LEUKOCORIA

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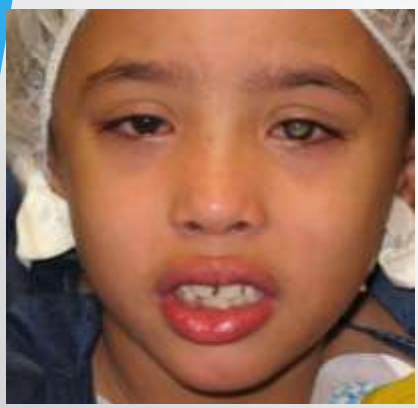
LEUKOCORIA



DD Of Leukocoria

- Retinoblastoma
- Coats' disease
- Persistent fetal vasculature (PFV)
- Toxocariasis
- Cellulitis
- Metastasis
- Cataract
- Coloboma
- Norrie's Disease
- Herpes Simplex
- Retinitis Cytomegalovirus
- Retinitis Toxoplasmosis
- Astrocytic hamartoma
- Retinopathy of prematurity
- Retinal detachment
- Combined hamartoma of the retinal pigment epithelium
- Myelinated nerve fiber

Leukocoria work up

- History (Trauma ,Prematurity , inflammation)
- Ocular examination:
  - Inspection (microphthalmia)
  - Lenticular or retrolental
  - IOP
  - **Fundoscopy and FA (RETCAM)**
  - OCT
  - Ultrasonography
  - MRI
- Systemic Evaluation

Retinoblastoma

Retinoblastoma (RB) is the most common intraocular malignancy of childhood and the most common cause of childhood leukocoria

Retinoblastoma

What is Retinoblastoma?

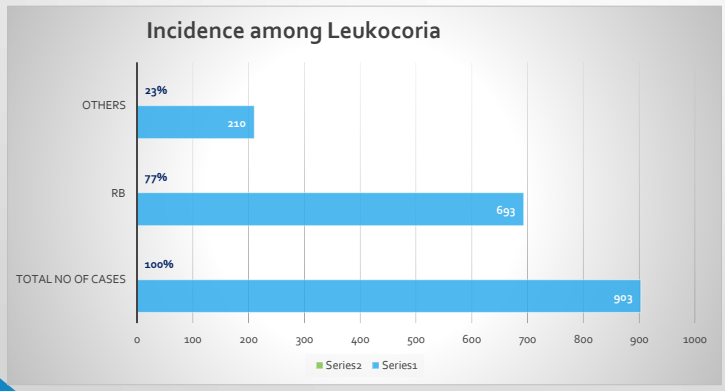
Malignant tumor of the Embryonic neural retina (fetal Retinoblasts) that normally differentiate into post-mitotic retinal photoreceptor cells and neurons.

Retinoblastoma

INCIDENCE

- 11% of Ped. Cancer in the 1st year but
- only 3% of all Ped. Cancers up to 15 years.
- 1/13000 birth
- Nearly 120 new case/year in Egypt.
- 250 case/year in USA.
- Increased in Africa – South America.

Retinoblastoma



RETINOBLASTOMA

AETIOLOGY

Genetic determined (2 Hits Hypothesis)

1st Step: Mutation of RB1 gene(13q14)

Tumour suppressor gene that regulates Cellular proliferation→

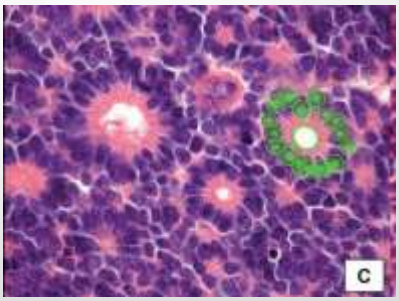
2nd Step: Anti Apoptotic Changes

Resulting in Decreased Cell death

RETINOBLASTOMA

PATHOLOGY

- The tumour is composed mainly of undifferentiated Anaplastic cells that arise from the nuclear layers of the retina.
- Aggregation around blood vessels, necrosis, calcification, and Flexner-Wintersteiner rosettes.
- Retinoblastomas are characterized by marked cell proliferations evidenced by high mitosis counts



RETINOBLASTOMA

CLINICAL PRESENTATION

-**Leukocoria**

-Squint

-Impaired vision

-Others:

