

Case Report

Primary diffuse large B-cell lymphoma of the choroid plexus: A case report and review of the literature

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Abstract

Background: Primary lymphomas in the choroid plexus are much less frequent than primary lymphomas in the brain parenchyma.

Case Description: A 66-year-old male patient was referred to our department with a right intraventricular mass that had been diagnosed by biopsy at another hospital as anaplastic ependymoma. The patient underwent subtotal removal of the tumor via a transcortical inferior temporal gyrus approach. The mass was attached to the choroid plexus in the right atrium. Histopathological examination showed diffuse large B-cell lymphoma. Ophthalmological examination, blood tests, computed tomography of the whole body, and bone marrow biopsy did not show any other lesion, leading to the diagnosis of primary choroid plexus lymphoma. The patient underwent chemotherapy with three courses of high-dose methotrexate and one course of carboplatin and etoposide followed by whole-brain irradiation (1.8 Gy × 22).

Conclusion: We present a rare case of primary choroid plexus lymphoma, which should be considered in the differential diagnosis of choroid plexus tumors.

Key Words: Diffuse large B-cell lymphoma, lateral ventricle, primary choroid plexus lymphoma

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INTRODUCTION

Primary central nervous system lymphoma (PCNSL) is typically a diffuse large B-cell lymphoma that accounts for about 2.4–3% of all central nervous system (CNS) tumors with an overall annual incidence rate of 0.47 cases per 100,000 population.^[10] The median age at diagnosis of patients with PCNSL is 56 years. More than 60% of PCNSL patients present with cognitive, motor, or constitutional symptoms; 30% with cerebellar or visual symptoms; and 20% with seizures.^[5] PCNSL is a more common diagnosis in patients with rapidly deteriorating neurological conditions who undergo brain biopsy.^[5]

PCNSL frequently presents as a solitary mass with profound peritumoral edema. The supratentorial compartment is involved in about 60% of PCNSLs,

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whereas the posterior fossa or spinal cord is less frequently affected.^[10] The most frequent locations are: the frontal lobe (15% of cases), temporal lobe (8%), parietal lobe (7%), followed by basal ganglia and periventricular brain parenchyma (10%).^[10] PCNSLs have rarely shown to originate in the ventricles. Here, we report a case of lymphoma that primarily involved the choroid plexus in the right lateral ventricle.

CASE DESCRIPTION

A 66-year-old male patient was referred to our department with a right intraventricular mass that had been diagnosed as anaplastic ependymoma on open biopsy at another hospital.

He had been followed because of diabetes mellitus, hyperlipidemia, and carotid artery stenosis but had been otherwise well. Two months before admission to our department, he had undergone magnetic resonance imaging (MRI) of the head, which incidentally revealed a mass lesion in the right lateral ventricle, for which he had been referred to the neurosurgery department at another hospital. He underwent a biopsy and the tumor was diagnosed as an anaplastic ependymoma. Although he had been initially referred to a Radiation Oncology Department for adjuvant radiation therapy, the provider referred him to our Department of Neurosurgery because of concerns that the residual tumor was not suitable for radiotherapy without prior cytoreduction.

Physical examination showed some short-term memory impairment, and ophthalmological examination revealed a visual field compromise (quadrantanopia) likely related to access corridor chosen for the biopsy. There was no focal motor weakness, major sensory deficit, dysmetria, or ataxia.

Computed tomography (CT) showed two hyperdense foci in the inferior horn and the atrium of the right lateral ventricle with peri-lesional edema of the ipsilateral temporo-occipital parenchyma [Figure 1a and b]. The intraventricular lesions showed heterogeneous enhancement on postcontrast T1-weighted imaging (T1WI), on which the lesions appeared to arise from the choroid plexus [Figure 1c–f]. The postcontrast T1WI also showed additional tumor dissemination in the left lateral ventricle and around the brain stem [Figure 1c]. Further whole-spine imaging showed leptomeningeal dissemination in the lumbar region (not shown).

The patient was taken to surgery and underwent subtotal removal of the tumor in the right lateral ventricle via an inferior temporal gyrus approach [Figure 2a and b]. The tumor was solid, densely adherent to the choroid plexus but floating in the ventricle [Figure 3a].

