Trilateral retinoblastoma with unilateral eye involvement

**RETINOBLASTOMA TRILATERAL COM ACOMETIMENTO OCULAR UNIVERSAL**

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**SUMMARY**

Retinoblastomas (RB) are the main forms of intraocular tumor in childhood, with a worldwide incidence of 1 case per 15,000 to 20,000 live births. Trilateral RB (RBT) is a rare combination of unilateral or bilateral RB with a midline intracranial neoplasm of neuroblastic origin, usually found in the pineal region or the suprasellar region, presenting variable incidence of 0.5% up to 6% among patients with RB. The article reports a case of unilateral RBT in a patient treated at Hospital A.C.Camargo.

**Keywords:** retinoblastoma, eye neoplasms, infant.
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FIGURE 1 (A) MRI with contrast sagittal T1-weighted image. (B) MRI with contrast coronal T1-weighted image. (C) MRI with contrast axial T1-weighted image.

FIGURE 2 Retinoblastoma. (A) Histological section of left eyeball showing a 1.3 cm tumor in the posterior chamber, connected to the retina without choroidal or optic nerve infiltration (H&E, 0.5x). (B) Greater magnification showing a malignant neoplasm of small round blue cells, (C) with multiple foci of dystrophic calcifications (H&E, 20x). (D) Homer-Wright rosettes were focally present, but Flexner-Wintersteiner rosettes were not seen (H&E, 20x).
steadily in subsequent follow-ups, without signs of systemic disease or impairment of the right eye.

With early diagnosis, the survival rate for retinoblastoma is good, reaching 93% at 5 years. In cases of RBT, prognosis is worse with survival rate between 9 and 13 months after diagnosis, mainly due to spinal tumor metastases, usually associated with delayed diagnosis of intracranial lesions. Some authors recommend routine assessment in patients with hereditary retinoblastoma with periodic MRI-SO until 4 years of age for early detection of asymptomatic tumors of the pineal or suprasellar region.8,9

**Resumo**

Retinoblastoma trilateral com acometimento ocular unilateral

O retinoblastoma (RB) é a principal forma de tumor intraocular na infância, apresentando uma incidência mundial de 1 caso em cada 15 mil a 20 mil nascidos vivos. O RB trilateral (RBT) é uma rara combinação de RB unilateral ou bilateral com uma neoplasia da linha média intracraniana de origem neuroblástica, geralmente na região da glândula pineal ou na região suprasellar, apresentando incidência variável de 0,5 a 6% entre pacientes com RB. O artigo relata o caso de uma paciente com RBT com acometimento unilateral atendida no Hospital A.C. Camargo.

**Palavras-chave:** retinoblastoma, neoplasias oculares, lactente.

**References**