Glaucoma-Hyphema- Uveitis - Orbital

Presentations of retinoblastoma

• Leukocoria - 60%

• Strabismus - 20%

• Secondary glaucoma

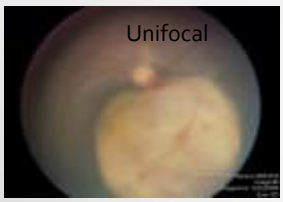
• Anterior segment invasion

• Orbital inflammation

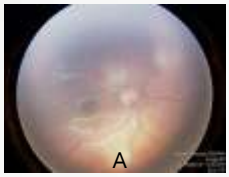
• Orbital invasion

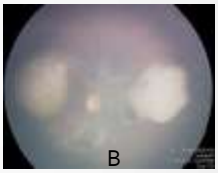
Classification and staging

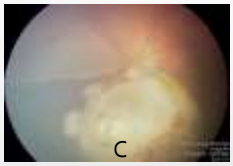
Unifocal Vs Multifocal

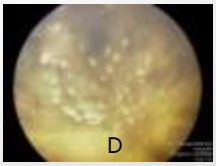


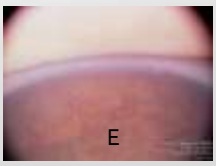
Retinoblastoma staging



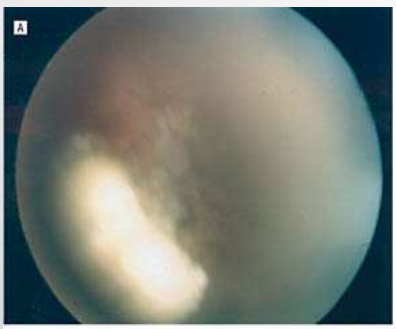


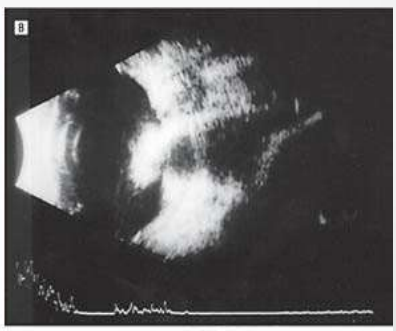


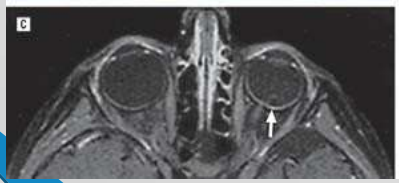


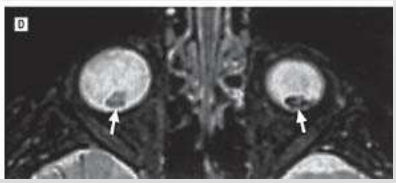


Ultrasonography and MRI Criteria

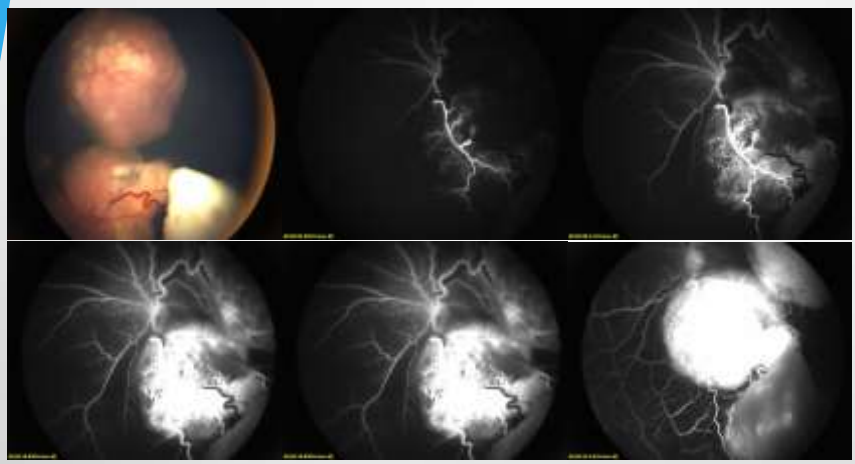








Fluorescin Angiogram Criteria

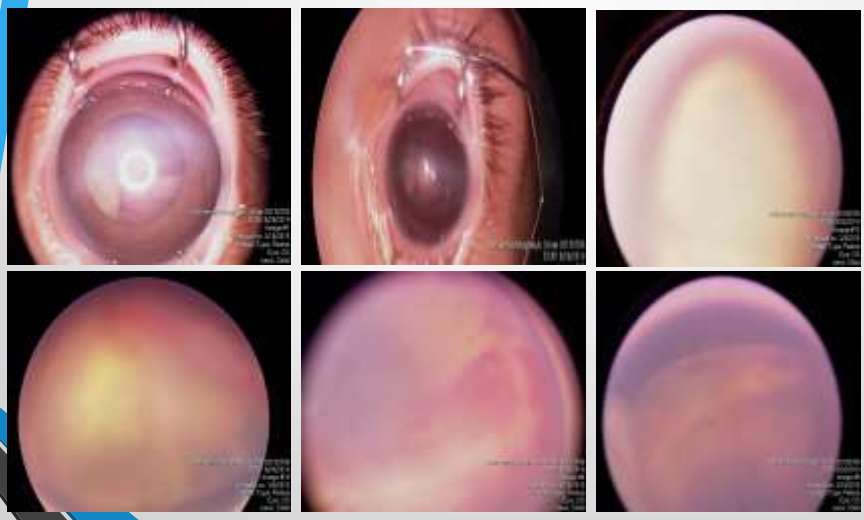


Intense hyperfluorescence and a fine vascular net- work within moderately sized tumors. Late phases of the angiogram show marked staining and leakage.

