CASO CLÍNICO/CASE REPORT

Incidental Third Ventricular Chordoid Glioma: Case Report

Glioma Cordóide do Terceiro Ventrículo Incidental: Relato de Caso Clínico Raro

Lídia Nunes Dias [1,*], João Pedro Oliveira [1], João Paulo Andrade [1], Sérgio Figueiredo [1]
1-Department of Neurosurgery / Hospital Egas Moniz – Centro Hospitalar Lisboa Ocidental, Lisboa, Portugal

DOI: https://doi.org/10.46531/sinapse/CC/200071/2021

Abstract

Chordoid glioma is a grade II (World Health Organization) rare type of brain tumor, frequently arising within the anterior part of third ventricle and/or suprasellar region, which can cause symptoms from obstructive hydrocephalus, local compression (as hypothalamic disfunction) or intracranial hypertension.

We report an incidental case of 57-years-old woman, scheduled to a neurosurgery appointment due to an infundibular tumor, found on requested image after presenting unusual headaches and facial paresthesia. The tumor was totally resected by microsurgery via right pterional/trans-lamina terminalis approach without any postoperative morbidity. Histological analysis confirmed the unexpected chordoid glioma diagnosis.

Chordoid glioma of the third ventricle is a rare tumor, which ideal treatment is surgical gross-total tumor removal. However, as it can carry a high risk of postoperative complications due to its location, one should make a careful and well-planed treatment decision on any case, especially in incidental and/or oligosymptomatic ones.

Resumo

O glioma cordóide é um tumor cerebral de grau II (Organização Mundial da Saúde), normalmente localizado no terceiro ventrículo, podendo causar sintomatologia de hidrocefalia obstrutiva, compressão local ou hipertensão intracraniana.

Apresentamos um caso incidental de uma mulher de 57 anos, com o diagnóstico imagiológico sugestivo de tumor infundibular, exame realizado no contexto de cefaleias incomuns e parestesias faciais recentes. Foi proposta a cirurgia e o tumor foi ressecado totalmente por cirurgia transcraniana com abordagem pterional direita, sem qualquer morbilidade pós-operatória. A análise histológica confirmou um diagnóstico pouco esperado.

O glioma cordóide do terceiro ventrículo é um tumor raro, cujo tratamento ideal é a exérese cirúrgica total. Apesar de ter um comportamento benigno, pode acarretar um alto risco de complicações pós-operatórias devido à sua localização, pelo que a decisão de tratamento deve ser tomada de forma cuidadosa, principalmente nos casos incidentais e/ou oligossintomáticos.

Informações/Informations:
Caso Clínico, publicado em Sinapse, Volume 21, Número 1, janeiro-março 2021. Versão eletrónica em www.sinapse.pt
Case Report, published in Sinapse, Volume 21, Number 1, january-march 2021. Electronic version in www.sinapse.pt

Keywords:
Cerebral Ventricle Neoplasms/surgery;
Glioma/surgery;
Third Ventricle.

Palavras-chave:
Glioma/cirurgia;
Neoplasias do Ventrículo Cerebral/cirurgia;
Terceiro Ventrículo.

***Autor Correspondente / Corresponding Author:**
Lídia Nunes Dias
Hospital Egas Moniz
Serviço de Neurocirurgia
Rua da Junqueira, 126
1349 - 019 Lisboa, Portugal
lidiandias.ld@gmail.com

Recebido / Received: *2020-12-07*
Aceite / Accepted: *2021-01-24*
Publicado / Published: *2021-04-16*

Introduction

Chordoid glioma is a grade II (World Health Organization) rare type of brain tumor, frequently arising within the anterior part of third ventricle and suprasellar region, which clinical, radiological and pathological features are usually pleomorphic and can mimic other (and more common) type of lesions.[1]

Case Report

We report an incidental case of 57-years-old woman, scheduled to a neurosurgery appointment after presenting recent unusual headaches and facial paresthesia. She brought an magnetic resonance imaging (MRI) showing an infundibular well-circumscribed mass **(Fig. 1)** and normal endocrinological pituitary function analysis. The tumor was apparently totally resected by microsurgery via right pterional / trans-lamina terminalis approach without any postoperative morbidity. She was release from our department 5 days after surgery.

