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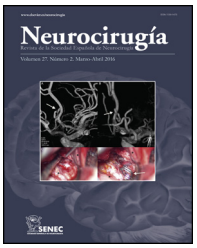

Case Report

Fluorescence guided resection with 5-aminolevulinic acid of a pilomyxoid astrocytoma of the third ventricle

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ABSTRACT

Fluorescence-guided resection with 5-aminolevulinic acid has been shown to be useful in the resection of certain brain tumors other than high grade gliomas, facilitating the intraoperative differentiation of neoplastic tissue. The technique enables the surgeon to ensure that no tumor fragments remain, thereby achieving higher rates of complete resection.

Tihan first described pilomyxoid astrocytomas in 1999. They are currently classified as grade II astrocytoma according to the WHO classification system and, because of their tendency to recur and their dissemination through the cerebrospinal fluid pathways, they are considered to be more aggressive than pilocytic astrocytoma. As a result, management of these tumors must be more aggressive, always aiming for complete macroscopic resection whenever possible.

In this article, we present a case of pilomyxoid astrocytoma of the third ventricle in which the use of fluorescence-guided resection with 5-ALA facilitated complete resection. Imaging tests performed after five years revealed no signs of recurrence and no adjuvant radiotherapy or chemotherapy was required. This article also comprises a review of the literature concerning the characteristics and management of this tumor, which was recently considered to be a different histopathological entity.

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Resección guiada por fluorescencia con ácido 5-aminolevulínico de un astrocitoma pilomixoide del tercer ventrículo

Palabras clave:
Resección guiada
por fluorescencia
Ácido 5-aminolevulínico
Fluorescencia intraoperatoria
Astrocitoma pilomixoide
Tumor del tercer ventrículo

RESUMEN

La resección guiada por fluorescencia con 5-ALA se ha mostrado útil en tumores diferentes a los gliomas de alto grado, permitiendo la diferenciación intraoperatoria del tejido tumoral. La técnica permite revisar el lecho quirúrgico para comprobar que no quedan fragmentos tumorales, consiguiéndose así mejorar las tasas de resección completa.

El astrocitoma pilomixoide, descrito en 1999 por Tihan, se clasifica actualmente como un astrocitoma grado II en la clasificación de la OMS y es considerado como una variante con mayor agresividad que el astrocitoma pilocítico por su tendencia a la recidiva y a la diseminación por el líquido cefalorraquídeo. Por ello el tratamiento debe ser más agresivo, fundamentado en una resección macroscópicamente completa siempre que se pueda.

En este artículo presentamos el caso de un astrocitoma pilomixoide del tercer ventrículo en el que la fluorescencia con 5-ALA permitió una resección completa, sin signos de recidiva en pruebas de imagen a los 5 años, sin haber precisado tratamiento complementario con radioterapia ni quimioterapia. Se hace además una revisión de la literatura acerca de las características y el manejo de este tumor recientemente considerado como una entidad histopatológica diferente.

Introduction

Pilomyxoid astrocytomas (PMAs) are primary tumors of the central nervous system that were recently identified as a variant of pilocytic astrocytoma (PA). But they exhibit clinicopathological and prognostic features that define them as more aggressive than PAs.[1] Because of this, treatment must be also more aggressive, with complete resection being the first goal of treatment.[2–5] Although this is usually precluded in certain locations such as the hypothalamic-chiasmatic area,[2–4,6] when located in more favorable areas such as cerebral hemispheres, cerebellum and even in the spinal cord, the goal of complete resection can be achieved improving the clinical outcome.

Fluorescence guided resection (FGR) with 5-ALA is an expanding surgical technique. After the first report of Stummer,[7] showing that high grade gliomas showed red fluorescence after administration of 5-ALA when illuminated with blue light, there have been numerous reports confirming the usefulness of the technique not only for high grade gliomas, but also for other types of tumor[8–21] as well as an adjuvant in the gathering of tumor biopsies.[22–24]

We are reporting a case of PMA of the third ventricle in which intraoperative fluorescence after administration of 5-ALA allowed us to completely remove the tumor. To the best of our knowledge this is the first time that FGR has been useful in the complete resection of a PMA in the hypothalamic area.

Clinical presentation

This 20-year-old male was admitted to our department with a history of progressive headaches for the previous two weeks. Past history was unremarkable. Neurological examination was unremarkable. Magnetic resonance (MR) imaging revealed a well-delimitated tumor in the third ventricle causing hydrocephalus. It was rounded, hypointense in T1-weighted, showing peripheral enhancement and no surrounding edema (Fig. 1A). Two days later a ventriculo-peritoneal shunt had to be inserted because clinical worsening due to increased hydrocephalus with immediate abatement of symptomatology.

A control computed tomography (CT) showed that the right ventricle was trapped. An endoscopic septostomy plus biopsy was performed. Pathological diagnosis was PMA (Fig. 2). A few days later a transcallosal approach to the tumor was performed after administration of 20 mg per kg weight of 5-ALA by mouth three hours before taking the patient to the operating room. The tumor was grayish and emitted intense red fluorescence when illuminated with blue light (Fig. 3). The tumor originated from the floor of the third ventricle and was readily dissected and removed. The attachment area in the floor of the third ventricle was inspected with blue light and every spot was removed until no red area was seen. MR performed 48 h later confirmed a complete resection of the tumor (Fig. 1B).

Pathological diagnosis of the removed tumor was PMA. Every single specimen showing fluorescence was informed as PMA. A total body MR imaging ruled out any pathological implant in the neuraxis.

The patient recovered uneventfully until three months later when he suffered acute neurological deterioration because right ventricle dilatation that required a ventriculo-peritoneal shunt on the right with immediate recovery.

No additional adjuvant therapy was instituted and 5 years later the patient is fully independent, although he has slight memory impairment and hypothyroidism. Sequential imaging studies have ruled out any recurrence (Fig. 1C).

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