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One of a kind—chordoid glioma in the fourth ventricle: a case report and literature review

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Abstract

Chordoid glioma (CG) is a rare brain tumor that is known for its characteristic location in the third ventricle. A wide spectrum of radiological presentations has been described, with few common features among them. Its radiological diagnosis is mainly suggested by location. However, several cases of CG with atypical locations have been described, illustrating that CG is not limited to the third ventricle, and should be considered in the list of radiological differential diagnosis for intraventricular masses. We present here a case of CG that was found in the fourth ventricle.

Keywords

Chordoid glioma, intraventricular tumor, fourth ventricle, magnetic resonance imaging

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Introduction

Due to its characteristic location in the hypothalamus and anterior third ventricle, chordoid glioma (CG) was officially named "chordoid glioma of the third ventricle" in the 2016 World Health Organization classification.[1,2] Four cases of CGs outside of the third ventricle have been reported.[3–6] Here, we describe a case of CG in the fourth ventricle, which has not been reported in the literature to the best of our knowledge.

Case presentation

Clinical information

A 53-year-old Chinese woman presented with an insidious onset of unsteady gait for 3 years, with no other complaints. She was previously healthy except for mild chronic rhinitis. She failed heel-toe walking during physical examination, but had no other neurological signs. Her blood tests came back normal. There was also no family history of malignancies.

Radiological findings

Magnetic resonance imaging (MRI) revealed a 3 cm × 2.9 cm × 2.9 cm intraventricular cystic lesion in the fourth ventricle which was causing moderate hydrocephalus (Fig. 1). The lesion was hypointense on T1-weighted images, and hyperintense on T2-weighted images, and showed ring enhancement with mural nodules. An intralesional T2W hypointense dependent fluid level with susceptibility artifact suggested history of tumoral hemorrhage. Restricted diffusion was absent. MRI of the whole spine showed no cerebrospinal fluid tumor seedling or synchronous spinal cord tumor.

The initial working diagnosis was intraventricular metastasis versus primary brain tumor. A whole body[17] FDG PET-CT subsequently excluded metastasis, and tumor resection was planned.

Surgical and pathological findings

Due to the tumor's tight adhesion to the anterior fourth ventricular wall, only subtotal tumor resection

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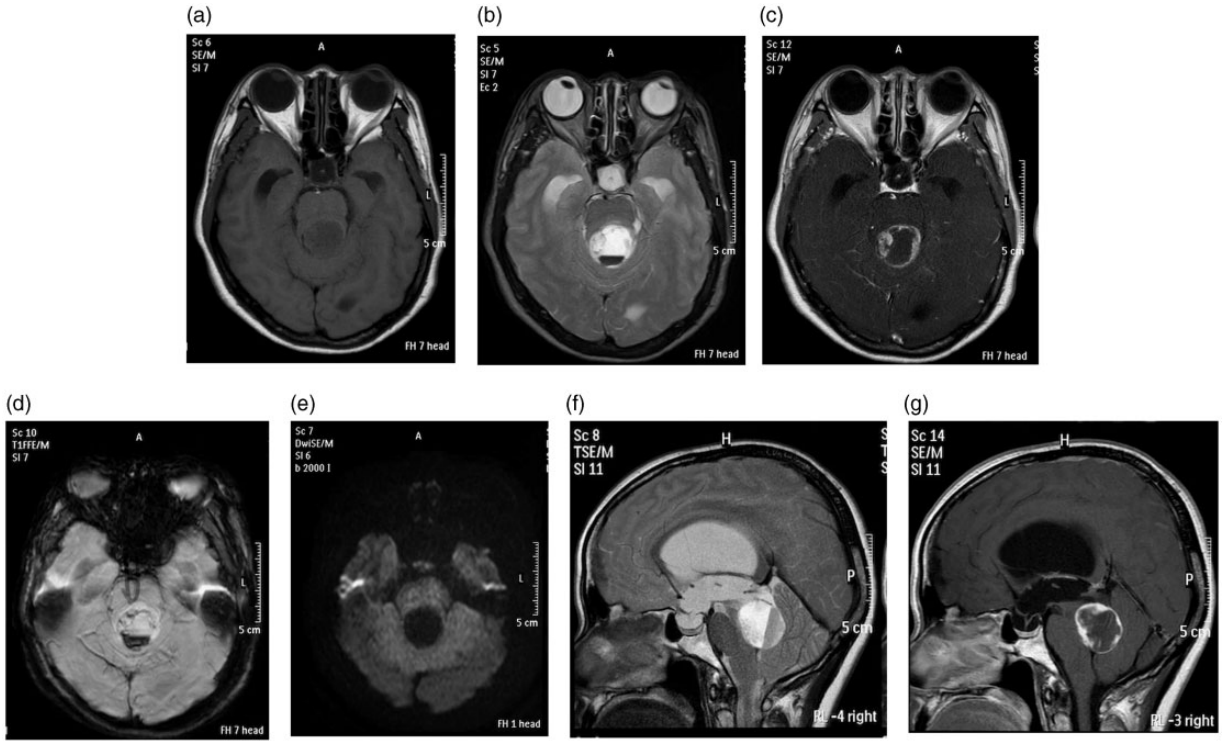

