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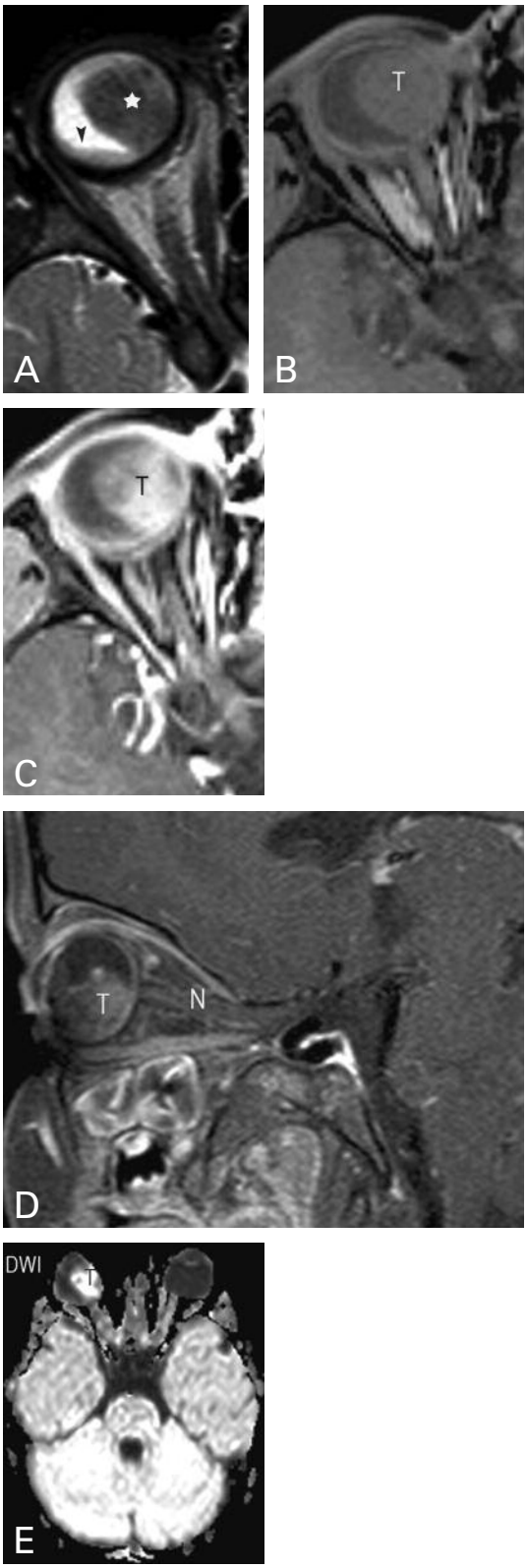
IMAGES IN CLINICAL RADIOLOGY



Unilateral sight loss in a 4-year-old girl

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For 6 weeks the parents of a 4-year-old girl had noticed a difference between the two eyes of their child. Ophtalmological examination revealed leukocoria. This finding raised clinical suspicion of retinoblastoma. MRI was performed. On T2 weighted images a hypointense mass relative to the vitreous humor was evident. There were several hypointensities compatible with calcifications. An area of retinal detachment was also seen. On T1 weighted images a mildly hyperintense mass relative to vitreous humor was seen. It showed marked contrast enhancement. On diffusion weighted images there was restricted diffusion suggestive for a tumoral lesion. The imaging findings were compatible with a retinoblastoma without transscleral or optic nerve extension. The tumor was complicated by retinal detachment reducing visual potential. The patient was treated with enucleation of the affected eye.

Comment

Retinoblastoma is the most common intraocular malignant tumor of childhood. It accounts for 5% of childhood blindness and 1% of cancer deaths. The incidence is 1/15000 to 1/30000 live births.
There are two forms. A sporadic nonfamilial form due to spontaneous mutation of both copies of the Rb 1 gene in a retinoblast. This form accounts for most unilateral disease. Another from consists of a familial heriditary form. In this form bilateral disease is present in 25%. Bilateral disease always points to the heriditary form. Trilateral disease includes bilateral disease and a pineal or suprasellar tumor. Quadrilateral disease consists of bilateral disease and a pineal and suprasellar tumor.
Imaging findings include an intraocular mass with multiple calcifications. In the absence of calcifications other mass lesions responsible for leukocoria should be suspected such as persistent hyperplastic primary vitrious, retrolental fibroplasia, toxocariasis, and Coat's disease. The primary role of imaging is to determine tumor extension. Important findings are optic nerve extension, scleral break through and metastasis. On T1 weighted images the tumor is mildly hyperintense relative to vitrious. Contrast enhancement is moderate to marked. On T2 weighted images the tumor is hypointense relative to vitrious. Retinal detachment is also best demonstrated on T2 weighted images.
Treatment consists of enucleation when vision can not be preserved. Other treatments consist of chemotherapy, external beam radiation therapy, plaque radiotherapy, cryotherapy, and photocoagulation. The rate of cure for noninvasive intraocular retinoblastoma is 90%. Extraocular disease has a more dismal prognosis.

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