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MR Imaging features of Retinoblastoma with relevance to diagnosis, staging and management

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INTRODUCTION

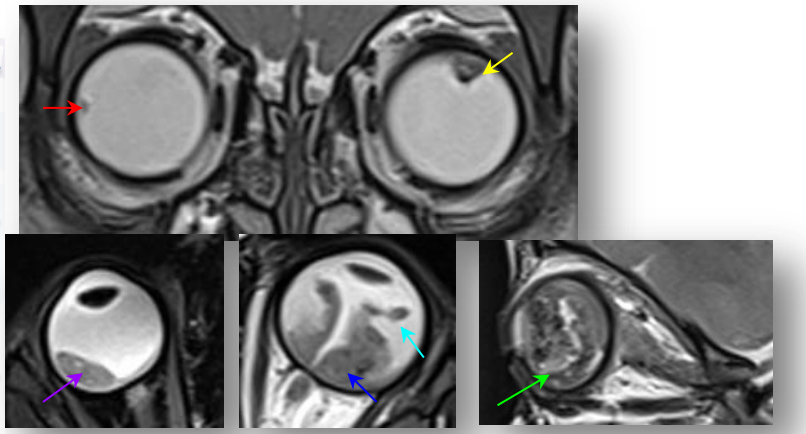
Most common intraocular malignancy in childhood.
Mutation in RB tumour suppressor gene in chromosome 13q14.
90% diagnosed before 5 years of age.
Extra ocular tumours carry worse prognosis, increased metastases.
Heritable forms – early onset, bilateral and multifocal.
Sporadic forms – late onset, solitary.
Clinical features - leukocoria, decreased vision, strabismus, proptosis, orbital pain, orbital inflammation.

AIMS / OBJECTIVES

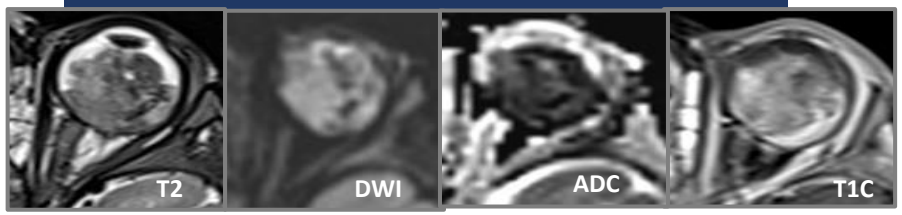
- To describe the MRI findings of retinoblastoma.
- To review the grouping classification of retinoblastoma relevant to radiologists.
- To provide a structured reporting template for ease of communication across multiple disciplines.

INTERNATIONAL INTRAOCULAR RETINOBLASTOMA CLASSIFICATION (IIRC) – The classification is according to increasing severity, the prognosis worsens from very low risk to very high risk and globe salvage becomes progressively unfavourable for some group C tumours and all group D and E tumours.

- **Group A** – Small intraretinal tumors (<3 mm) away from fovea and optic disc (Laser / Cryotherapy)
- **Group B** – All remaining discrete tumors confined to the retina . Sub retinal fluid less than 3 mm from the tumor with no sub retinal seeding (Intravenous / Intra arterial chemotherapy)
- **Group C** – Discrete well-defined tumors with small amounts of sub retinal or vitreous seeding (Intravitreal / Intra arterial chemotherapy)
- **Group D** – Diffuse / large tumors with significant vitreous or sub retinal seeding (light blue arrow) (Intravitreal / Intra arterial chemotherapy +/- Enucleation)
- **Group E** – Presence of poor prognosis features - **Tumor touching the lens, ciliary body, anterior segment/ Diffuse infiltrating RB/ Neovascular glaucoma/Tumor necrosis with aseptic orbital cellulitis/ Phthisis bulbi** (Enucleation / Chemotherapy and/or RT)



TYPICAL MRI FEATURES OF RETINOBLASTOMA



Retinoblastomas are intermediate on T2-weighted image. Tumor calcification is seen as signal void on susceptibility-weighted image. Mass shows restricted diffusion in the form of ADC hypointensity and heterogeneous enhancement.

PROPOSED REPORTING TEMPLATE FOR RETINOBLASTOMA

History: Clinical findings / evaluation / family history
Tumor: Laterality /number / size / location / distance from optic disc
Orbits:
Globe Size/ retinal detachment / hemorrhage / vitreous or retinal seeding
Choroid enhancement / Scleral rim involvement **(worse prognosis)**
Anterior segment – iris, ciliary body, lens
Peri orbital soft tissue involvement
Retro orbital fat involvement
Optic Nerve: Thickening / enhancement / distance of involvement from nerve head insertion **(worse prognosis with increased risk of metastases)**
Brain: Suprasellar / Pineal region
Metastases: Leptomeningeal, bones, nodes, lung, liver