Cortical ependymoma. Case report

Evolución y Tasa de Recuperación en el Tratamiento Ependimoma Cortical. Relato de caso

Antonio Daher
Adriana Brito
Daniela Diaz

ABSTRACT
Ependymomas are rare central nervous system tumors derived from ependymal cells, located in the ventricular system, choroid plexus and central canal of the spinal cord. Ependymomas could appear throughout the craniospinal axis, even outside the ventricular system, although, pure cortical ependymoma are very rare, and only a few cases have been reported in the literature. We hereby present the case of a 46 years-old female, admitted to the hospital, presenting a new-onset seizure. Magnetic Resonance Imaging (MRI) showed a large bilateral frontoparietal tumor, associated to important perilesional edema. Radical resection of the tumor was performed, and the biopsy reported anaplastic ependymoma. Patient underwent complementary radiotherapy.

Keywords: Ependymoma; Cortical ependymoma; Brain tumor

INTRODUCTION
Ependymomas are rare central nervous system tumors that arise from differentiated ependymal cells located in the ventricular system, choroid plexus, and along the central canal of the spinal cord. Representing 2% to 9% of all neuroepithelial tumors, localizations outside the ventricular system are uncommon, and affect pediatric and adult patients equally. Few cases of pure Cortical Ependymomas (CEs) have been reported in the literature. CEs usually manifest with a size larger than 4cm, and symptoms are relatively mild until a later stage. Patients tend to present focal neurologic deficits, and most frequently seizures. We present a case of a 47-year-old female with a frontoparietal cortical ependymoma manifested as new-onset seizure and left hemiparesis.
Right handed female, 46-year-old, was admitted to the ER with a new-onset seizure and left hemiparesis. Neurosurgical service was consulted. Laboratory examinations were normal. MRI showed a large, bilateral frontoparietal tumor with more extension to the right hemisphere, associated to important perilesional edema (Figures 1, 2). Meningioma and anaplastic meningioma were the primary differential radiological diagnosis.

Bifrontal craniotomy with extended resection to the right side was performed disclosing important edema and bleeding. Patient underwent radical resection of the tumor located in the parenchyma with no dural attachment. Histological examination demonstrated findings compatible with high grade anaplastic ependymoma. Radiotherapy with linear accelerator (LINAC) was performed. The patient reached a satisfactory recovery with a left hemiparesis as sequel.

**CASE REPORT**

Ependymomas are rare central nervous system (CNS) tumors that arise from differentiated ependymal cells located in the ventricular system, choroid plexus, and along the central canal of spinal cord. Representing 2% to 9% of all neuroepithelial tumors, it may manifest at all ages, affecting more frequently children and young adults, with no gender predilection.

Ependymomas can appear in any site of the craniospinal axis, even outside the ventricular system, however, they’re more frequently located in the cervico-thoracic segment of the spinal central canal and fourth ventricle, and only a third are supratentorial.

From all supratentorial ependymomas, it is estimated that about 50% are originated from ependymal cells related to the ventricular system and choroid plexus. The remaining...
is distributed throughout brain parenchyma. Localization outside the ventricular system is uncommon, and affects pediatric and adult patients equally. Few cases of pure Cortical Ependymomas (CEs) have been reported in the literature, which are located in the cortical ribbon with no identified connection to the ventricular lining. In the series of cases presented at the Mayo Clinic in 2011, the most common localization was the frontal lobe, followed by parietal and occipital lobes. A very low frequency of CEs in the temporal lobe has been observed.

CEs usually are manifested with a size larger than 4cm, and symptoms are relatively mild until a later stage. Patients tend to present focal neurologic deficits and most frequently seizures, regardless of their rare localization in the temporal lobe. CEs have also been reported as incidental during workup for headaches.

Even though there is no clear explanation for the histogenesis of CEs, it has been proposed that supratentorial ependymomas could be originated from rests of ependymal cells located in the angle of the ventricle, deep into the adjacent parenchyma. On the other hand, due to the wide distribution of ependymomas without restriction to the mentioned area, this mechanism seemed doubtful. Progenitor cells hypothesis has been proposed as an explanation of cortical ependymomas pathogenesis. In 2005, Hegyi et al. based on the pathologic features of an ectopic retinal ependymoma suggested that the origin of this neoplasm could be explained for Muller cells (with progenitor properties).

CEs had shown a wide spectrum of morphologies, and could be difficult to distinguish from some infiltrative gliomas, typically showing the characteristic perivascular pseudo-rosettes, and in some cases true ependymal rosettes. Less classic features could be found, as tanyctic, epithelial and clear cells, and even rarely spindle cell component and schwannian-like nodules, important for differential diagnosis with Angiocentric Glioma.

According to the World Health Organization (WHO) classification, for CNS tumors, ependymomas have been traditionally divided into low grade (Grade II), and high grade (Grade III) lesions, reserving Grade I for myxopapillary ependymoma. A higher proportion of supratentorial ependymomas are high grade when compared with infratentorial ependymomas. Nevertheless, cortical ependymomas are most commonly low grade with a relatively better clinical course.

Recently in 2016, the WHO CNS tumor was updated, including now molecular parameters in addition to the histological findings, to define many tumor entities. In this update, a genetically defined ependymoma subtype has been accepted: Ependymoma, RELA fusion–positive, responsible for the majority of supratentorial tumors in children. It is expected that continuing studies of the molecular features will provide a more objective classification to this tumors and, therefore, a major clinical significance to the histological classification.

Magnetic Resonance Imaging (MRI) is the primary modality, for assessment of ependymoma. Intracranial ependymomas conventionally demonstrate hypo-intensity on T1, hyper-intensity on T2, and intermediate-to-high FLAIR signal intensity, relative to gray and white matter. Supratentorial ependymomas tend to demonstrate more heterogeneous T1 and T2 due to a greater tendency toward cyst formation, calcifications and hemorrhage, when compared with infratentorial ependymomas. Commonly, on post gadolinium T1-weighted images, there is avid enhancement of the soft tissue within the tumor, intermixed with poorly or none enhancing areas.

Radical surgical resection is considered to be the best treatment, in light of easier approach for CEs, than intraventricular tumors. Postoperative radiotherapy is used for anaplastic ependymoma, and partially resected tumor, due to the possibility of recurrence, and 30 to 40% chances of leptomeningeal dissemination at recurrence. Chemotherapy and prophylactic craniospinal irradiation are not indicated.

CEs appear to have a relatively favorable prognosis when compared to other supratentorial ependymomas.
According to Metellus et al. the 5 year survival rate is 57.1% +/- 8.7%; although, histopathological type, localization, extent of resection, age at diagnosis and gender must be considered.

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CORRESPONDING AUTHOR

Antonio Daher, MD, PhD
Neurosurgeon
Adjunct Professor of Neurosurgery
Centro Médico Guerra Mendez
Hospital Oncológico Miguel Perez Carreño
Valencia, Venezuela
E-mail: antonio.daher@gmail.com