Histopathologically, the tumor showed diffuse, tightly packed monotonous neoplastic cells with large, round,

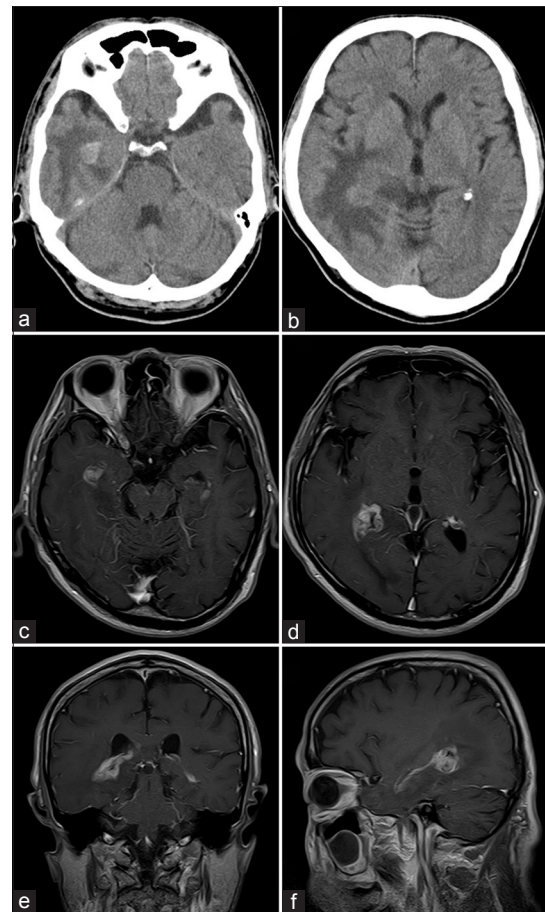


Figure 1: Preoperative images. Axial CT of the head (a and b) and postcontrast T1WI in the axial plane (c and d), coronal plane (e), and sagittal plane (f). Note that intraventricular mass lesions exist along the choroid plexus

or slightly irregular nuclei and a scant to moderate amount of eosinophilic cytoplasm [Figure 3b]. There were numerous mitotic figures and foci of focal necrosis. Perivascular pseudorosettes were not discernible. On immunohistochemical staining panels, the tumor cells were positive for leukocyte common antigen (LCA), CD20 [Figure 3c], CD30, CD79a, bcl-2, bcl-6 [Figure 3d], and MUM-1; and negative for cytokeratins, epithelial membrane antigen, S-100 protein, and glial fibrillary acidic protein.

Ophthalmological examination showed no intraocular lesions. CT of the chest, abdomen, and pelvis was interpreted as normal and bone marrow biopsy showed no atypical lymphocytic infiltrate. The patient was thus diagnosed with primary diffuse large B-cell CNS lymphoma likely originating in the choroid plexus.

The patient underwent chemotherapy with high-dose methotrexate (3500 mg/m²) every 14 days for three courses, followed by a course of chemotherapy with carboplatin (360 mg calculated by using the Calvert formula) and etoposide (80 mg/m²) 14 days after the third administration of high-dose methotrexate. After

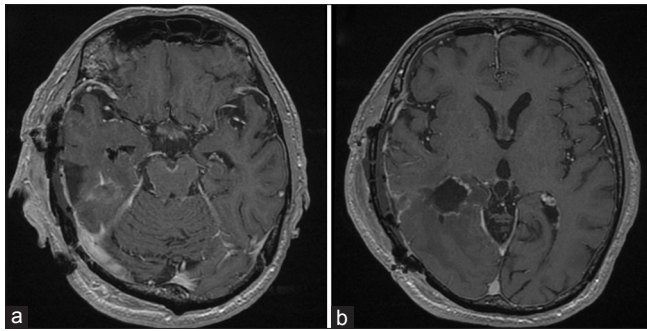


Figure 2: Postoperative images. Axial postcontrast T1WI (a and b) showing subtotal removal of the intraventricular mass lesions

chemotherapy, cerebrospinal fluid (CSF) samples were obtained for cytology, which was negative for malignant cells. After the patient had undergone whole-brain irradiation (1.8 Gy \times 22), postcontrast T1WI showed complete resolution of all previously visible CNS disease. Physical examination showed persistent quadrantanopia—unchanged from the patient's status prior to subtotal removal of tumor—and amelioration of his short-term memory impairment. Because meningeal dissemination recurred during the follow-up period, the patient is currently again receiving chemotherapy with high-dose methotrexate in our department (8 months after surgery).