COAT,S

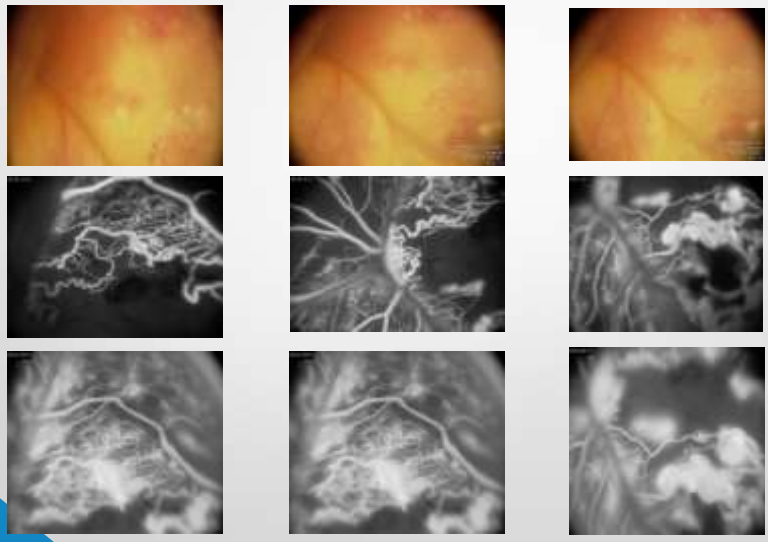
- Coats' disease is one of the most common lesions simulating retinoblastoma. This retinal vascular disorder is characterized by retinal telangiectasia, intraretinal exudation, and exudative retinal detachment
- This condition produces leukocoria and can sometimes be difficult to differentiate clinically from exophytic retinoblastoma
- MRI shows lack of enhancement of the subretinal space, absence of calcifications and high signal intensity on T2-W images due to high fat content.

COAT,S



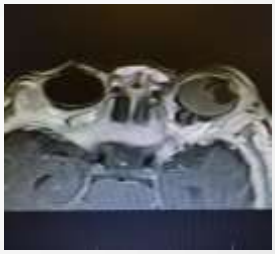
COAT,S

FA Criteria



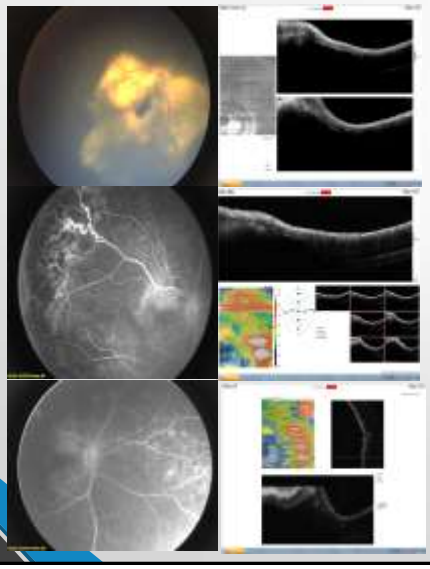
COAT,S
MRI Criteria

- Abnormal signals of vitreous with low signal intensity in T2 and high signals in T1
- Irregular areas of high signal intensity in T2

