4.11.10. Retina & Retinal pigment epithelium (X): Neoplasia (II)

Fuchs Adenoma

- acquired tumor of nonpigmented ciliary body epithelium
- may cause sectoral cataract
- histology
  - hyperplastic nonpigmented ciliary epithelium
    - arranged in sheets and tubules
  - alternating areas of PAS-positive basement membrane material

Combined hamartoma of the retina and retinal pigment epithelium

- often diagnosed in childhood
- slightly elevated variably pigmented mass
- distorts the tumor inner retinal surface
- preretinal membrane
- thickening of optic nerve head and peripapillary retina
- histology
  - increased number of vessels
  - perivascular hyperplastic RPE
  - vitreous condensation and fibroglial proliferation

Adenoma/Adenocarcinoma of RPE

- differentiates from RPE hyperplasia
- uncommon
- no history of prior trauma or eye disease
- adenoma
  - basement membrane
  - cell junctions
  - microvilli
  - retain characteristics of RPE cells
- adenocarcinoma
  - greater anaplasia
  - mitotic activity
  - retinal or choroidal invasion
  - does not metastasize

Medulloepithelioma (Diktyoma)

- congenital neuroepithelial tumor
- arises from primitive medullary epithelium
- sites
  - ciliary body
    - most common
  - retina
  - optic nerve
- clinical
  - non- or lightly pigmented cystic mass
  - ± iris/anterior chamber erosion
- histology
  - undifferentiated round-oval cells with little cytoplasm
  - cell nuclei are stratified in 3-5 layers
  - ribbonlike structure with distinct cellular polarity
  - lined on one side by thin basement membrane
  - secretes mucinous substance rich in hyaluronic acid
    - resembles primitive vitreous
    - mucinous cysts
  - ± rosettes
    - Homer-Wright
    - Flexner-Wintersteiner

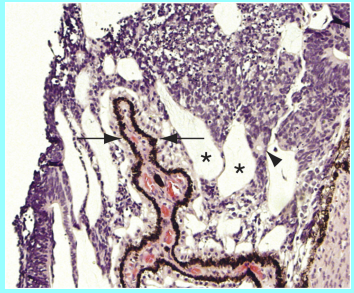

Figure 11-45 (© 2020 American Academy of Ophthalmology)