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Chordoid glioma: Case report and review of the literature

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ABSTRACT

INTRODUCTION: Chordoid glioma is a rare low-grade brain tumor originating from the anterior wall of the third ventricle.
CASE PRESENTATION: A 13-year-old female with progressive intermittent holocranial headaches and a diagnosis of chordoid glioma underwent tumor resection in our neuro-oncology unit.
DISCUSSION: We review all 79 cases of chordoid glioma reported in the literature so far, focusing on the diagnostic criteria, treatment options and prognosis.
CONCLUSION: Efficient treatment of chordoid glioma depends upon radical surgical resection. Based on the reviewed data, which showed high morbi-mortality rates for this kind of tumor, we recommend a more conservative treatment approach.

1. Introduction

Chordoid glioma (CG) is a rare low-grade tumor that arises from the anterior wall or roof of the third ventricle. It was first described in 1998 by Brat et al. [1]. In 2000, it was incorporated into the World Health Organization (WHO) classification as grade II [2]. Chordoid glioma affects women at a rate of 2:1, and most patients are between 30 and 60 years of age [3].

This tumor was named chordoid glioma because of its distinctive histologic appearance, reminiscent of chordoma, and its avid staining with glial fibrillary acidic protein (GFAP) in immunohistochemical analyses [4]. After a cross-referenced PubMed search that yielded 79 published cases, we present one illustrative case and review the diagnostic criteria, treatment options and prognosis.

2. Case presentation

A 13-year-old girl presented with 2 weeks of progressive intermittent holocranial headaches, with no specific behavioral alterations. Magnetic resonance imaging (MRI) showed a well-defined lesion adjacent to the floor of the third ventricle, with little surrounding edema. The initial diagnosis was intraventricular meningioma, which was resected in 2003 at a regional hospital in Sao Paulo, Brazil.

In 2011, a new resection was performed due to lesion regrowth. The new histological diagnosis was gliosarcoma. An oncologist conducted 28 sessions of adjuvant radiotherapy (50, 4 Gy). Postoperatively, the patient evolved with diabetes insipidus (DI) and hypothyroidism. A follow-up with serial MRI was conducted and three months after this surgery, the patient began experiencing convulsive seizures.

In 2012, tumor regrowth in the hypothalamic region was detected via MRI. At this time, the patient presented with behavioral alterations characterized by psychomotor impairment, despite a Karnofsky score of 90 and an Eastern Cooperative Oncology Group (ECOG) score of 1 (Fig. 1).

When the patient was then referred to our oncology unit, we opted for a total transcortical resection guided by neuronavigation (Fig. 2). The material from the second surgery was sent to a different laboratory for a second histological diagnosis, along with material from the third surgery (Fig. 3). The anatomo-pathological report revealed a CG of the third ventricle. Postoperatively, the patient had improved psychomotor function and mild mental confusion, and maintained her previous Karnofsky and ECOG scores (90 and 1, respectively).

3. Discussion

3.1. Clinical presentation

Initial presentations of CG vary from asymptomatic to aggressive, and most cases show slow and mild progression. Specific symptoms are attributable to the specific tumor location and to the involvement of adjacent structures responsible for possible endocrine alterations, visual deficits and behavioral alterations,

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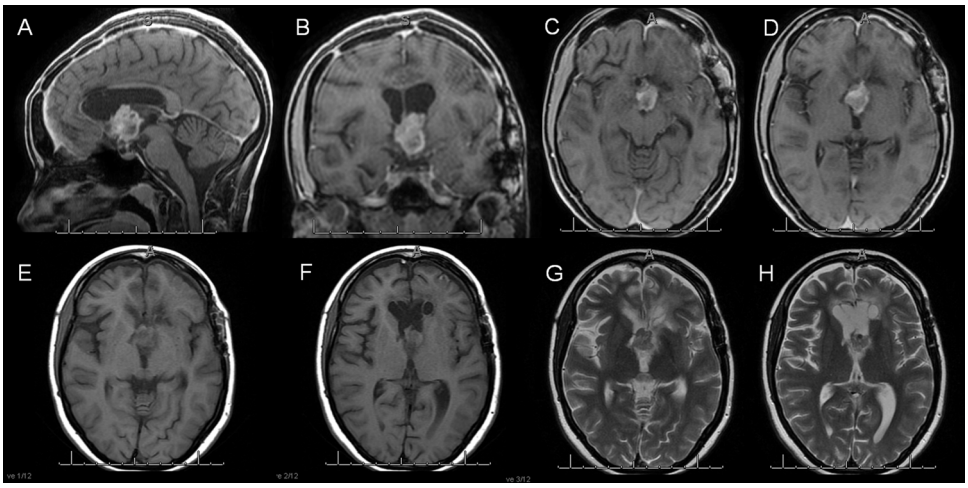