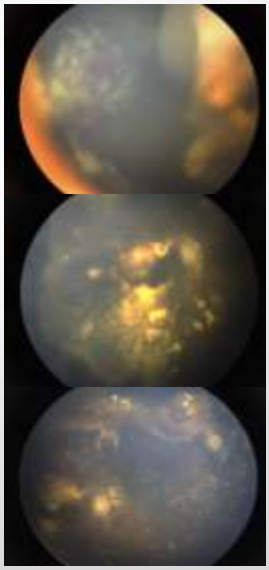


COAT,S prognosis

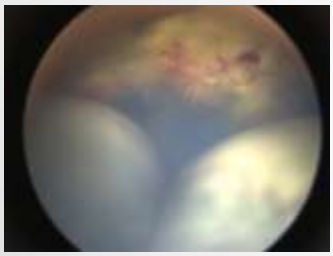
At diagnosis



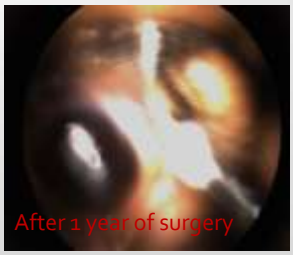
Post 6 ms of treatment



COAT,S prognosis







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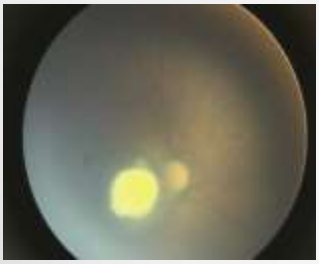
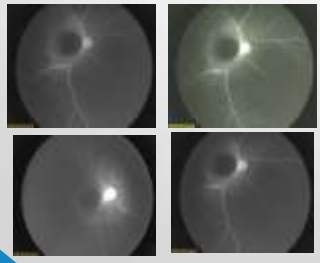
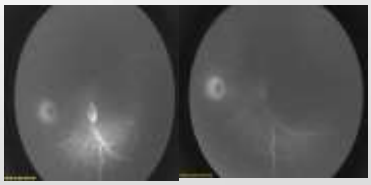
INFECTION AND INFLAMMATIONS

TOXOCARIASIS

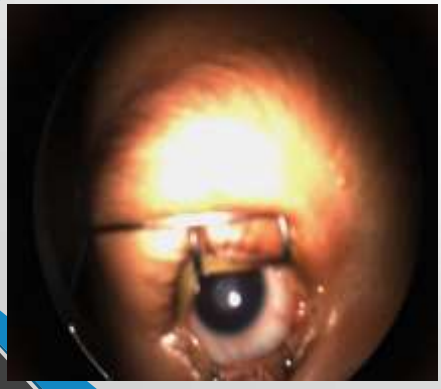
Premature infant
2ms of age

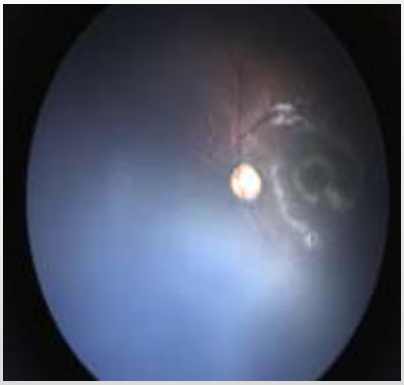






One month later
TORCH Negativa
HIV negative





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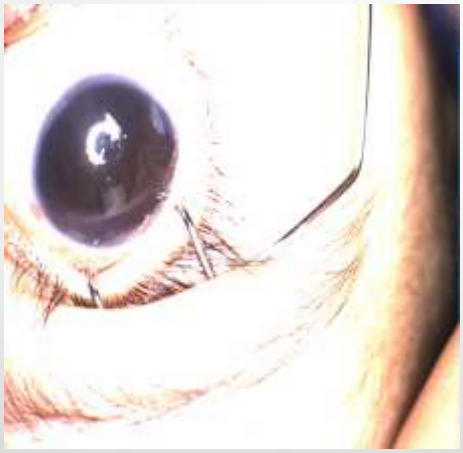
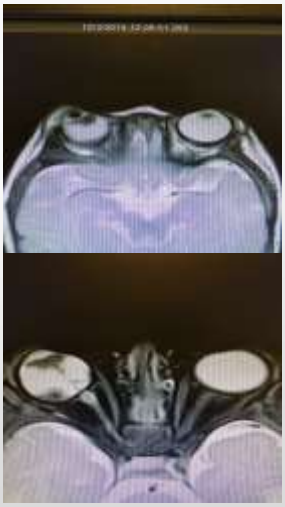
DEVELOPMENTAL VASCULAR ABNORMALITIES

PERSISTANT FEATAL VASCULATURE
PFV

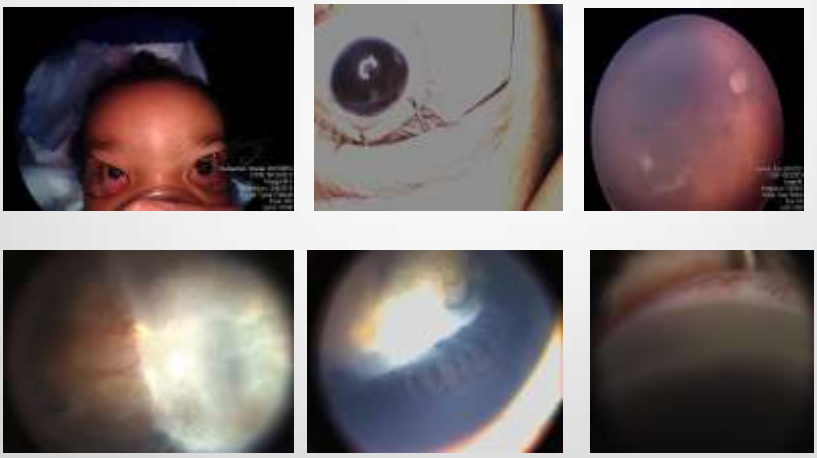


PHPV
MRI

- -The Vitreous shows bright T1 &T2 signals with no contrast enhancement
- -Retrolental soft tissue
- -Evidence of retinal detachment retinal hges
- -No definite retinal masses



