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Clinical report

Optic chiasm glioma in children: Endocrine disorders in 14 cases☆

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ABSTRACT

Background and objectives: To describe the frequency of endocrine disorders in children with optic chiasm glioma and analyze related factors.
Patients and methods: Review of medical records by collecting sex, age, history of neurofibromatosis, clinical presentation, treatment of tumour, and presence of endocrine abnormalities. Statistical tests Wilcoxon and Fisher.
Results: Fourteen patients (6 female) with age at diagnosis of 0.5–7.0 years (mean ± standard deviation: 2.97 ± 2.32) and follow-up of 10.64 ± 3.30 years (range 6.0–16.0). 12/14 presented endocrinopathy at follow-up: 8 precocious puberty, 5 hypopituitarism, and 5 obesity. The onset of deficits was related to the neuroophthalmological symptoms under the age of five ($p = 0.02$) and treatment of the tumour was required ($p = 0.03$).
Conclusions: Children with optic chiasm gliomas may present endocrine disorders from the time of diagnosis of the tumour and, in particular as they develop on. The most common of these is precocious puberty. Pituitary deficits are associated with more aggressive tumours (those presenting with neuroophthalmological signs and symptoms before the age of five and requiring treatment).

Glioma del quiasma óptico en niños: complicaciones endocrinológicas en 14 casos

Palabras clave:
Glioma óptico de los niños
Neoplasias hipotalámicas
Hipopituitarismo
Pubertad precoz
Niños

RESUMEN

Fundamento y objetivos: Describir la frecuencia de endocrinopatías en niños con glioma del quiasma óptico y analizar los factores relacionados.
Pacientes y métodos: Revisión de historias recogiendo las variables sexo, edad, antecedente de neurofibromatosis, forma de presentación, tratamiento del tumor y presencia de endocrinopatías. Pruebas estadísticas: Wilcoxon y Fisher.
Resultados: Catorce pacientes (6 mujeres) con edad al diagnóstico de 0,5 a 7,0 años (media ± desviación típica: 2,97 ± 2,32) y tiempo de seguimiento de 10,64 ± 3,30 años (rango 6,0–16,0). Doce de 14 presentaban endocrinopatía al final del seguimiento: 8 pubertad precoz, 5 hipopituitarismo y 5 obesidad. La aparición de déficits se relacionó con la clínica neurooftalmológica antes de los 5 años de edad (p = 0,02) y con el requerimiento de tratamiento de la lesión (p = 0,03).
Conclusiones: Los niños con gliomas del quiasma óptico pueden presentar endocrinopatías desde el momento del diagnóstico del tumor y, sobre todo, a lo largo de su evolución. La más frecuente es la pubertad precoz. Los déficits hipofisarios se relacionan con los tumores más agresivos (aquellos que debutan con clínica neurooftalmológica antes de los 5 años de edad y que requieren tratamiento).

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Introduction

Gliomas account for 40% of tumours of the central nervous system in children. They can be found in cerebellum, brainstem, and visual pathway, which is the most common location, with half of the cases originating in these. Within this last location, those who settle in the chiasmal or suprasellar region are often associated with endocrine disorders that may even be the first manifestations of neoplasm[1–4] (Fig. 1).

Before suggesting a therapeutic approach, we must establish a differential diagnosis with other lesions of this region, because treatment is very different depending on its nature. Craniopharyngioma, the most frequent sellar lesion in the first 2 decades of life, often require surgery; prolactinoma and pituitary hyperplasia secondary to long-standing primary hypothyroidism respond to medical treatment, while germ cell tumours are treated with chemotherapy and radiotherapy.[5–7] Gliomas, according to their degree of aggressiveness, sometimes require surgery, other chemotherapy and/or radiotherapy, and many require no action.[8–10]

The objectives of this study are to describe the frequency of endocrinological complications in optic chiasm gliomas in children, at the time of diagnosis and throughout their progression, and analyze the factors that relate to them.

Patients and methods

A retrospective study of patients under 14 years of age referred to a paediatric endocrinology consultation of a tertiary hospital due to the presence of an optic chiasm glioma over a period of 15 years (2000–2014). The injuries were diagnosed by magnetic resonance imaging (MRI).

The following variables were collected from the medical records of each patient: sex, age at diagnosis, history of type 1 neurofibromatosis (NF), symptoms and signs of clinical presentation, primary treatment of tumour and presence of endocrine disorders at diagnosis and throughout the follow-up period.

Symptoms and signs of clinical presentation were defined as the first related to the lesion that led to consultation. Different forms were distinguished based on whether these were derived from neuropathies (headache, signs of intracranial hypertension, seizures, focal signs, etc.), eye diseases (decreased visual acuity or visual field, strabismus, nystagmus, etc.) or endocrine disorders.

