

Case Report

Chordoid Glioma : A Case Report of Unusual Location and Neuroradiological Characteristics

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Since the World Health Organization (WHO) classification for central nervous system neoplasms was declared in 2000, chordoid glioma of the third ventricle has been noted as a newly recognized tumor for central nervous system neoplasms. Although there is not enough universal experience to know the nature of this tumor due to its rarity, the origin of chordoid glioma was guardedly proposed to be the ependymal cells of the third ventricle. Such an idea has been primarily based on the specific location of the tumor, that is, third ventricle, suprasellae, and hypothalamus. However, we report a rare case of histologically confirmed chordoid glioma located in the left thalamus, not attached to any of the midline structures having unusual neuroradiological characteristics.

KEY WORDS : Chordoid glioma · Third ventricle · Radiological feature.

INTRODUCTION

In 1998, Brat et al.²⁾ introduced a tumor of the third ventricle presenting with both glial and chordoid features, and a new clinicopathological entity was named as "chordoid glioma of the third ventricle". From then on, approximately 50 cases of chordoid glioma, to our best knowledge, have been reported^{11-5,7,8,11-18,20-23,25,28)}. Chordoid glioma is a rare low-grade tumor of the brain, most of which are known to be located in the third ventricle, frequently attached to the hypothalamic or suprasellar area.

Though this neoplasm is currently incorporated into the WHO classification of gliomas according to the context mentioned above, the histogenesis of the tumor is still unclear. Some previous reports have suggested that this tumor might arise from ependymal cells of the subcommissural organ^{6,17,19,25)}. However, we present a case of a chordoid glioma of the left thalamus without involvement of the structures of third ventricle and suprasellae.

CASE REPORT

A 27-year-old woman with a two-month history of worsening headache and visual disturbance underwent computed tomography (CT) as recommended by a primary physician. A low-density lesion in left thalamus associated with mild hydrocephalus was found. The patient was then referred to our institution. She had no significant past medical history. Neurological examination revealed a visual field defect on the left temporal side on confrontation test. However, ophthalmologic examination was normal. Magnetic resonance (MR) images demonstrated a well-defined large mass in the left thalamus. The tumor showed high signal intensity on T2-weighted images, and low signal on T1-weighted images (Fig. 1). It was entirely located in the left thalamus, compressing the third ventricle, without any involvement of the third ventricle, hypothalamus and suprasellar region. After gadolinium injection, scattered dot-like subtle enhancement was found (Fig. 1). Since the tumor was presumed to be a diffuse glioma, MR spectroscopy and ¹⁸F-FDG-positron emission tomography (PET) were performed.

With single-voxel MR spectroscopy with a point-resolved spectroscopy (PRESS) sequence (echo time 288 ms), high choline, low N-acetyl aspartate (NAA) and equivocal lactate were detected (Fig. 2). A choline peak was highest in the central portion of the tumor and so the central portion was tho-

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ught to be composed of the tumor cells with higher histological grade. Brain ^{18}F FDG-PET demonstrated diffuse hypometabolism of the tumor with scattered foci of intermediate metabolism within the tumor in the left posterior thalamus.

The patient underwent a left fronto-temporal craniotomy using a transcortical approach via the inferior temporal gyrus. Near total resection of the tumor was performed in spite of the difficult tumor location. Post-operative pre-contrast CT demonstrated no unusual findings. In addition, her neurological status was not changed after the surgery. Unfortunately, on the third day after the surgery, the patient had respiratory failure of unknown cause. Despite efforts to resuscitate her, she died on the 10th post-operative day.

Pathological findings

Surgical specimens were fixed in formalin and embedded in paraffin. Hematoxylin-eosin (HE), periodic acid-Schiff (PAS), D-PAS, Alcian blue, and Masson's trichrome stains were performed. In addition, a broad range of immunohistochemical staining was performed. Histologically, the tumor showed clusters and cords of epithelioid cells, abundant myxoid (mucinous) stroma, well developed capillary network, mild nuclear pleomorphism, no mitosis, no vascular endothelial hyperplasia, and no necrosis (Fig. 3A). However, prominent lymphoplasmacytic infiltration with Russell bodies was exceptionally

absent in this case.