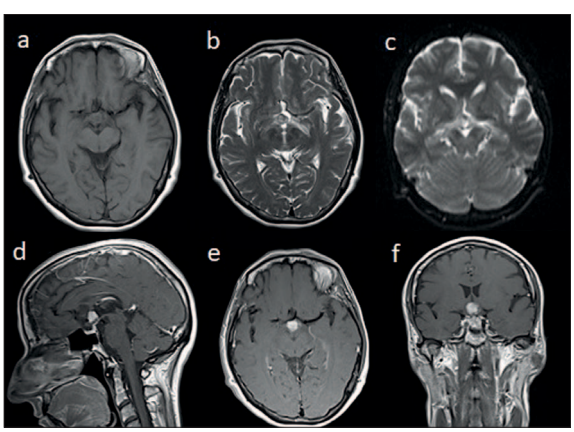

Figure 1. Pre-operative MRI.

a) T1 axial showing an isointense tumor, as well on T2 (b) and no water diffusion (c). After gadolinium the tumor was highly and homogenous hyperintense (d, e, f).

The resected specimen consisted in fragments of a partly well-defined neoplasm, composed of cells with a regular nucleus and vast, eosinophilic cytoplasm, organized in cords and trabeculae within a myxoid stroma. There was a moderate lymphoid infiltrate, generally perivascular, with germinal centers, some plasmocytes with Russel bodies and areas of fibrosis. No mitosis or necrosis was observed. Immunohistochemical profile consisted in positivity for: GFAP, CD34, TTF1; doubtful S100 and EMA; negativity for AE1AE3 and IDH1. Ki67 was less than 3%. All supported the unexpected chordoid glioma diagnosis **(Fig. 2)**.

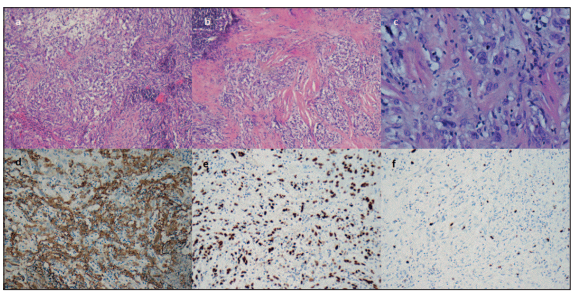

Figure 2. Pathology analysis.

Pathology analysis showed a), b) (both in 100x) and c) (400x) fragments of a partly well-defined neoplasm, composed of cells with a regular nucleus and vast, eosinophilic cytoplasm, organized in cords and trabeculae within a myxoid stroma. There was a moderate lymphoid infiltrate, generally perivascular, with germinal centers, some plasmocytes with Russel bodies and areas of fibrosis. Immunohistochemical profile consisted in positivity for: GFAP (d) and TTF1 (c). Ki67 was less than 3% (f).

Postoperative MRI **(Fig. 3)** showed no tumor and the patient remains clinically silent and recovered.

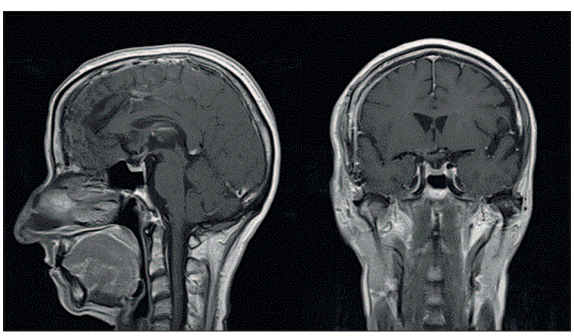

Figure 3. Postoperative MRI – SAG and CORONAL T1 + gadolinium showed no tumor or complications.

Brief Review of the Literature and Discussion

Chordoid glioma (CG) is a rare type of brain tumor, frequently arising within the third ventricle, originally described as a distinct meningioma variant by Wanschitz et al in 19952, and, in 1998, as a "third ventricle chordoid glioma" by Brat *et al*.[3] However, only in the 2000 version of World Health Organization (WHO) classification of tumors of the central nervous system it was categorized as a novel grade II tumor entity named "chordoid glioma of the third ventricle".[4]

Its histogenesis remains uncertain and the current hypotheses states a glial origin from the ependymal cells of the lamina terminalis and/or from the multipotent stem cell of Rathke's pouch.[1] It seems that, to date, less than 100 cases of chordoid glioma have been reported, only three cases outside the third ventricle.[1]

The clinical picture can vary from being completely asymptomatic to space-occupying general and/or local effect symptoms as headache, visual changes, memory

deficits, endocrinal disturbances (due to hypothalamic dysfunction)[5] or obstructive hydrocephalus symptoms, namely headache, nausea, vomiting, and ataxia.[1,5]

Enhanced brain MRI is the standard diagnostic imaging which usually shows a well-circumscribed, round or oval lesion, iso or hypointense in T1 and slight hyperintense in T2-weighted sequences. After gadolinium injections, they can present strong homogenous enhancement or heterogeneously due to cysts, necrosis or even rarer calcifications.[1,6] Diffusion-weighted image (DWI) frequently shows no diffusion restriction and spectroscopy (MRS) elevated choline and reduce N-acetyl aspartate (NAA) values. Additionally, (obstructive) hydrocephalus can accompanied the tumor.[1]

