Chordoid glioma of the third ventricle attached to the optic chiasm
Successful removal through a trans-lamina terminalis approach

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Received 5 February 2008; received in revised form 6 May 2008; accepted 13 May 2008

Abstract

Chordoid glioma of the third ventricle constitutes a rare, very recently recognized histological entity. Most reports of this neoplasm, focused on its distinct histological features, have hypothesized about a probable origin of the lesion at the third ventricle floor and/or the lamina terminalis. We report on a new case, presenting neuroradiological and intraoperative pictorial evidences of the tumoral attachment, limited to the chiasm-lamina terminalis junction.

A 53-year-old woman debuted with acute symptoms of obstructive hydrocephalus, visual disturbances and confusion. MRI investigation showed a large solid-cystic third ventricle mass bulging through the lamina terminalis and ventricular floor. After placing a ventriculoperitoneal shunt, the tumor was completely removed through a trans-lamina terminalis approach. A tight tumoral attachment to the junction of the posterior chiasm to the lamina terminalis was identified and dissected. No other adhesions to the third ventricle boundaries were found. A chordoid glioma was diagnosed on histological examination. One year after the surgical procedure the patient does not present new neurological deficits, and there are no signs of tumoral regrowth on the follow-up postoperative MRI.

Chordoid glioma should be included in the differential diagnosis of third ventricle tumors. Preoperative neuroradiological suspicion of this lesion should alert the neurosurgeon about the presence of a tight tumoral adherence at the level of the chiasm-lamina terminalis junction. The trans-lamina terminalis approach provides a suitable route for an early control of this attachment under direct vision, allowing a safe dissection of the mass from the third ventricle.

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Keywords: Chordoid glioma; Third ventricle; Lamina terminalis; Intraventricular tumor

1. Introduction

Chordoid glioma of the third ventricle is a rare and relatively recently described neoplasm that has been assigned by the World Health Organization (WHO) to the group of “Tumors of Uncertain Origin”, as a glial grade II tumor [1,2]. Topographically, this tumor typically occupies the third ventricle-hypothalamic region as a solid, homogeneously enhancing mass, constituted by epithelial cells arranged in clusters and cords, embedded within a mucinous matrix [3,4]. Immunohistochemically, the lesion is characterized by its strong diffuse GFAP reactivity [5]. Due to the short number of cases reported to date, the exact origin and best surgical management for this lesion remain unclear [6–9]. Surprisingly, for a lesion considered as a low-grade glioma and operated under standard microsurgical techniques, the overall outcome is rather poor, with a postoperative mortality reaching the 25% of the cases, and permanent hypothalamic morbidity affecting one out of four survivors [8]. The recurrence rate is also high, involving nearly the 20% of patients. These figures are probably related more to the uncertainty about anatomical relationships and degree of adherence of this tumor than to its biological behaviour. We present neuroradiological and surgical evidences obtained from a successfully removed chordoid glioma showing a definite band of tight adherence to the lamina terminalis.
attachment to the optic chiasm-lamina terminalis junction. A trans-lamina terminalis approach allowed a safe detachment of the tumoral mass under continuous direct view, minimizing the potential distortions of the hypothalamic walls caused by surgical manipulations.

2. Case report

A 53-year-old woman was transferred from another hospital to our neurosurgical department, for surgical evaluation of a large tumoral lesion occupying the third ventricle area. The patient had symptoms for one year, when she complained of sudden visual disturbances, confusion and dizziness. A relative had noticed that she did not recognize her name during phone conversations. The ophthalmologic exploration revealed a superior bitemporal quadrantanopia without fundoscopy signs of papilledema. Endocrinologic tests evidenced reduced plasmatic estradiol levels and an increased prolactin value of 1.7 ng/ml. An obstructive hydrocephalus, caused by a tumor occupying the third ventricle cavity, was then observed in the MRI study, and a ventriculoperitoneal shunt was implanted with resolution of symptoms related to high intracranial pressure. On admission in our department, the patient was alert and oriented in time and place, and did not show focal neurological deficits. The ophthalmologic study showed no changes in her visual field deficit yet an increased latency in the visual evoked potentials of both eyes was observed. The endocrinologic investigations for pituitary functions were normal.