Pathologies of hypothalamic origin were included in the last group (new onset obesity, eating, thirst, sleep or temperature disorders) as well as pituitary, either due to a deficiency of some of the hormones of the anterior pituitary (growth hormone [GH], adrenocorticotropic hormone [ACTH], thyrotropic hormone [TSH], luteinizing hormone [LH] and follicle-stimulating hormone [FSH]) or posterior pituitary (central diabetes insipidus), or by overproduction of the same (hyperprolactinemia, early puberty and gigantism). Clinical and laboratory evaluations followed the typical care protocols in these patients and were performed initially at diagnosis of the lesion and then semi-annually.

Quantitative variables (age and time periods) were expressed as mean values ± standard deviation and were compared using the Wilcoxon statistical test. The qualitative variables were expressed as proportions and compared by the Fisher test.

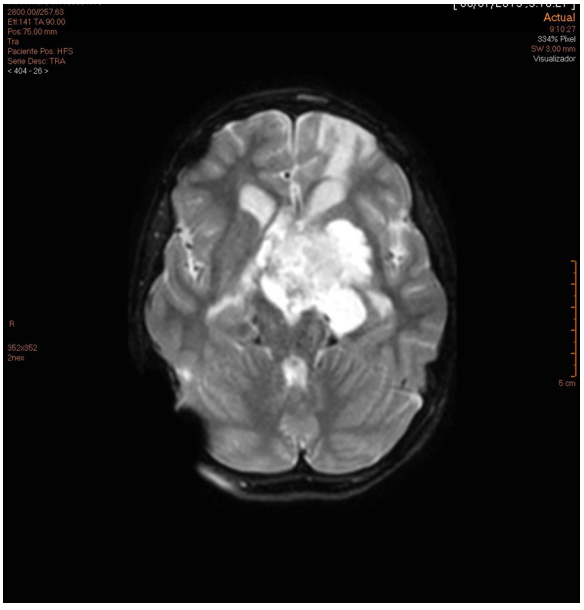

Fig. 1. Nuclear magnetic resonance image (sequence T2) of a case showing a solid heterogeneous, hyperintense, mass located within the sella and suprasellar cistern, extending to the floor and anterior portion of the third ventricle.

Results

Fourteen patients were included in the study (6 women) with an age range at diagnosis of 6 months to 7 years (mean 2.97 ± 2.32 years) and a follow-up time of 10.64 ± 3.30 years (range 6.0–16.0 years). Six of them were previously diagnosed with NF. Table 1 shows the clinical data of patients and differences between subgroups with and without a history of NF.

The reasons for consultation were neurological and visual symptoms or signs in 8 cases and hormonal in 3: precocious puberty in 3 cases, associated with obesity (for recent weight gain) on one of them. Three more patients had a subclinical diagnosis (through neuroimaging), as they were being monitored for NF. The neuro-ophthalmological signs and symptoms were significantly more frequent in children without this disease (6/8 cases versus 2/6 in those affected by NF, $p = 0.05$).

The glioma was treated surgically or oncologically only in 8 children (receiving chemotherapy [carboplatin and vincristine regimen] and/or radiotherapy); in the other 6 cases, no therapy was required for the lesion. The need for primary treatment of the neoplasm was significantly more frequent in patients without NF (7/8 cases versus 1/6, $p = 0.02$).

Although at the time of diagnosis of glioma only 3 children manifested some endocrinopathy (all precocious puberty, in one case with obesity, and no hypopituitarism), the proportion of patients with any hormonal disorder amounted to 12/14 at the end of follow-up period (Table 2): 8 children with precocious puberty (7 of them isolated), 5 with obesity (4 became obese during follow-up, 1 of them fulfilled criteria for metabolic syndrome) and 5 with pituitary deficits: 2 of them isolated (1 of GH and other gonadotropins) and 3 multiple (having previously presented one precocious puberty). There has been no disorder of the neurohypophysis or other different hypothalamic disorder besides obesity. A girl whose glioma was treated with surgery and chemotherapy developed a chronic autoimmune thyroiditis 8 years after treatment, and a GH deficiency. The time of onset of early puberty was 3.7 ± 3.0 years (range 0–7.0), while the pituitary deficiencies occurred at 7.9 ± 3.4 years (range 2.6–11.5).

The onset of hypopituitarism was associated with the more aggressive tumours: in the 5 cases that presented them, there were neuro-ophthalmological signs and symptoms before 5 years of age and had required surgical or oncological treatment for the lesion (Table 2). Neither the onset of early puberty nor the weight gain was associated with any variable studied (Table 2).