Trilateral retinoblastoma: A rare tumor involving the “third eye”

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Procedure: Diagnostic procedure
Technique: MR
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 2 years, female

Clinical History:

A 2 year-old girl initially presented with esotropia of the left eye. A left intraocular tumour was found on ophthalmologic exam. A brain MRI was performed and showed bilateral intraocular masses with calcifications and a pineal mass.

Imaging Findings:

Multiplanar MRI of the brain and orbits with and without gadolinium was performed. The gradient images (Figure 1) show susceptibility artefacts within the bilateral intraocular masses indicating the presence of calcification. These heterogeneous masses are located in the posterior chamber without evidence of optic nerve involvement or extraorbital extension (Figures 2 and 3). A third lesion involving the “third eye” in the pineal region also shows a lobulated and heterogeneous mass without local invasion or evidence of CSF dissemination (Figure 4 and 5).

Discussion:

Retinoblastoma is the most common intraocular malignancy of childhood with 95% of cases developing before age 5. All cases of bilateral retinoblastoma are caused by a germline mutation of the RB1 gene [1, 2]. Approximately 5% of these children also develop an additional midline neuroectodermal tumour, more commonly (75%) in the pineal region [2-4]. Since the pineal gland in some animals has vestigial retinal elements, it is sometimes referred to as the "central or third eye". Also, these midline tumours are similar to retinoblastoma as they both arise from primitive neuroectodermal tissue and hence described by some as trilateral retinoblastoma (TRb) when they occur together. Classic trilateral retinoblastoma is associated with poor prognosis having a 5-year survival rate of only 22% [2]. However, a case series of children with bilateral retinoblastoma who developed benign pineal neoplasms suggest that a benign variant of trilateral retinoblastoma exists and that it might have a better prognosis than classic TRb [5]. Over the last decade, survival has increased substantially with earlier detection coupled with advances in chemotherapy [2, 3, 6].

Magnetic Resonance Imaging (MRI) is the preferred modality for staging retinoblastoma prior to treatment. The retinal mass can show variable enhancement depending on the size of the tumor, associated calcifications and necrotic changes. When conservative management is contemplated, it is important to assess the optic nerve and the presence of extraocular tumor extension [7].

MRI findings of pineal involvement will show an enlarged gland with irregular enhancement and necrotic or cystic
changes [5]. Although less common than germ cell tumours, primary pineal parenchymal tumors are mostly represented by pineocytomas and pineoblastomas. Pineoblastomas are more common in children and tend to be more aggressive with associated local invasion and often metastasis though the CSF. Pineocytomas on the other hand are usually slow growing and often non-invasive but can also metastasize.

This patient was found to have bilateral retinoblastoma and a pineal neoplasm on MRI compatible with trilateral retinoblastoma. She subsequently underwent enucleation of the left eye and laser resection of the right retinal tumour to spare the right eye, as well as removal of the pineal mass, followed by chemotherapy. Final histopathology of the pineal tumor demonstrated a benign pineocytoma. She is now 8 years old with 20/25 vision of her right eye.

**Differential Diagnosis List:** Trilateral retinoblastoma, Intraocular infection, Pineal germinoma

**Final Diagnosis:** Trilateral retinoblastoma

**References:**


Description: Axial GRE shows susceptibility artifact within the two intraocular masses (arrows) indicating calcifications. Origin: UC Davis Medical Center
Description: Coronal T2 Fat Sat – Normal sized globes with two separate intraocular masses (arrows).
Origin: UC Davis Medical Center
Description: Axial T1 Fast Sat Post Gadolinium – Heterogeneous enhancement of the bilateral intraocular masses (arrows). The optic nerves are not involved and there is no evidence of extraocular extension. Origin: UC Davis Medical Center
Description: Sagittal T1 FLAIR demonstrates the heterogeneous midline pineal (arrow). Origin: UC Davis Medical Center
Description: Axial T1 post Gadolinium shows a lobulated and heterogeneously enhancing mass in the region of the pineal gland. **Origin:** UC Davis Medical Center