PERSISTANT FEATAL VASCULATURE (PFV)



LEUKOCORIA

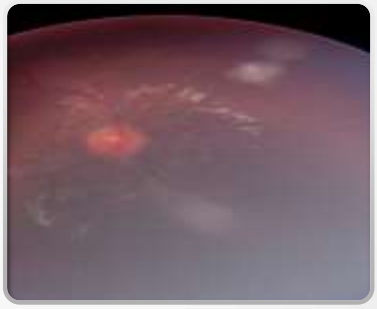
SYSTEMIC DISEASE

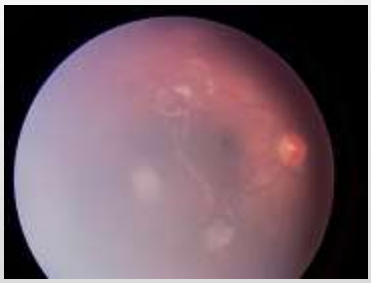
Astrocytoma

-Retinal astrocytic hamartomas are usually located in the inner layers of the retina or the optic disc, obscuring the retinal vessels. They have a milky gray–white appearance, depending on their amount of calcification .

-They are composed of spindle-shaped or even pleomorphic retinal astrocytes and can have various amounts of calcification

-On rare occasions retinal astrocytomas can show progressive growth and have associated features such as vitreous seeds, vitreous hemorrhage and exudative retinal detachment, which can simulate retinoblastoma





Astrosytoma

-Retinal astrocytic hamartoma or retinal astrocytomas are benign, acquired, retinal or papillary neoplasias, which are often found in association with the tuberous sclerosis complex

-They can also be associated with neurofibromatosis type 1 (NF-1), and can even rarely be found independently to any systemic diseases.

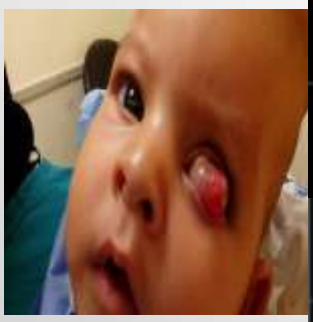
LEUKOCORIA

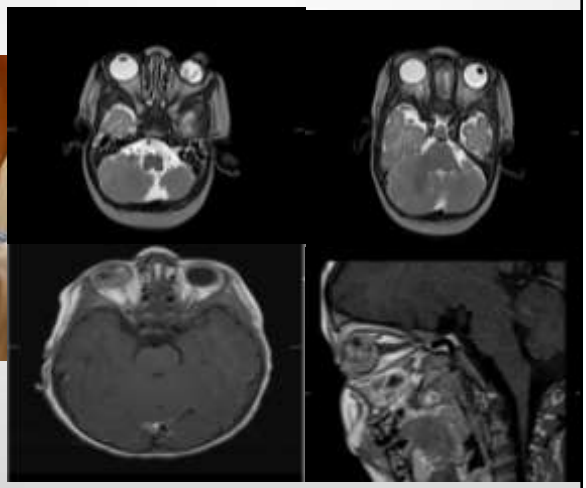
DEVELOPMENTAL MALFORMATIONS

Diktyoma

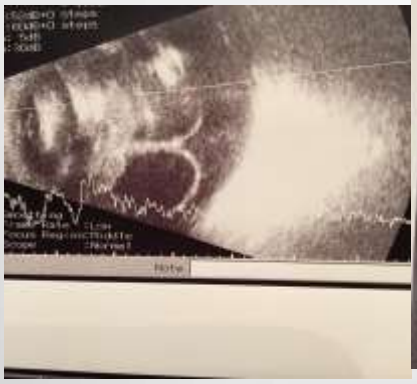
- It originates from undifferentiated nonpigmented epithelium of the ciliary body and most often presents as an iris mass during the first decade of life.
- Secondary glaucoma, hyphema, and ectopia lentis are less frequent initial manifestations.
- This rare lesion shows a spectrum of clinical and pathologic characteristics, ranging from benign to malignant. Although metastasis is rare, local invasiveness can lead to death.
- Teratoid elements are often present.
- Enucleation is usually required and is curative in a large majority of cases.