Fig. 1. (a and b) MRI study shows a T1-weighted hypointense, T2-weighted hyperintense intraventricular lesion in the fourth ventricle, complicated by moderate obstructive hydrocephalus. (c and g) Ring enhancement and mural nodules are seen. (d and f) Intralesional fluid–fluid level with susceptibility artifacts indicates tumor hemorrhage. (e) No restricted diffusion is demonstrated.

was achieved during surgery. No choroid invasion was detected intra-operatively.

Histopathological examination (Fig. 2(a)) showed cords and clusters of eosinophilic tumor cells in a myxoid background, with mild lymphocytic infiltrate. Pericellular clearing reminiscent of chondroid pattern was focally observed. There was no abnormal mitotic activity, microvascular proliferation, or necrosis. Immunohistochemical analysis showed positive staining for S-100, Olig2 (Fig. 2(b)), vimentin, and CAM5.2. EMA (epithelial membrane antigen) was focal. Staining was negative for glial fibrillary acidic protein (GFAP), CD34, PR, brachyury (Fig. 2(c)), neurofilament protein, synaptophysin, and somatostatin receptor (SSTR) (Fig. 2(d)). INI1 staining was preserved. IDH1 and IDH2 mutation were absent.

Chordoid meningioma, chordoma, chondrosarcoma, and CG were considered as pathological differential diagnoses. Chordoid meningioma was unlikely given absence of a typical meningothelial proliferation and negative SSTR staining. Chordoma and chondrosarcoma were also unfavored on account of the immunoprofile. A final diagnosis of CG was deduced, despite a negative GFAP.

Follow-up findings

Our patient had a short course of radiation therapy after her surgery. Follow-up MRI studies at 6 months, 1 year, and 2 years post-resection showed a residual ~1 cm intraventricular tumor occupying the lower part of the cerebral aqueduct (Fig. 3). The lesion remained stable in size to date – 2 years after the surgery. Clinically, the patient underwent an unremarkable recovery, walking unaided a year after the surgery and free from neurological symptoms.

Discussion

CG was first described in 1995 by Wanschitz et al., but was at that time considered a peculiar variation of meningioma that expresses GFAP.[7] In 1998, Brat et al. described CG as a distinct clinicopathologic entity, based on its mixed glial and chordoid features.[2] CG is now recognized by WHO as a WHO grade II slow growing, well-circumscribed brain tumor.[8] Globally, less than 100 cases of CG have been reported, therefore there has not been a universal understanding on its epidemiology or pathophysiology. It is observed

