Case Report

An incidentally detected third ventricle chordoid glioma

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Chordoid glioma is a rare low-grade tumor located in the third ventricle-hypothalamic region. Since its first report, 37 cases have been described in the literature. We report on an additional case that we considered significant because of its incidental detection and its uneventful surgical removal.

Key words: Brain surgery, chordoid glioma, immunohistochemistry, third ventricle

Chordoid glioma (CG) of the third ventricle is a rare low-grade tumor described for the first time by Brat in 1998 as a distinct clinical-pathological entity.[1] The histopathological features of CG are consistent with a slowly growing neoplasm, thus successfully curable by surgery. However, due to its anatomical neighborhood, its prognosis is comparatively poor.

Case Report

A previously healthy 56-year-old woman was admitted after a head injury due to a road accident. Physical and neurological examination was unremarkable. Routine computerized tomography (CT) scans revealed a circumscribed, round, hypodense suprasellar mass. No hydrocephalus was detected. On contrast-enhanced magnetic resonance (MR) imaging, the lesion appeared well-circumscribed, solid and homogeneously enhanced without signs of mass effect [Figure 1]. Despite the little size of the lesion and the absence of neurological signs or symptoms and considering the advantage of obtaining histopathological diagnosis, surgical removal was proposed. A right fronto-temporal craniotomy was performed, the Sylvian fissure was split to expose the chiasmatic region and then the suprasellar cistern was widely opened. The nodule, located between the optic nerves, was first gently dissected from the chiasm and the right optic nerve. Once isolated from the anterior-superior wall of the third ventricle, it was removed “en bloc”. The postoperative course was totally uneventful.

Particularly, no signs of hypothalamic dysfunctions were observed. Postoperative MR imaging confirmed complete removal of the lesion and no further therapy was recommended. At the 48-month follow-up, the patient remained asymptomatic and MR imaging showed complete anatomical normalization of the area [Figure 2].

Histopathological findings

The lesion presented a fibrous capsule, sharply demarcating it from the parenchyma. It consisted of clusters and cords of epithelioid cells with abundant eosinophilic cytoplasm and isomorphic nuclei. Abundant lymphoplasmacytic infiltration was appreciable, particularly at the tumor-parenchyma interface. The lesion did not show necrosis and the mitoses were exceptional. Immunohistochemically, the tumor cells showed diffuse expression of glial fibrillary acidic protein (GFAP), neurofilaments (NF), vimentin.
(VIM), CD34 and a weaker expression of neuron specific enolase (NSE) [Figure 3].

**Discussion**

In the last revised edition of the WHO classification of the nervous system tumors (2000), CG has been incorporated under the category of glial tumors of uncertain origin.[2] It typically shows chordoma-like histological features and glial fibrillary acid immunoreactivity; it mainly occurs in middle-aged women and is usually found in the hypothalamic-suprasellar-third ventricle region. Chordoid glioma is usually revealed by signs of obstructive hydrocephalus, visual field loss, endocrine abnormalities and/or personality changes. Due to its small size and thus absence of compression on the neighboring structures, the lesion in the described case represents the only reported CG of incidental detection.

The precise anatomic origin of the lesion remains unclear but according to other authors the chordoid glioma seems to rise from the vicinity of the lamina terminalis and infundibular recess and such a hypothesis has been confirmed in our case by the anatomical exposure at the surgical time.[3]

Three cases described in the literature underwent a biopsy (open surgery, stereotaxy, endoscopy). All the CG lesions described in the literature were surgically treated through different approaches:pterional, interemispheric (trans lamina terminalis) and less frequently transcallosal or transphenoidal (in the latter case the lesion was initially taken for a pituitary adenoma). From the reports in which treatment and follow-up are available, in only 17 cases (including ours) complete removal was achieved.[4,5] Eight of these cases presented postoperative complications including severe hypothalamic disturbances that were transitory in five and lethal in three. In this group, within a 6-68 month follow-up, no cases of recurrence were observed and no adjuvant therapy was followed. Of 17 patients who underwent only partial surgical removal of the lesion seven presented complications that were lethal in two cases and transitory in five. In four of these cases the remnant was treated with external radiotherapy, with radiosurgery in three cases and in one case, the patient underwent re-surgery, external radiotherapy and radiosurgery. Within this group of partially removed lesions, six patients are well after a 9-42 month follow-up, while two died independently from the CG.

The CG radical removal that represents the recommended treatment is not easily reachable because of the strict closeness of critical structures. Moreover, surgery, also in cases of subtotal removal, often presents complications, even death. External radiotherapy does not seem to give benefits while the role of radiosurgery cannot be evaluated as it was practiced only in one case on the residue after surgery. Our case represents a relatively small asymptomatic tumor, and should support the early removal of lesions of the third ventricle needing diagnostic confirmation.

**References**


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