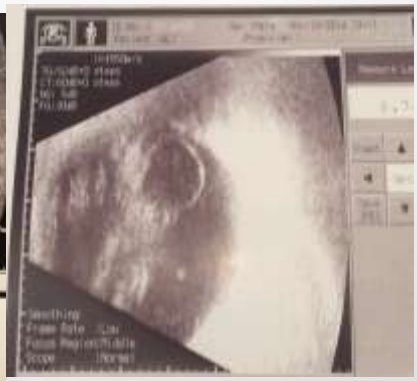
MEDULLOEPITHELIOMA(Diktyoma)





US Criteria

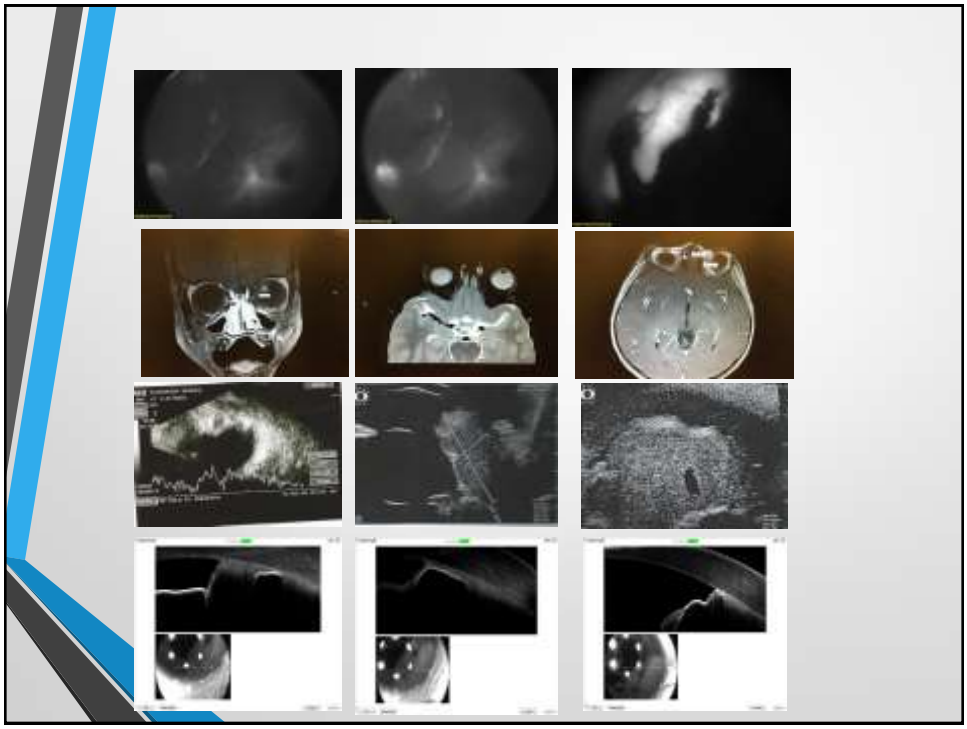




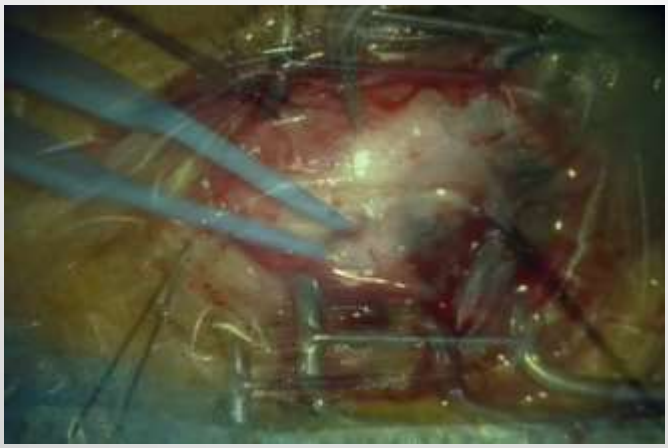
Progressive medulloepithelioma

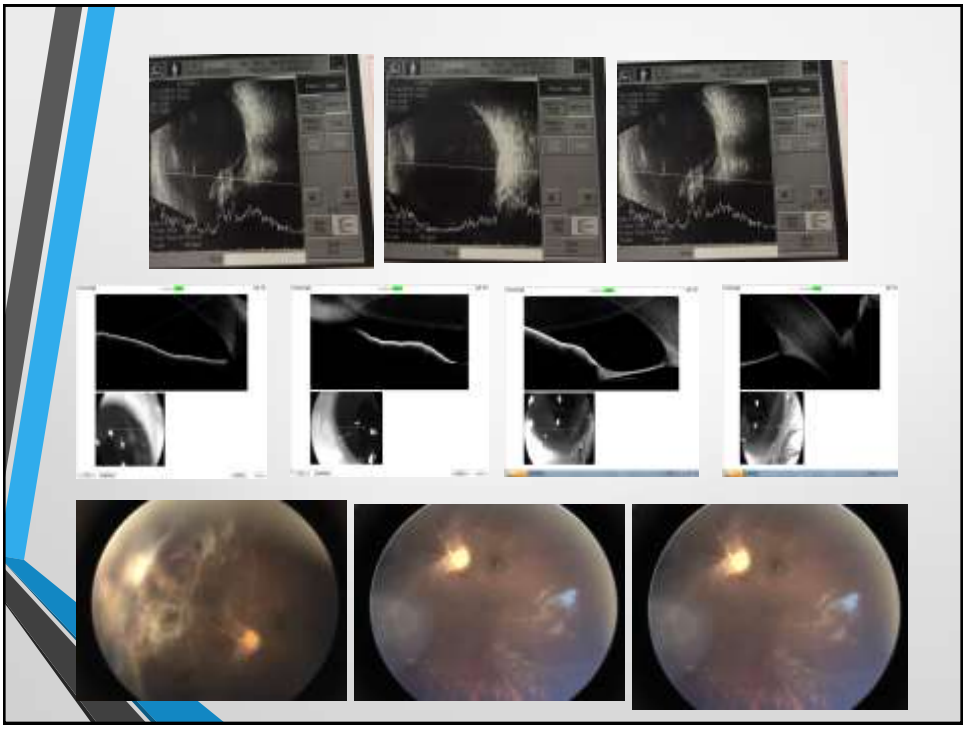
MRI Criteria of Medulloepethelioma

- MRI demonstrate a solid mass with cystic components, usually arising from the ciliary body . The solid component generally enhances markedly on MRI after contrast administration .
- postcontrast-enhanced T1-W MRI show marked enhancement of medulloepithelioma (M) arising from lateral ciliary body . The intense enhancement, tumor location and cysts are features typical of medulloepithelioma.