All CG share similar pathological findings, including ovoid or polygonal epithelioid cells with abundant cytoplasm organized in clusters and chords, typically found within a mucinous vacuolated stroma.[5] Immunohistochemistry shows positivity to GFAP, EMA, CD 34, cytokeratin, S100 and vimentin, more commonly. It can also be found some lymphocytic infiltrate. Mitosis are consistently low as well as proliferation indexes (low Ki-67 percentages) without necrosis.[5]

Surgical resection is the mainstream treatment, however in the majority of the cases gross-total resection (GTR) is not possible, due to its location and tight adherences which makes *en bloc* resection difficult and dangerous, leading to a high postoperative morbidity risk (for hypothalamic-associated complications or panhypopituitarism).[5] Immediate postoperative mortality with this approach can be as high as 29%, and morbidity among survivors can reach 67%.[7] So, despite being a low-grade tumor, the prognosis can be quite poor.[6] When GTR cannot be accomplished, adjuvant radiotherapy or radiosurgery can and has been administered, however their optimal role still remains nuclear.[5]

It's interesting to verify that, in terms of morbidity and progression-free, surgical approach seems to be more important than the proper extent of resection.[5] Trans-lamina terminalis approach seems to be related to less adverse effects, comparing to transcortical and transcallosal approaches ($p=0.051$).[5]

Regarding the literature, our case carried out a very simple, benign and uncomplicated course, from the incidental and/or oligosymptomatic presentation, a very favorable radiological and intraoperative features and an uneventful surgical procedure (with apparent GTR) and postoperative period.

Conclusion

Chordoid glioma of the third ventricle is a rare, low-grade and slow-growing tumor, for which ideal treatment is total surgical removal of the tumor. However, as it can carry a high risk of postoperative (treatment-related) complications, mainly due to its location, one should make a careful decision whenever incidental and/or oligosymptomatic cases are found. ■

Acknowledgement / Agradecimento:

We would like to acknowledge and thank the contribution of Martinha Chorão (MD) from the Pathology Department of Hospital Egas Moniz, for the acquisition, analysis and interpretation of gross pathology images and/or photomicrographs presented in this article.

Responsabilidades Éticas

Conflitos de Interesse: Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.

Fontes de Financiamento: Não existiram fontes externas de financiamento para a realização deste artigo.

Confidencialidade dos Dados: Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.

Consentimento: Consentimento do doente para publicação obtido.

Proveniência e Revisão por Pares: Não comissionado; revisão externa por pares.

Ethical Disclosures

Conflicts of Interest: The authors have no conflicts of interest to declare.

Financing Support: This work has not received any contribution, grant or scholarship.

Confidentiality of Data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Patient Consent: Consent for publication was obtained.

Provenance and Peer Review: Not commissioned; externally peer reviewed.

References / Referências

1. Yang B, Yang C, Du J, Fang J, Li G, Wang S,et al. Chordoid glioma: an entity occurring not exclusively in the third ventricle. Neurosurg Rev. 2020;43:1315-22 10.1007/s10143-019-01161-w. doi:10.1007/s10143-019-01161-w
2. Wanschitz J, Schmidbauer M, Maier H, Rossler K, Vorkapic P, Budka H. Suprasellar meningioma with expression of glial fibrillary acidic protein: a peculiar variant. Acta Neuropathol. 1995; 90: 539–44
3. Brat DJ, Scheithauer BW, Staugaitis SM, Cortez SC, Brecher K, Burger PC. Third ventricular chordoid glioma: a distinct clinicopathologic entity. J Neuropathol Exp Neurol. 1998; 57:283–290
4. Gonzales M. The 2000 World Health Organization classification of tumors of the nervous system. J Clin Neurosci. 2001; 8:1–3. doi: 10.1054/jocn.2000.0829
5. Ampie L, Choy W, Lamano JB, Kesavabhotla K, Mao Q, Parsa AT, et al. Prognostic factors for recurrence and complications in the surgical management of primary chordoid gliomas: a systematic review of literature. Clin Neurol Neurosurg. 2015; 138:129–36. doi: 10.1016/j.clineuro.2015.08.011
6. Chen X, Zhang B, Pan S, Sun Q, Bian L. Chordoid glioma of the third ventricle: a case report and a treatment strategy to this rare tumor. Front Oncol. 2020; 10:502. doi:10.3389/fonc.2020.00502
7. Cunha PR, Rebelo O, Barbosa M. Chordoid glioma of the third ventricle, a rare tumor with an unexpected outcome. Arq Bras Neurocir. 2017;36:32–37