Immunohistochemically, the tumor cells showed diffuse expression of glial fibrillary acidic protein (GFAP), vimentin, S-100 protein, neuron-specific enolase (NSE), CD56, epi-

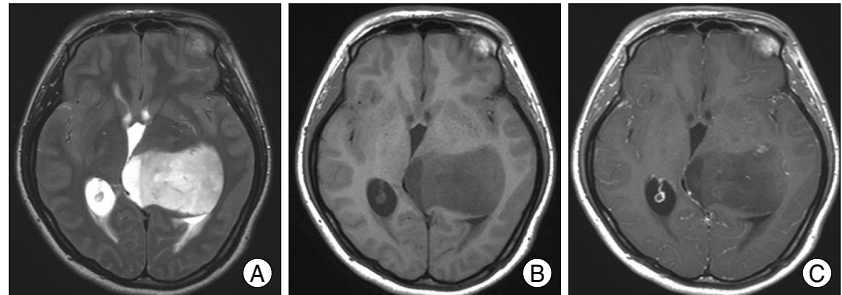


Fig. 1. Magnetic resonance images of the tumor. A : T2-weighted image shows a well-defined high signal mass in the left thalamic pulvinar area without any involvement of the third ventricle. B : T1-weighted image shows low signal intensity of the mass. C : Gadolinium-enhanced T1-weighted image shows scattered dot-like subtle enhancement of the tumor, suggestive of the possibility of intermediate grade glioma.

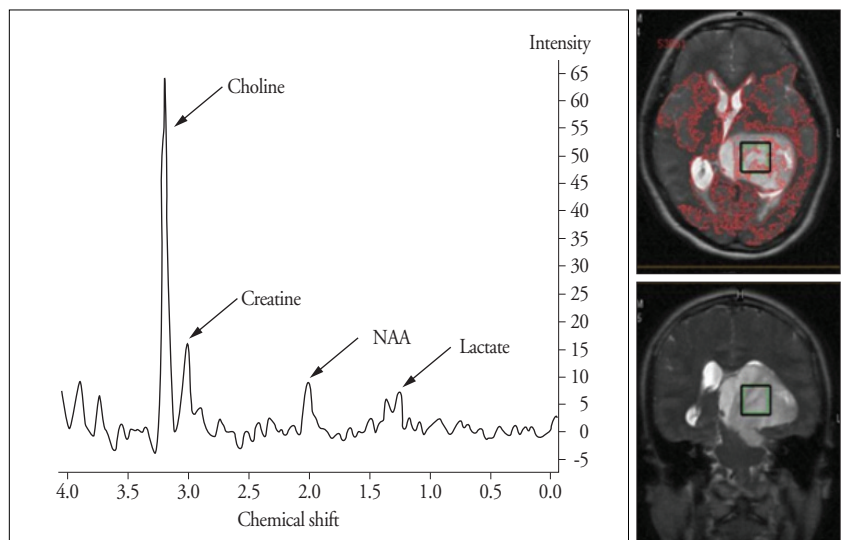


Fig. 2. Magnetic resonance spectroscopy. Single-voxel MR spectroscopy performed in the central portion of the tumor shows high choline, low N-acetyl aspartate (NAA), and equivocal lactate. The box on the tumor indicates the voxel.