IRIDOCYCLECTOMY

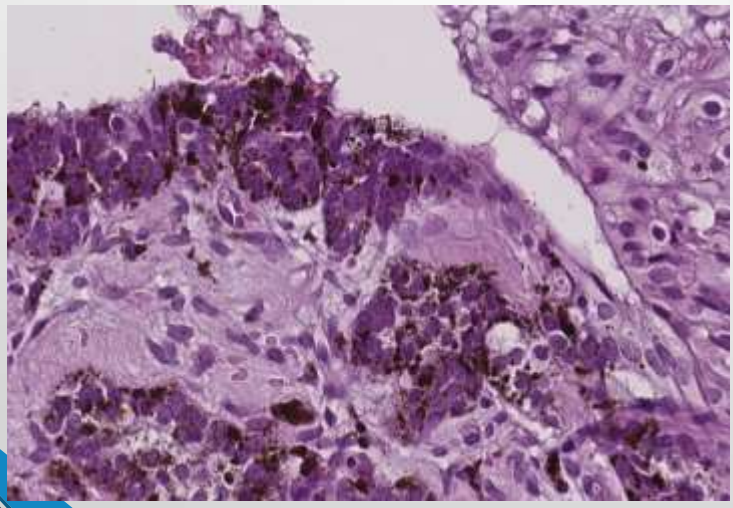




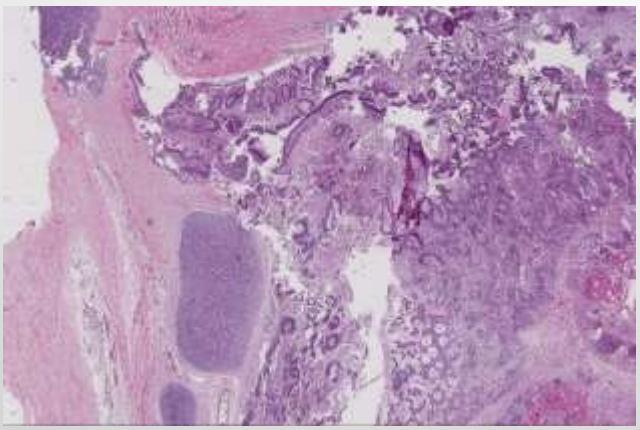
Pathology

- Benign Teratoid medulloepithelioma
- The group of medullo-epitheliomata which mimics embryonic retina before its strata are differentiated is called embryonal medulloepithelioma or diktyoma. The name, diktyoma was coined by Fuch in 1908 because of the net-like structure of the tumour cell arrangement.
- Although closely related to retinoblastoma, diktyomata differ from the former in several ways- neither bilateral nor multicentric in origin nor show any hereditary tendency.

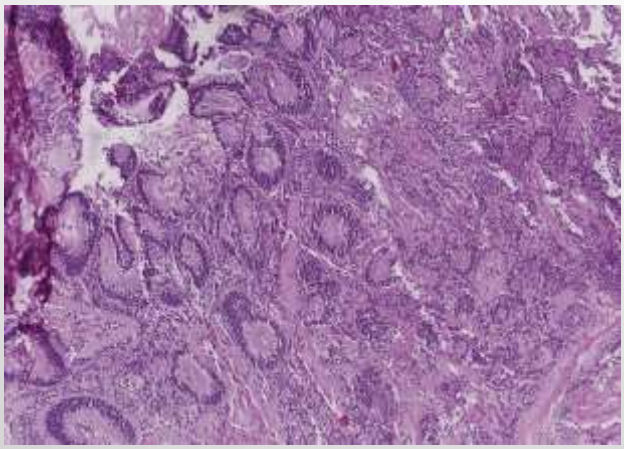
Embryonal cells invading pigmented ciliary body



