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Case Report

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Chordoid Glioma of Third Ventricle: About an Extensive and Rapidly Progressive Presentation

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Abstract

Chordoid glioma is a rare tumor of the Third Ventricle (V3), representing a very distinct clinico-pathological entity in the WHO classification of central nervous tumors, who classified it as grade II [1]. It has usually a slow growing. In this paper, we report the case of a rapidly progressive and lethal Chordoid Glioma (CG) in 48-year-old Moroccan a women. The rarity and the morphological similarities of this neoplasm with other suprasellar tumors constitute a challenge for the pathologist especially in extemporaneous condition, which complicates the diagnosis and the management.

Keywords: Chordoid glioma; Third ventricle; Low grade; Poor outcome

Abbreviations

CG: Chordoid Glioma; V3: Third Ventricle; WHO: World Health Organisation; GFAP: Glial Fibrillary Acidic Protein; CD34: Cluster Differentiation 34; EMA: Epithelial Membrane Antigen; TTF-1: Thyroid Transcription Factor-1; MRI: Magnetic Resonance Imaging

Introduction

Chodoid glioma is a rare and discrete glial tumor of Third Ventricle (V3) of adults, characterized by chordoid architecture and composed of oval-to-polygonal epitheloid cells with abundant eosinophilic cytoplasm organized in clusters and cords within a myxoid background. The cells may resemble ependyma of subcommissural organ, present in dorsocaudal third ventricle during embryonic life, which regresses after birth [2]. It's known as a slow-growing noninvasive tumor, but- through this case report- we confirm that it may be rapidly progressive, aggressive and lethal.

Case Report

We present the case of a rapidly progressive and lethal V3 chordoid glioma in a 48-year-old patient admitted to the emergency with intracranial hypertension, Left hemiparesis, opto-chiasmatic syndroma and frontal syndroma. Symptoms dated back to 15 days before admission. Brain MRI showed an expansive process (4 cm in diameter) of the antero-inferior wall of the V3 in favor of an ependymoma. The lesion marks intensely the contrast product, exerting a mass effect on the two holes of monro, the optical chiasm with reduction of the supra sellar cistern, the two capsulo-lenticular and thalamic regions (Figue1). The patient underwent partial surgical excision.

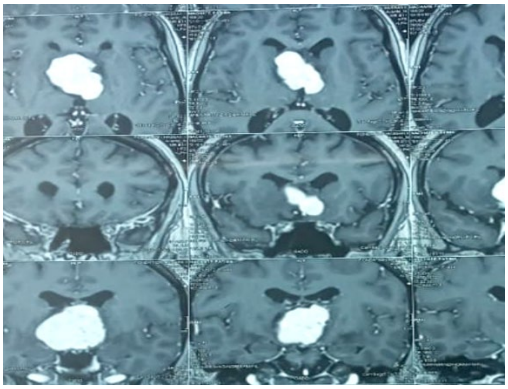

Figure 1: (A) Brain MRI shows an expansive process (4 cm in diameter) of the antero-inferior wall of the V3. (B) The lesion marks intensely the contrast product.

A sample was sent to our laboratory for extemporaneous analysis. The Smear showed a tumor proliferation of epithelioid architecture, supported by a fibrillar background, discussing an ependymoma, or a meningioma. Histological examination of the specimen showed a tumor proliferation with chordoid appearance, arranged in cords and nests. Tumor cells appear cohesive, with an abundant eosinophilic cytoplasm with a round nucleus, fine and nucleolated chromatin, without mitotic activity (Figure2). They develop on a myxoid and microcystic background harboring an important mononuclear inflammatory infiltrate which is rich in plasma cells with the presence of numerous Russell bodies (Figure 2).

