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## ANSWERS TO MEDICAL QUIZ

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A1. The unenhanced axial CT images of the orbit show dense calcification within the posterolateral aspect of the right globe associated with a soft tissue mass. The uveal sclera shows minimal diffuse thickening. The anterior aspect of the globe is not involved .The optic nerve is not thickened and no bony destruction is present. The left globe and orbit is normal.

A2. Retinoblastoma of the right globe.

A3. Leukokoria (white pupillary reflex) is the classical presentation of retinoblastoma, however this is not a specific finding and may also be seen in several other conditions such as: Coat's disease, persistent hyperplasia primary vitreous (PHPV), retrolental fibroplasia and ocular toxicariasis.

Coat's disease is a retinal telangectasia with associated sub-retinal fluid. It is usually seen in males and presents in an older age group (6-8 years) than retinoblastoma.

PHPV does not have calcification and is usually associated with other congenital anomalies like microophthalmos.

Retrolental fibroplasia is seen in premature infants with history of oxygen therapy.

Ocular toxicariasis is usually seen in older children (average age 6 years) with history of close contact with dogs and usually have positive ELISA test.

### DISCUSSION

Retinoblastoma is the most common intraocular malignancy in childhood with an incidence of 1: 20,000 live births. It has no sexual or racial predilection, 90% occur in children younger than 5 years of age. The average age of presentation is 18 months. These tumours are familial in 6-10% and about 25% of retinoblastoma can be either multifocal or bilateral.

Bilateral retinoblastoma is usually transmitted by autosomal dominant inheritance with incomplete penetrance. Bilateral hereditary retinoblastomas are occasionally associated with a pineal neuroectodermal (trilateral retinoblastoma) or with pineal and hypothalamic neuroectodermal tumour (quadrilateral retinoblastoma). Children with the hereditary form are at risk of developing second non-ocular malignancy either within or out of the radiation field. Osteosarcoma is the commonest tumour, other tumours include fibrosarcoma and rhabdomyosarcoma.

Imaging studies like ultrasound and CT are commonly performed and these confirm the diagnosis, although at times MRI is performed to differentiate retinoblastoma from its benign mimic. CT is the commonly performed it can identify calcification in

more than 90% of cases, it can also demonstrate the extension of tumour into the optic nerve and other extra ocular sites.

Treatment of choice in retinoblastoma is enucleation, although other adjunct therapy like external beam radiation may be sometimes indicated. Overall survival rate is more than 90%; survival rate with tumour extension beyond the globe is around 20%.

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