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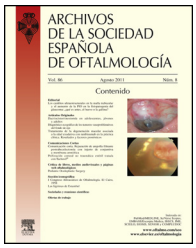

Short communication

Retinoblastoma in the adolescent. Unusual clinical and histopathology findings☆

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ARTICLE INFO

Article history:
Received 27 November 2014
Accepted 7 December 2015
Available online 4 March 2016

Keywords:
Retinoblastoma
Enucleation
Immunohistochemistry
Synaptophysin
CD99

ABSTRACT

Case report: A 17-year-old male with 2 years history of an intraocular mass and progressive visual loss of the left eye. Spontaneous sclera rupture occurred during enucleation. Microscopic evaluation with H–E, PAS and immunohistochemistry (NSE, GAFP, SYN, CD99) revealed a small blue round cell malignant neoplasm with extensive necrosis and apoptosis. The optic nerve, ciliary body, choroid, anterior chamber, and sclera were infiltrated. SYN was positive and CD99 was negative in neoplastic cells, consistent with a poorly differentiated retinoblastoma.

Discussion: Retinoblastoma is the most frequent primary intraocular malignant tumor in childhood, but occasionally older patients can be affected. Immunohistochemistry is mandatory in poorly differentiated retinoblastomas.

Retinoblastoma en un adolescente. Hallazgos clinicopatológicos poco frecuentes

Palabras clave:
Retinoblastoma
Enucleación
Inmunohistoquímica
Sinaptofisina
CD99

RESUMEN

Caso clínico: Hombre de 17 años, con un tumor intraocular izquierdo de 2 años de evolución y pérdida visual progresiva. Presentó rotura escleral durante la enucleación. Microscópicamente, las tinciones de H-E, PAS e inmunohistoquímica (NSE, GAFP, SYN, y CD99) demostraron un tumor maligno de células pequeñas, redondas y azules, con necrosis, apoptosis e invasión al nervio óptico, cuerpo ciliar, coroides, cámara anterior y esclerótica. La SYN resultó positiva y el CD99 negativo en células neoplásicas, confirmándose un retinoblastoma pobremente diferenciado.

☆ Please cite this article as: Hernández-Ayuso I, Ríos y Valles-Valles D, Lome-Maldonado C, Gómez-Leal A, Rodríguez-Martínez HA, Rodríguez-Reyes AA. Retinoblastoma en un adolescente. Hallazgos clinicopatológicos poco frecuentes. Arch Soc Esp Oftalmol. 2016;91:145–148.

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Discusión: El retinoblastoma es el tumor intraocular maligno primario más frecuente en niños, aunque ocasionalmente afecta a otros grupos de edad. La inmunohistoquímica es obligada en los retinoblastomas pobremente diferenciados.

Introduction

Small, rounded and blue cell tumors comprise a group of potentially malign neoplastic proliferations that, in differential diagnostic, comprise lymphoma/leukemia, rhabdomyosarcoma, neuroblastoma and peripheral primitive neuroectodermal tumor (PNET)/Ewing sarcoma (EWS). Confined within the ocular globe, said tumors give rise to a specific group of diseases having the main diagnostic possibilities as retinoblastoma, medulloepithelioma,[1] rhabdomyosarcoma originated in medulloepithelioma,[2] lymphoma/leukemia and primary retinal[3] or choroidal metastatic[4] PNET/EWS. When the clinic course and histopathological appearance of the tumor are atypical, the use of immunohistochemistry markers are required for adequate classification, considering the implications for treatment and prognosis.

Clinic case report

Male, 17, without relevant history, exhibiting a large size intraocular mass in the left eye with 2 years evolution and progressive loss of vision reaching absence of luminosity perception. In the past 6 months the patient exhibited painful proptosis and conjunctival chemosis. Mode B echography did not show intraocular calcifications (Fig. 1). Due to the presence of an intraocular tumor together with absence of light perception, ocular globe enucleation was indicated. In the course of this procedure, anterior scleral spontaneous rupture occurred (Fig. 2). The contralateral eye did not exhibit alterations.