Computed tomography performed on admission revealed a 3 cm × 2.5 cm isodense mass centered at the third ventricle area, which enhanced homogeneously after contrast administration. No signs of active hydrocephalus were observed. On the high resolution 3 T MRI study a rounded and well-delimited lesion filling the third ventricle cavity was evidenced (Fig. 1A–B). It consisted mainly of a solid isointense mass on both T1 and T2 weighted images, associating some peripheral cystic regions that were hypo and hyperintense on T1 and T2 sequences respectively. The solid component enhanced homogeneously after Gadolinium administration. The morphology of the sellar region and pituitary gland was intact and the suprasellar cistern was free, but the pituitary stalk could not be visualized. The third ventricle floor limits were not identified but both the mamillary bodies and the optic chiasm were clearly depicted. Hyperintense T2 signals were observed along both optic tracts and also at the level of the third ventricle walls (Fig. 1A). Selective cerebral angiography of carotid and vertebral systems was performed, without evidence of significant neovascularization or displacement of anatomic vascular structures. The differential diagnosis of this lesion included the following lesions: optic-hypothalamic glioma, intraventricular cranio-pharyngioma and chordoid glioma of the third ventricle.

The lesion was surgically approached through a standard left pterional craniotomy. The choice of a left approach was made in order to gain access to the cystic component of the lesion. After opening the Sylvian fissure and completing the dissection of the optic-carotid area, there was no evidence of the tumoral mass at the suprasellar area. The optic chiasm presented a prefixed position and a bulging mass pushing forward through the thin wall of the lamina terminalis was appreciated (Fig. 2A). The lesion was then exposed through a trans-lamina terminalis approach and a tough pinkish mass abutting the third ventricle was disclosed. The lesion was well-delimited from the third ventricle walls but it showed a band of tight attachment to the posterosuperior edge of the chiasm (Fig. 2B). The tumoral mass was first debulked using microsurgical technique with bipolar forceps and pituitary curettes (Fig. 2C). Once it could be mobilized, the mass was released from its adherence to the chiasm-lamina terminalis junction by sharp dissection (Fig. 2D). Finally, the two cystic areas were emptied and the tumor was completely removed without damaging the surrounding hypothalamic structures (Fig. 2E–F). The pituitary stalk could not be identified during the surgical procedure.

The postoperative clinical course of the patient was good, showing only a transitory short memory deficit, associated with some episodes of disorientation, without any other focal neurological deficit. She initially presented panhypopituitarism, requiring total hormone replacement, as well as permanent diabetes insipidus treated with fluid restriction and intranasal vasopresin. Follow-up endocrinological test performed 2 months after the surgical procedure showed a recovery of the gonadotrophic and tirotrophic axes. The visual field deficit has not varied since the operation. The postoperative MRI study showed no remnants and the third ventricle morphology had recovered its normal appearance. The ventricle boundaries displayed hyperintense signals on T1 and T2 weighted sequences that probably correspond to reactive peritumoral gliotic areas (Fig. 1C–D). The third ventricle floor, the suprasellar cistern and the mamillary bodies were easily recognizable on the postoperative MRI, as well as the surgically disrupted lamina terminalis.

Histologically the tumor consisted of a homogeneous proliferation of eosinophilic epithelioid-like cells arranged in cords and clusters, which were linked by cytoplasmic extensions and were embedded in a myxoid stroma (Fig. 3A). Prominent lymphoplasmocytic infiltrate areas were found among the epithelial cell clusters. There was no evidence of histological malignancy, such as nuclear atypia, mitotic activity or necrosis. Neither infiltration of normal parenchyma was observed among the pathologic samples studied. From an immunohistochemical standpoint, the tumoral cells showed strong and diffuse reactivity to glial fibrillary acidic protein (GFAP) and slight focal reactivity to neurofilaments (NF) and epithelial membrane antigen (EMA) (Fig. 3B–C). S-100 protein, carcinoembryonic antigen (CEA), synaptophysin and chromogranin immunostaining were all negative. The tumoral proliferation index estimated by MIB-1 labelling was less than 5%. The final pathological diagnosis was chordoid glioma of the third ventricle.
3. Discussion

The chordoid glioma of the third ventricle was defined as a new pathological entity in 1998 by Brat et al. [1], although the tumor reported by Wanschitz et al. in 1995 [10], that was diagnosed as a third ventricle meningioma with atypical GFAP positive immunoreactivity, would probably correspond to a chordoid glioma. The WHO 2000 CNS tumor classification [2] included this neoplasm in the group of glial tumors of uncertain origin. Typically, the tumor shows the microscopic features of a low growing lesion, without signs of malignancy, such as necrosis, anaplasia or neovascular proliferation, therefore it has been ascribed as a WHO grade II. Macroscopically, chordoid glioma is a solid tumor composed of epithelioid eosinophilic cells arranged in cords and clusters which are embedded within a mucinous basophilic matrix with scattered areas of lymphoplasmacytic infiltrates. Immunohistochemically, the most distinctive finding is the strong diffuse reactivity to GFAP shown by the tumor [2]. The staining for vimentin and CD34 is usually diffuse and strong, whereas cytokeratin, S-100 protein and EMA immunoreactivity are observed inconstantly. The absence of neuronal markers (synaptophysin, neurofilaments, neuron-specific enolase and chromogranin) is also a common characteristic. However our case is the third one presenting a positive immunostaining for neurofilaments, together with the lesions reported by Buccoliero et al. [11] and Gallina et al. [12]. This fact would support a divergent glial and neuronal differentiation within this tumoral type. As a consequence, the presence of neuronal markers should not exclude the chordoid glioma from the pathological differential diagnosis of any tumoral mass located at the third ventricle [11].