Fig. 1. Coronal, sagittal and axial T_1 gadolinium-contrast and T_2 axial images showing a tumor in the 3rd ventricle.

as well as symptoms secondary to an obstructive hydrocephalus [4–6].

In the case reported herein, the patient initially presented with a headache and as the tumor progressed, developed behavioral and endocrine complications.

3.2. *Differential diagnoses*

Suprasellar masses (including pituitary macroadenomas, craniopharyngiomas, meningiomas, metastases, optic and hypothalamic pilocytic astrocytomas) account for more than 75% of these lesions. Tumors of the third ventricle constitute an uncommon subset of suprasellar masses, and offer differential diagnostic value, excluding meningiomas, craniopharyngioma, hypothalamic gliomas, hypothalamic hamartoma, germ cell tumors, and others [7,8].

Meningioma is among these differential diagnoses, which was our patient's initial diagnosis. Kobayashi et al [2] also reported three cases with initial diagnoses of ventricular meningioma. These neoplasms had histologic appearance of cords and clusters of epithelioid cells within a mucinous background, along with a low-grade lymphoplasmacytic infiltrate reminiscent of a chordoma or chordoid meningioma. Unlike meningiomas, which are not glial in origin, all CG stained avidly for the glial cell marker GFAP [2,9].

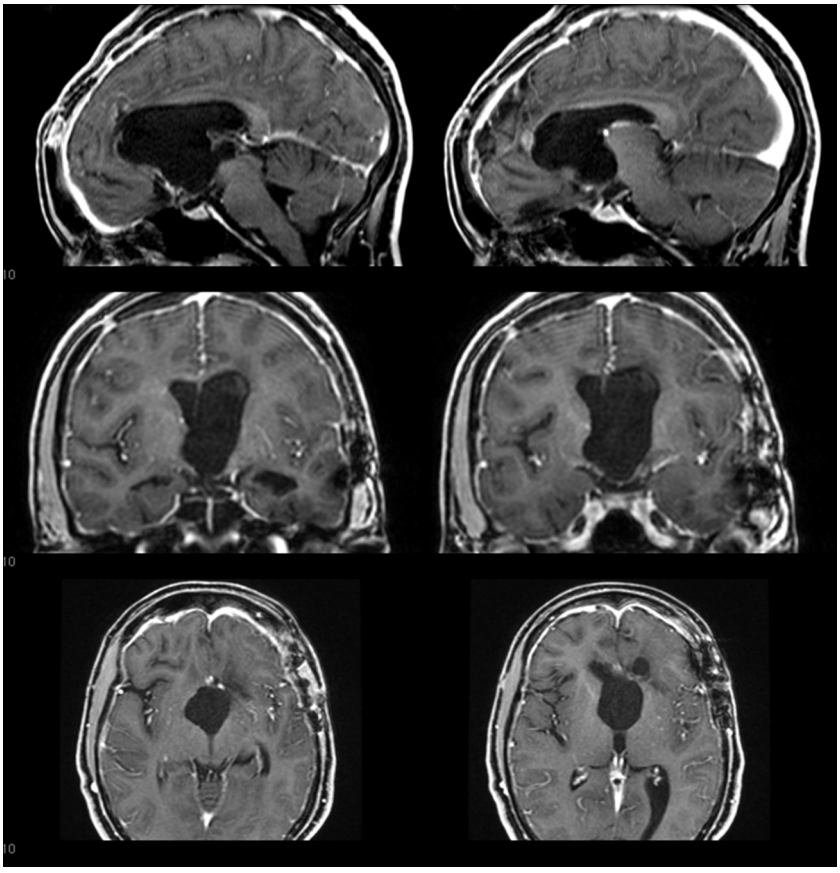

Fig. 2. Immediate Postoperative coronal, sagittal and axial T_1 gadolinium-contrast images.