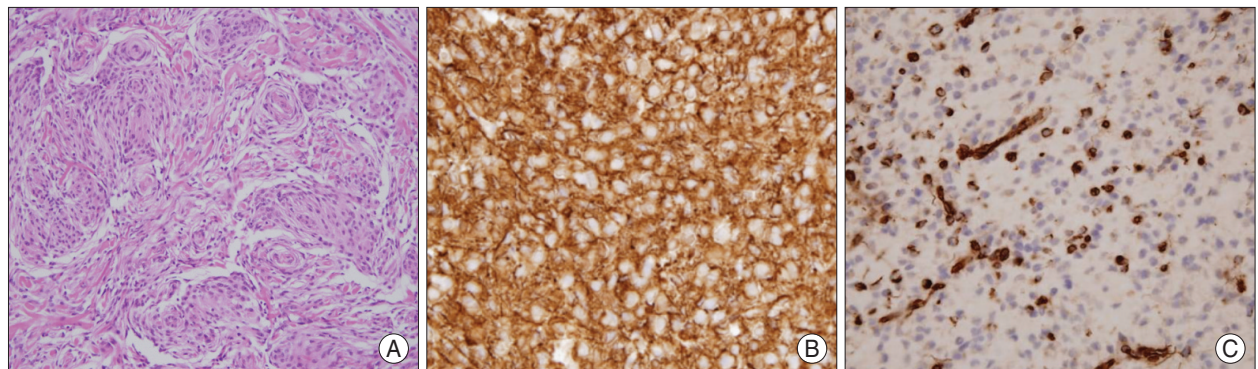


Fig. 3. Histological findings. A : Hematoxylin and eosin staining revealed clusters and cords of epithelioid cells, abundant myxoid and mucinous stroma, well developed capillary network, mild nuclear pleomorphism, no mitosis, no vascular endothelial hyperplasia, and no necrosis (H & E, $\times 100$). B and C : Photographs of immunohistochemical stainings demonstrating diffuse cytoplasmic expression for glial fibrillary acidic protein (B) and vimentin (C) ($\times 200$).

thelial membrane antigen (EMA), and cytokeratin, also with platelet-derived growth factor receptor-alpha (PDGFR-alpha), and glucose transporter-1 (Glut-1) (Fig. 3B, C). But, expressions of neurofilament and synaptophysin were absent. Based on the pathological findings above, the tumor was diagnosed as chordoid glioma^{3,5,10,11,17,26,27}.

DISCUSSION

Chordoid glioma of the third ventricle was incorporated into WHO classification in 2000. The name itself included the stereotypical location of the tumor. It seemed suitable to name it "chordoid glioma of the third ventricle" because, according to the literature at that time, all of the chordoid gliomas were located in the third ventricle, hypothalamus and suprasellae^{2,19,24,25}. However, to our best knowledge, approximately 50 cases have been reported so far, which inevitably limits the scope of understanding this rare disease.

Radiological findings from neuroimaging studies of this tumor were also consistent and typical, in that they characteristically exhibit a well-circumscribed border, uniform contrast enhancement, and a fusiform gross morphology^{2,19,27,28}. On the histological side, it was cautiously suggested that the tumor showed features of ependymal differentiation^{6,19,23,25}. Taken altogether, it was even proposed that the tumor be renamed "chordoid ependymoma of the lamina terminalis"^{6,19,23,26}. It was inferred likewise, not only due to the ultrastructural findings, but also due to its unique midline location.

However, there are some differences between our case and typical chordoid glioma of the third ventricle. This case is not associated with any of the midline anatomical landmarks that were mentioned above : third ventricle, hypothalamus, and suprasellae. It is also different from others in that the mass was not uniformly enhanced, rather only focally enhanced from a radiological standpoint. Due to the rarity of this tumor, it can be risky to form a hasty conclusion on the characteristics and on the origin of this tumor. Our case only suggests a good possibility that this tumor entity is composed of heterogeneous subgroups. Archiving more cases of the kind, and correctly identifying their location, histology, neuroimaging findings, and clinical course, are warranted to define this relatively new neoplasm entity.

As previously described, despite the successful surgery and favourable recovery process, the patient had respiratory failure of unknown cause on the third day after the surgery. Such a clinical course is in line with previous reports. Vanhauwaert et al.²⁸ reported overall immediately postoperative mortality rate was about 30% through the literature reviews. They were considered to have had a thromboembolism or to suffer from hypothalamic injury. Poor prognosis might be

due to the tumor location in the deep structure of the brain, making it difficult to approach surgically. The possible cause of death and unfortunate poor prognosis of our case is consistent with previously reported cases^{9,11,17,25,28}.

CONCLUSION

We have presented a case of a chordoid glioma that has unusual location and neuroradiological characteristics.

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