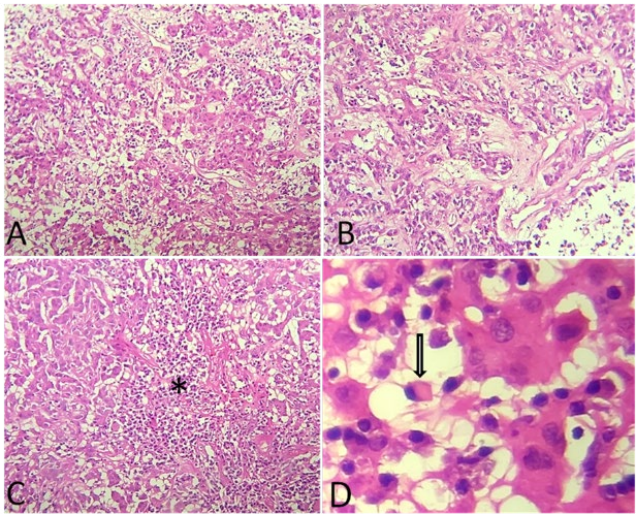

Figure 2: Histologicaly, (A) the tumor shows a chordoid appearance proliferation, arranged in cords and nests (Hematoxylin-eosin, original magnification x 10). (B) They develop on a myxoid and microcystic background with (C) lymphoplasmocytic infiltrate (star), (D) and numerous Russell bodies (arrow).

Immunostaining showed a strong expression of CD34, GFAP and vimentin by tumor cells, associated with a positive labeling of PS100, Pancytokerain AE1 / AE3 and EMA (Figure 3). Analysis for Thyroid Transcription Factor-1 (TTF-1) was not performed.

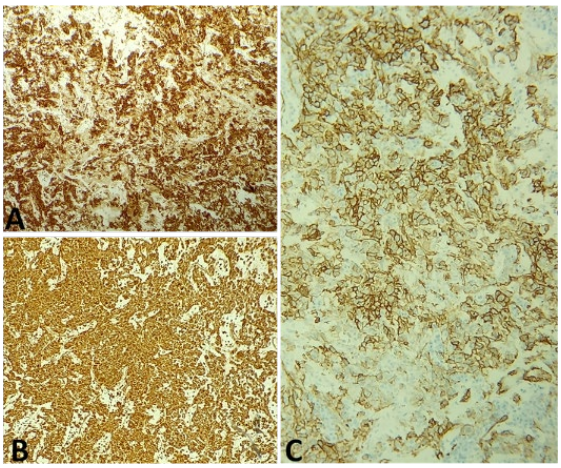

Figure 3: In immunohistochemical stain, the tumor cells expressed GFAP (A), Vimentin (B) and CD34 (C).

Thus, the diagnosis retained is a Chordoid Glioma of V3, grade II of WHO 2016. The patient died after the operation following hemorrhagic and ischemic complications.

Discussion and Conclusion

Chordoid Glioma (CG) is a rare central nervous system neoplasm (less than 100 cases reported in the literature to our Knowledge). It was first reported by Brat et al in 1998 as a new distinct histopathological tumor entity, and the World Health Organization (WHO) 2007 and 2016 classified it as a grade II neoplasm [1]. Non consensus has been retained on the histogenesis of the tumor, but a common hypothèses support a glial origin, particularly from the ependymal cells. Other hypotheses favor divergent neuronal, glial, circumventricular and subcommissural differentiation [1,2]. It's typically arising from the anterior well or roof of the third ventricle, which may adhere to the hypothalamus and extend to supra sellar structures 3. This topography associated with a clinical, radiological and histological polymorphism complicates its pre and intraoperative diagnosis which can be challenging [3]. Through this case, we point out the importance and usefulness of immunostaining in the positive diagnosis and elimination of other differential diagnosis namely ependymoma, pilocytic astrocytoma, chordoid meningioma or chordoma. Indeed, CG is characterized by a strong expression of Glial Fibrillary Acidic Protein (GFAP), CD34, and Vimentin suggesting an origin from tancycytes which are located in the subcomissural organs. In addition, most CGs contained small fractions of neoplastic cells immunoreactive for epithelial membrane antigen, S-100 protein, or cytokeratins [4,5]. Thyroid Transcription Factor-1 (TTF-1)

expression has also been proposed as a good differential marker for chordoid glioma, but it is not specific, this marker could help in the differentiation of atypical cases. However, the immunoreactivity for CD34 is most useful for making differential diagnosis between chordoid glioma and the others tumors which are negative for CD34 [4,5]. The clinical outcome is generally poor because of the location of the lesion and its close relation to the hypothalamus. Only complete excision favors a better outcome, although surgery in this area carries significant operative risks [5].

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Consent for Publication

Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient.

Competing Interests

The authors declare that they have no competing interests.

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