The histogenesis of chordoid glioma remains unknown. Several studies have claimed that this lesion would be originated from immature ependymal cells of the third ventricle [9,13]. This hypothesis has been supported by the regular description of this tumor as developing along the antero-inferior ependymal lining of the third ventricle, including the lamina terminalis, the infundibular recess and the median eminence. Some morphological and histological features of the lesion have linked its origin to two of the known circumventricular organs: the organum vasculosum of the lamina terminalis and the subfornical organ. These structures lack the property of blood brain barrier and contain a specialized type of ependymal cells, the tanyocytes, which show, unlike ordinary ependymocytes, strongly positive reactivity for GFAP [7,14]. The usual appearance of the chordoid glioma as an intraventricular, rounded mass that shows an intense
homogeneous Gadolinium enhancement on MRI, but is poorly vascularized on angiography, provides additional support for this hypothesis. Although some cases with small size are circumscribed to the antero-inferior part of the third ventricle and displays an ellipsoid morphology oriented along the vertical axis of the lamina terminalis [11,12,15–19], many tumors have occupied the whole third ventricle area when diagnosed, it being impossible to identify neither the anatomical third ventricle boundaries nor the exact site of tumoral origin [20–23].

Given the uncertainty about the exact site and cell type from which chordoid glioma is originated, those evidences...
obtained at the surgical field about the anatomical relationships between the tumor and the third ventricle boundaries are of paramount importance. Most of the cases of this entity reported so far focus the discussion on the histological features of the tumor, without offering a detailed description of the findings observed during the surgical procedure. In the case we report on a horizontal gliotic band of tight attachment that could be identified between the antero-inferior tumoral surface and the junction of the lamina terminalis with the optic chiasm. Apart from this area, the tumoral mass showed no other adhesions to the third ventricle boundaries, and it could be easily removed from the ventricular cavity once this gliotic attachment was freed by sharp dissection. Our surgical findings provide additional evidence about the specific site of origin of chordoid glioma at the lamina terminalis area, as depicted in the intraoperative microphotographs (Fig. 2). However, it is not clear whether this finding can be generalized, as other authors have noted more extensive attachments along the third ventricle walls, precluding a total resection [20].

Tumoral lesions growing mainly at the third ventricle can be removed through any of the three following surgical routes: frontal transventricular, interhemispheric transcallosal and trans-lamina terminalis approach [24,25]. There are some advantages and possible complications associated with each one. The opening of the lamina terminalis can be used in combination with any surgical exploration of the suprasellar area performed through either a subfrontal or a pterional approach [26]. This surgical strategy is specially suited for small and moderate sized lesions centered at the antero-inferior third ventricle area [27]. It allows a safe exploration of the mass as well as its dissection from the optic chiasm and the third ventricle walls under permanent visual control. For larger lesions occupying the whole third ventricle or the upper third ventricular area, at the region of the foramina of Monro, the transcallosal approach provides a wider exposure. Nevertheless if the lesion is attached at the floor of the third ventricle, the surgical manipulations during its removal without direct control of the tumor attachment, may cause harmful distortions on the hypothalamic nuclei. Therefore, it is of paramount importance when choosing the surgical approach to know preoperatively the typical location and extent of attachment for every pathological entity occupying the third ventricle. Most of the data provided so far about the site of origin of the chordoid glioma point to an anterior-inferior attachment at the third ventricle. As the most dangerous surgical maneuvers would occur during the dissection of the mass from the chiasm and the anterior-inferior recesses of the third ventricle, which are limited by the hypothalamic nuclei, we think that the trans-lamina terminalis approach is especially appropriated for this lesion, even when approaching large sized tumors. As a support of our opinion, a null surgical-related mortality has been reported among those chordoid gliomas operated on through the trans-lamina terminalis approach (n = 13 cases) [3,12,18,28–32]. As a contrast, a 75% postoperative mortality has been observed among the patients operated through a frontal transventricular approach (n = 4 cases) [5,33–35] and a 33% mortality was found in the subgroup of patients approached through a transcallosal route (n = 6 cases) [8,15,20–23]. Although the final decision about the surgical route to be chosen for the removal of such a rare lesion will be based on the neurosurgeons’ experience and preferences, the analysis of the global outcome from the cases reported so far should alert about the specific risks associated with each type of approach, taking into account the characteristic adhesion shown by this type of tumor [36].
References