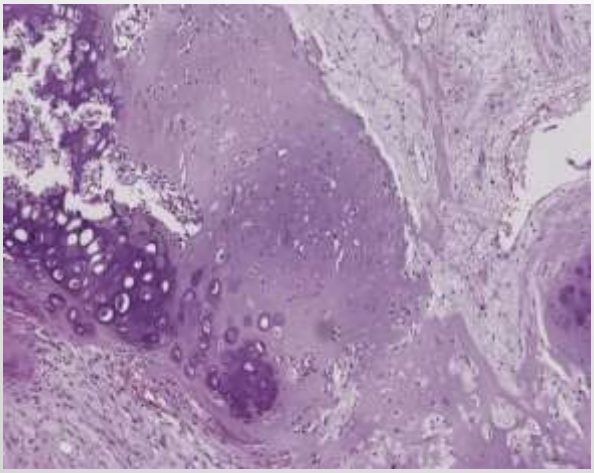
Diktyoma formed of heteroplastic cartilage and neuroepithelial element



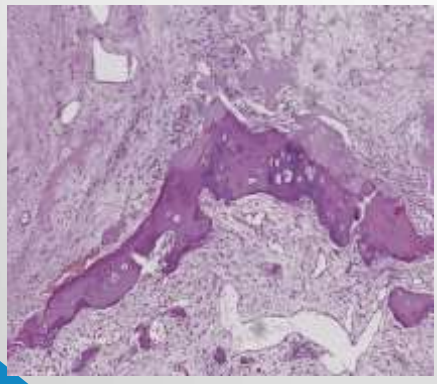
Primitive neuroepithelial neoplasm formed of trabeculae and glands with pseudostrafication

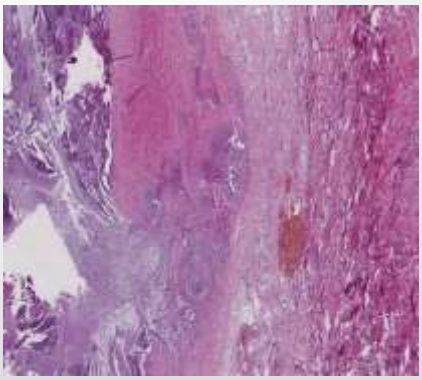


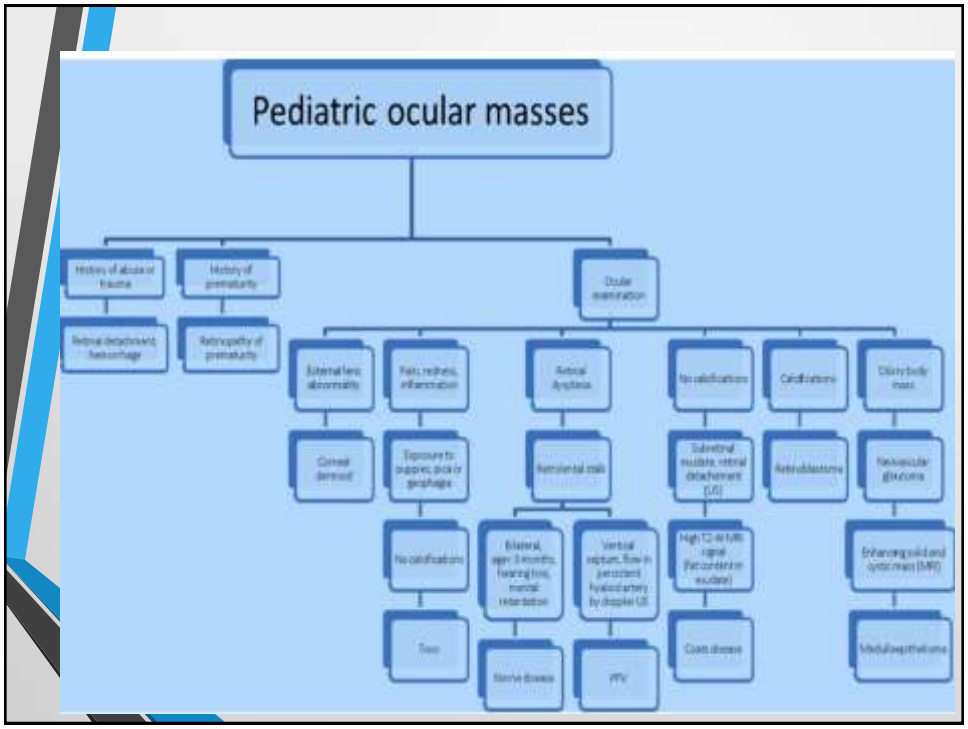
Cartilage with ossification



Heteroplastic bone and cartilage








Thank You