathy, macular edema, cataract, glaucoma).**
13. Interpret the results of laboratory investigations and adjust management accordingly.**
14. Discuss prognosis and various management options with patients and their families in a detailed, ethical, and compassionate manner.**

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)**
   b. 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).**

B. Technical/Surgical Skills

1. Perform or request appropriate examinations and investigations according to differential diagnosis.**
2. Perform or refer for treatment for conjunctival or intraocular tumors, demonstrating awareness of the indications, contraindications, and complications of each treatment and having skill to administer short-term and long-term postoperative care:**
   a. Radiotherapy (eg, brachytherapy, external beam radiotherapy)**
   b. Phototherapy (eg, photodynamic therapy, transpupillary thermotherapy)**
   c. Surgical excision (eg, local resection, enucleation, exenteration)**
   d. Ocular pharmacological therapy by various routes (ie, topical, intravitreal, ophthalmic artery infusion, subtenon, systemic)**
      i. Chemotherapy and biological therapy
      ii. Antiangiogenic agents
      iii. Steroids
3. Interpret results of relevant laboratory tests and communicate results to patients, relatives, and health care workers; and adjust patient management accordingly.
4. Communicate prognosis with patients, relatives, and health care workers; and adjust patient management accordingly in collaboration, if necessary, with a subspecialist.**
5. Use information technology and other aids to cope with lack of expert knowledge.**
6. Assist patients with selecting the most appropriate management in collaboration, if necessary, with a subspecialist in ocular oncology.
7. Provide or organize appropriate psychological support, demonstrating empathy and an adequate awareness of the principles of this aspect of care (eg, giving bad news).**
8. Collaborate with subspecialists and other health care professionals to provide patient-focused care.**
9. Develop protocols and infrastructure for practice-based learning and improvement (eg, access to information, outcomes data).

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)
         1.1. 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
         1.1. 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

1.1. 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
3.1. 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:
   a. Principles of epidemiology

11. Describe the principles of examination techniques:**
   a. Inspection**
      i. Slit-lamp examination
      ii. Gonioscopy and 3-mirror examination
      iii. Ophthalmoscopy
   b. Transillumination**
      i. Transpupillary
      ii. Transscleral
   c. Color photography**
      i. Standard ocular photography
      ii. Specialized cameras (eg, RetCam, Optos)
      iii. Autofluorescence photography
   d. Angiography**
      i. Fluorescein angiography
      ii. Indocyanine green angiography
   e. Ultrasonography**
      i. A-scan ultrasonography
      ii. B-scan ultrasonography (including high frequency)
      iii. Doppler ultrasonography
   f. Magnetic resonance imaging**
   g. Computerized tomography**
   h. Positron emission tomography**
      i. Biopsy**
      i. Aspiration
      ii. Incisional
      iii. Excisional
      iv. Impression cytology
      j. 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

12. Describe the clinical features of each tumor type:**
   a. Inspection/color photography**
   b. 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:**
   a. Radiotherapy**
      i. Radiation
         1.1. Radioactive sources (eg, iodine, ruthenium)
         2.1. Types of radiation (eg, gamma, beta, proton)
      ii. Biological effects
      b. Chemotherapy**
      c. Phototherapy**
      d. Cryotherapy**
      e. Surgical resection**

15. Describe how the following statistics can be applied to ocular oncology:

a. Statistical correlations
   i. Univariate
   ii. Multivariate
b. Survival statistics
   i. Kaplan-Meier analysis
   ii. Cox analysis
   iii. Neural networks
   iv. Accelerated failure time
c. Bias
d. Power calculations
e. Other relevant statistical methods