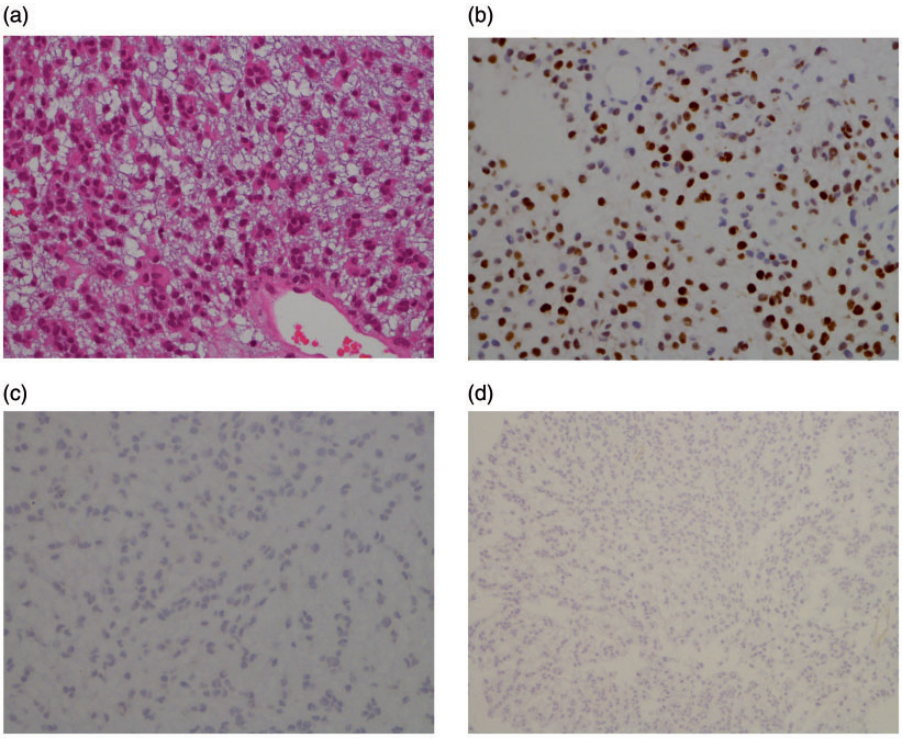

Fig. 2. Photomicrographs of the tumor. (a) Hematoxylin and Eosin stained slides show cords and clusters of eosinophilic abundant tumor cytoplasm on myxoid basophilic matrix. (b) Immunostained slide for glial nucleic marker Olig2 shows strong expression in neoplastic cells. (c) Negative immunostaining for Brachyury, a sensitive and fairly specific diagnostic marker for chordoma. (d) Immunostaining for somatostatin receptor (SSTR) is also negative.

that GC occurs mostly in adults (mean age 47), having a female predominance.[9] Five pediatric cases of GC have been reported in the literature at the time of this report.[3]

The clinical manifestations of CG depend on its location. As CG is characteristically at or near the third ventricle, patients most commonly present with obstructive hydrocephalus. More location specific symptoms that have been reported include endocrine imbalance, visual disturbances, behavior alterations, and autonomic dysfunction.[10]

Our patient is a good representation of the commonest patient group and clinical presentation; however, her CG has a peculiar location at the fourth ventricle. Apart from our case, four other cases of CG locating outside of the third ventricle have been reported, being at the frontal horn of the right lateral ventricle,[3] the left thalamus,[4] the occipital horn of the right lateral ventricle,[5] and the left temporoparietal region, respectively.[6] Three of these atypical cases are from pediatric patients.

Radiological assessment

CG is often a well-circumscribed iso- to hyper-dense oval mass on computed tomography (CT), with its greatest diameter expected in the craniocaudal direction.[11] An extension into the hypothalamus is usual.[12,13] The tumor shows homogeneous contrast enhancement. Tumor apoplexy and calcification are rare findings, and were observed only three times in all reported cases.[6,14–17]

Like in all brain tumor assessments, MRI is a superior imaging modality. CG is a solid tumor on MRI, with cystic components observed in 26% cases, mostly at the periphery. T1-weighted and T2-weighted signals are highly variable. Fan Chen et al. reviewed 86 published cases of CG, and summarized that they can be isointense (48.15%), hyperintense (25.93%), hypointense (22.22%), or heterogeneous (3.70%) on T1-weighted images; and hyperintense (50.00%), isointense (25.00%), hypointense (15.63%), or heterogeneous (9.38%) on T2-weighted images. Significant gadolinium contrast enhancement is common and usually homogenous (75%) due to a blood–brain barrier breakdown.[3,13] No restricted diffusion has been observed in all reported cases.

Achieving a diagnosis of CG by imaging alone is often challenging, as CG has variable radiological presentations and overlapping features with other tumors. Other common intraventricular tumors at the fourth

