CHORDOID GLIOMA OF THIRD VENTRICLE: A CASE REPORT AND A REVIEW OF CURRENT LITERATURE.

AUTHORS:
Lucas Piason de Freitas Martins, MBBS¹; Renato Teixeira Conceição Junior, MBBS¹; Vitor Nagai Yamaki, MD²; Davi Jorge Fontoura Solla, MD²; Iuri Santana Neville, Ph.D²,³

1: Escola Bahiana de Medicina e Saúde Pública (EBMSP), Salvador, Bahia, Brazil.
2: Division of Neurosurgery, Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo (USP), São Paulo, São Paulo, Brazil.
3: Division of Neurosurgery, Instituto do Câncer do Estado de São Paulo (ICESP), São Paulo, São Paulo, Brazil.
DISCLOSURES

• I have read the AANS/NREF/NPA Corporate Guidelines and AANS/NREF/NPA Conflict of Interest Policy, and understand that my decisions as an AANS/NREF/NPA Board, Committee Member, speaker, content planner or Staff must conform to this policy.

• I have read the AANS Non-Discrimination, Anti-Retaliation, and Harassment Policy and AANS Policy Regarding Harassment and Disruptive Behavior at Meetings and Courses and understand that as an AANS/NREF/NPA Board, Committee Member, speaker, content planner or Staff, I need to be aware of this policy and will report any perceived violations to the AANS Executive Director or to AANS/NREF/NPA legal counsel.

• I have read and I understand and agree with the AANS/NREF/NPA Content Validation Statements and CME Policies.

• I DO NOT have any financial or organizational relationships with commercial interests or other entities. I hereby certify that to the best of my knowledge, no aspect of my current personal or professional circumstances places me in the position of having a conflict of interest with my duties, responsibilities and exercise of independent judgement as an Officer, Member of the Board of Directors, Nominee for Office, Educational Presenter and/or a representative of AANS/NREF/NPA.

• I have read and understood the Personal Information Consent. I consent to the AANS retaining personal information about me and using such information to send emails and other communications to me. I further consent to the AANS sharing my personal information with its third-party vendors, when necessary.

• I consente with the Recording Release and with the Final Statement Response.
INTRODUCTION

Chordoid Glioma Of Third Ventricle is a rare entity with a variable clinical presentation and a challenging treatment. Chordoid Glioma’s (CG) features was first described by Wanschitz et al. in 1995, but it was first named by Brat et al. only in 1998 with 8 case reports, and was incorporated into the World Health Organization (WHO) classification as grade 2, in 2000. 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. Since then, a few case reports have been published.

CG is now known as a slow-growing, non-invasive primary neuroepithelial tumor of central nervous system, but its origin is still an incognate. Initial presentations vary from asymptomatic to severe symptoms, depending on the degree of obstruction and compression to adjacent structures. The most common symptoms are visual impairment, headache, nausea, vomiting, endocrine alterations and behavioral changes. The outcomes of gross total resection are often unfavorable despite its benign behavior.

We performed a systematic review to address different aspects of this rare neoplasm.
A 45 year old, female, presented with acute hidrocefalous (figure 1) in January 2019 at the emergency room and underwent a right frontal VP shunt procedure to improve her symptoms. Prior to admission, the patient was left-sided paretic and had been having focal seizures three times a week, since June 2018. She performed a MRI that showed a large mass well-demarcated arising from the supra-sellar area and occupying the third ventricle (figure 2). A stereotactic biopsy was performed in February 2019 and the diagnosis of Chordoid Glioma of Third Ventricle was confirmed. She was referred to our department, Instituto do Câncer do Estado de São Paulo (ICESP), and we repeated the MRI exam (figure 3) one month later, that showed an important growth of the tumor. It was heterogenously enhenced by contrast on T1 weighted sequence, with cystic area and it was isointense to grey matter on T2 weighted sequence.

In July 2019, we performed a interhemispheric approach and a partial resection was achieved, with a small residual tumor remaining, confirmed by post-operative MRI (figure 4). The post-op recovery was prolonged because the patient developed sepsis and hydroelectrolytic disorders due to central diabetes insipidus. She was readmitted twice on account of diabetes insipidus and seizures. She was, then, transferred to the oncology service from ICESP, but no adjuvante tratment has been done so far.
METHODS

The terms “Chordoid” and “Glioma” were searched at PubMed and EMBASE databases. All reports in English and Spanish were included in this study. The exclusion criteria considered were as follow: Only abstract available, technical notes, multiple publications, non English or Spanish papers, not sited on third ventricle, non Chordoid Glioma or without immunohistochemical analysis. The data was collected by two independent researchers. Of all, 63 case reports were selected and then analysed. The variables of gender, age of onset, signs and symptoms on presentation, kind of approach, use of adjuvant therapy, patient outcomes, time of follow-up and recurrency were collected and recorded in a secure database.

A descriptive analysis of the frequencies of variables was performed using PAST 3.0.
RESULTS

We included 63 case reports with 89 patients. The mean age of diagnosis was 45.6 (±11.5) years with prevalence of women. The most common symptoms were headache (46%), visual impairments (39%), mental status alterations (25%), and endocrine disorders (19%). 43 (48.3%) patients were treated with a complete resection approach while 41 (46%) had a partial resection approach. Only 10 (11.2%) patients underwent to radiotherapy treatment. The recurrence rate found was 13.4%. The mean follow-up period was about 19.6 (±19.2) months. The major post-operative complications presented in 15 (16.8%) patients while the mortality rate was 11.2%.
DISCUSSION

Efficient treatment of chordoid glioma depends upon radical surgical resection. However, this entity is often associated with high perioperative mortality due its topography and adherence to the surrounding structures. Thus, the outcomes are often unfavorable, despite of its grade II classification.