B. Technical/Surgical Skills

1. Perform or request the following examinations, interpreting and documenting any findings, demonstrating awareness of the indications, contraindications, and limitations of each investigation:**
   a. Slit-lamp examination of conjunctiva and assessment of conjunctival fornices**
   b. Slit-lamp examination of anterior chamber and gonioscopy**
   c. Binocular indirect ophthalmoscopy with indentation**
   d. Transpupillary transillumination**
   e. A-scan and B-scan ultrasonography of anterior and posterior eye**
   f. Color and autofluorescence photography**
   g. Fluorescein angiography**
   h. Indocyanine green angiography**
   i. Magnetic resonance imaging**
   j. Incisional and excisional conjunctival tumor biopsy**
   k. Aspiration, incisional, or excisional biopsy of intraocular tumor**
   l. Other relevant examinations and investigations**
2. Perform or refer for the following treatments for conjunctival tumors, demonstrating awareness of the indications, contraindications, and complications of each treatment:**
   a. Surgical excision**
   b. Cryotherapy**
   c. Brachytherapy**
   d. External beam radiotherapy, including proton beam radiotherapy**
   e. Topical therapy (eg, mitomycin C, 5-fluorouracil, interferon)**
3. Perform or refer for the following treatments for intraocular tumors, demonstrating awareness of the indications, contraindications, and complications of each treatment:**
   a. Radiotherapy**
      i. Brachytherapy (eg, iodine, ruthenium, strontium, palladium, iridium)
      ii. External beam radiotherapy
      iii. Stereotactic radiotherapy
      iv. Charged particle radiotherapy (eg, proton beam)
   b. Phototherapy**
      i. Photocoagulation
      ii. Transpupillary thermotherapy
      iii. Photodynamic therapy
   c. Surgical excision**
      i. Iridectomy
      ii. Iridocyclectomy
      iii. Transscleral choroidectomy
      iv. Transretinal choroidectomy
      v. Enucleation
      vi. Exenteration
   d. Ocular pharmacological therapy by various routes (ie, topical, intravitreal, ophthalmic artery infusion, subtenon, systemic)**
      i. Chemotherapy and biological therapy
      ii. Antiangiogenic agents
      iii. Steroids
4. Request the following investigations, interpreting and communicating the results to patients, relatives, and health care workers, adjusting patient management accordingly:**
   a. Histopathological assessment of tumor samples**
   b. Genetic assessment of tumor samples**
   c. Laboratory investigation of vitreous samples**
   d. Other**
5. Estimate the prognosis and communicate the following implications with patients, relatives, and health care workers, adjusting patient management accordingly:**
   a. Visual acuity**
   b. Local tumor control**
   c. Possible side effects and complications of therapy**
   d. Ocular conservation**
   e. Systemic manifestations of disease, including metastasis**
   f. Systemic complications and side effects of therapy**
   g. Survival probability and chances of disease-related mortality**
   h. Heritability**
   i. Use information technology and other aids to enhance prognostication**
6. Communicate the following to patients, relatives, and health care workers:**
   a. Diagnosis, extent and severity of disease, including diagnostic uncertainty**
   b. Natural history without treatment**
   c. Therapeutic options with advantages and limitations of each therapy, including methods available elsewhere**

d. Logistical implications of selected treatment**
e. Use information technology and other aids to support this process**
   i. Websites
   ii. Printed leaflets
   iii. Audio recordings
f. Other relevant materials**

7. Assist patients with selecting the most appropriate management, taking into account:**
   a. Patient age, gender, culture, wishes, needs, and fears**
   b. Costs and logistics**
   c. Availability of health care resources, locally and elsewhere**

8. Provide or organize appropriate psychological support, demonstrating empathy and an adequate awareness of the principles of this aspect of care, such as:**
   a. Giving bad news**
   b. Communicating with relatives**
   c. Enabling long-term communication and support**

9. Develop and maintain a multidisciplinary team of health care professionals to provide patient-focused care by activities, such as:
   a. Recruiting staff and coworkers
   b. Developing service operating procedures.
   c. Maintaining efficient and varied methods of communication and education
      i. Between multidisciplinary team members (MDT)
      ii. Between MDT and other practitioners (eg, pathologists)
      iii. Between MDT and patient

10. Develop protocols and infrastructure for practice-based learning and improvement, including:
    a. Proformas and databases for storing data
    b. Protocols for extracting and analyzing data
    c. Application of study designs and statistical methods
    d. Adherence to clinical governance
       i. Informed consent
       ii. Confidentiality
       iii. Ethical committee approval