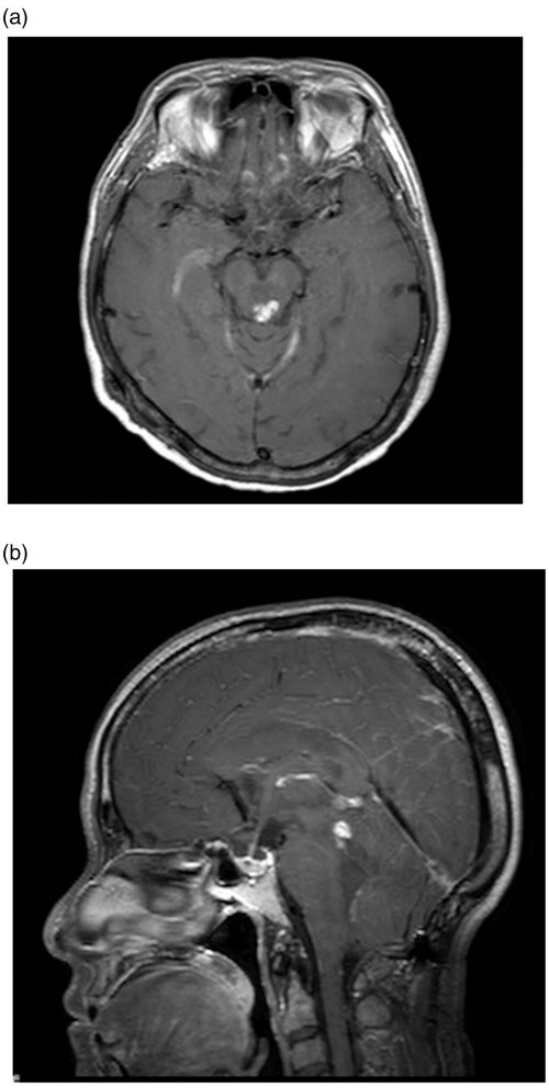

Fig. 3. Two-year postoperative MRI shows ~1 cm residual intraventricular tumor tissue at lower part of the cerebral aqueduct.

ventricle include ependymomas, subependymomas, pilocytic astrocytomas, meningiomas, and choroid plexus tumors. More age-specific differentials include medulloblastoma in pediatrics, and intraventricular metastasis and lymphoma in older adults. In our case, the cystic component of the tumor, lack of restricted diffusion, and close proximity to the posterior third ventricle could be clues to diagnosing CG. Currently, based on the few cases of CG of atypical locations, no radiological difference has been identified from those arising at the third ventricle.

Pathological characteristics

CG is fundamentally a histopathological diagnosis, supported by a coherent immunohistochemistry. Typical histopathologic findings of CG include: (1) clusters and cords of epithelioid tumor cells with abundant eosinophilic cytoplasm, embedded in an abundant PAS (periodic acid-Schiff stain)-positive myxoid and mucinous stroma, (2) prominent lymphoplasmacytic infiltrates and Russell bodies throughout the tumor, (3) rare or absent high-grade features such as mitotic activity and anaplastic features (nuclear polymorphism, vascular endothelial proliferation, necrosis), and (4) minor tendency to infiltrate the surrounding brain parenchyma.[1,2,18]

Immunohistochemically, CGs are usually strongly and diffusely positive for GFAP and vimentin. The positive rate for EMA, S-100 protein, CD34, synaptophysin, cytochrome, and neuron specific enolase was reported at 79.38%, 78.26%, 97.56%, 7.69%, 90.91%, and 50% respectively.[3,19] In our case, the negative GFAP could reflect a possible origin from a distinct subtype of glial cells—Olig2-lineage glial cells—which are much more abundant than GFAP astrocytes in the brainstem.[20]

On-going debates on whether CG is of astrocytic or ependymal origin mean a lack of a defining immunohistochemical profile. This may finally change after Goode et al. identified a recurrent PRKCA D463H oncogene mutation in 2018, shedding light to a possible genotypic identification for this uncommon tumor.[21]

Treatment and prognosis

Although being a low grade tumor, CG carries a poor prognosis because of its close vicinity to vital structures. Surgical resection aiming at gross tumor resection is the treatment of choice for CG, with repeated resections or adjuvant gamma knife radiosurgery often needed for incompletely resected tumors.[22] Postoperative mortality reaches 29%, with complications (cognitive, optical, endocrinological, and thromboembolic) seen among 67% of its survivors.[23] The treatment outcome is most affected by tumor location and surgical approaches.

No chemotherapeutic regimen has yet been proven effective on CG.

In conclusion, CG is a difficult diagnosis for radiologists to commit on. Its typical proximity to the third ventricle is a major hint. However, we should keep in mind that CG is not limited to the third ventricle, and it should be a part of the differential diagnoses for intraventricular masses. In our opinion, sagittal contrast enhanced MRI images may provide the most helpful information to the diagnosis of CG, by directly evaluating the proximity and infiltration of the lesion to the third ventricle.

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