The histopathological study demonstrated malign neoplasia comprised by small, round and blue cells with extensive necrosis and without dystrophic calcification areas (Fig. 3). The feasible tumor was arranged in perivascular rods invading

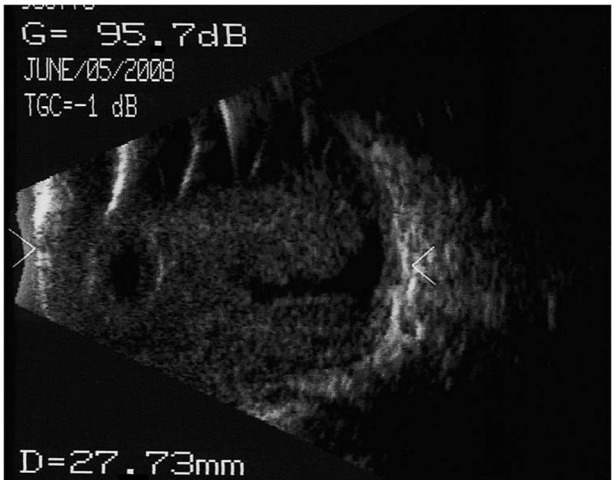

Fig. 1 – Mode B ultrasound showing phakic ocular globe with anteroposterior axis measuring 27.7 mm, with a mass having homogeneous internal appearance and focus the increased density occupying most of the vitreous cavity.

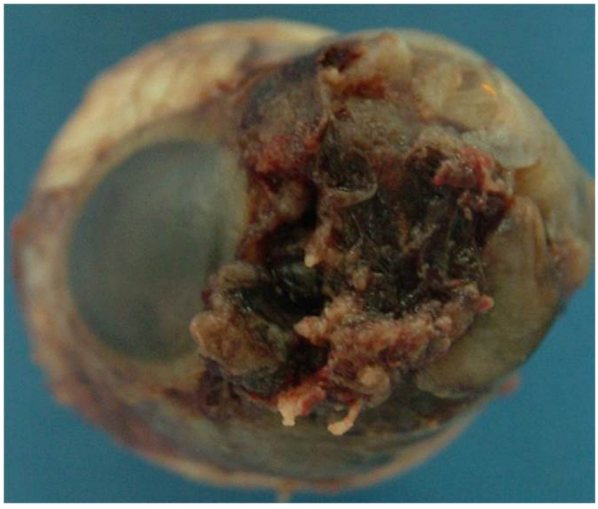

Fig. 2 – Macroscopic image showing thinned sclerocorneal limbus allowing the exit of pro-hemorrhagic tumor.

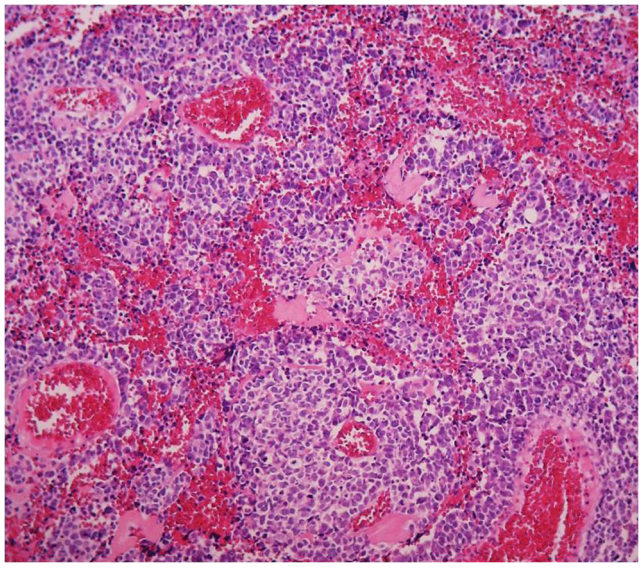

Fig. 3 – Photomicrograph showing malign neoplasia of small, round and blue cells arranged in some areas appearing as perivascular rods. Hematoxylin–inosine staining, 10× enlargement.