DISCUSSION

Primary lymphomas in the brain have been reported to occur as single or multiple space-occupying lesions that usually occur in the parenchyma, are deep-seated and adjacent to the ventricular system. Lymphomas originating in the choroid plexus itself are rare. To our knowledge, this is only the seventh reported case of such primary choroid plexus lymphoma.^[3,4,6,7,15,17]

The choroid plexus is a lobulated structure found in the walls of the ventricles. It has a central highly vascularized stroma surrounded by neuroectodermal cells.^[2] The capillaries of the stroma of the choroid plexus allow the passage of fluid and lyophilic molecules, and the cells of the choroid epithelium secrete CSF.^[12] The choroid plexus secretes factors that guide neural tissue development and it has recently been discovered to contain stem cells.^[2]

More typical choroid plexus tumors consist of choroid plexus papilloma, atypical choroid plexus papilloma, and choroid plexus carcinoma, accounting for as little as 0.77% of all brain tumors and 14% of those occurring in the first year of life.^[2] The average annual incidence is 0.03 cases per 100,000 population.^[9] Among these, choroid plexus papilloma is the most common subtype, accounting for 58.2% of choroid plexus tumors.^[2] Approximately 80% of lateral ventricular choroid plexus tumors present in patients aged <20 years.^[9] Although

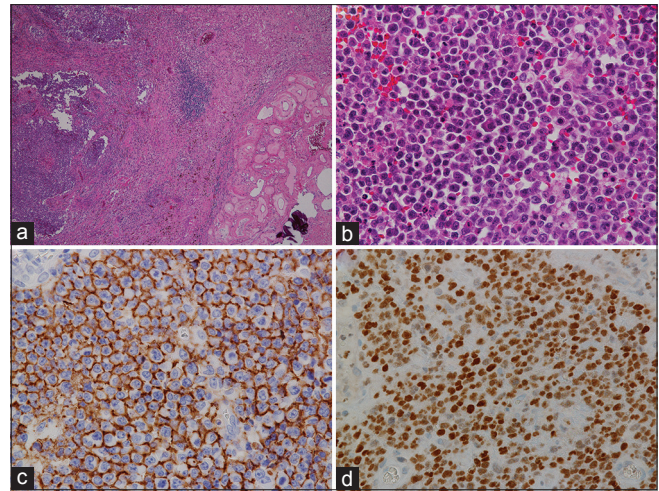


Figure 3: Histopathological features of the tumor. Hematoxylin and eosin staining at 20 \times magnification (a) and 400 \times (b), and immunohistochemistry for CD20 at 400 \times (c) and bcl-6 at 400 \times (d). Note that the tumor is attached to the choroid plexus (on the right lower part) in (a)

dozens of cases of metastatic choroid plexus tumors and a large number of intraventricular meningiomas attached to the choroid plexus have been reported, there have been only few previous reports of primary choroid plexus lymphoma.^[3,4,6-8,11,15,17]

In all reported cases of primary choroid plexus lymphoma, as well as in our case, the tumors were located in the lateral ventricle [Table 1]. Five of the patients were male and two were female; all were older than 50 years (53–73 years), except for a 27-year-old male. Patients with primary choroid plexus lymphoma presented with various combinations of headache ($n = 3$), seizures ($n = 2$), cognitive dysfunction ($n = 3$), and focal neurologic deficit ($n = 2$). Although our patient had leptomeningeal dissemination, physical examination showed no eye movement abnormalities or any other cranial nerve deficit; similarly, no ocular abnormalities were observed in any of the previously reported cases of primary choroid plexus lymphoma. Four of the patients underwent surgical resection and chemotherapy, one underwent surgical resection, one underwent external ventricular drainage and chemotherapy, and one underwent radiation therapy. In two patients who did not undergo surgical resection, diagnosis was made by CSF examination. Four patients remained disease-free for at least 1 year (12–26 months). The majority of PCNSL cases (>95%) are diffuse large B-cell lymphomas.^[1] In contrast, three of the cases of primary choroid plexus lymphoma were classified as extranodal marginal zone B-cell lymphomas of mucosa-associated lymphoid tissue (MALT), three were diffuse large B-cell lymphomas, and one was a T-cell lymphoma [Table 2]. The phenotypes of primary choroid plexus lymphomas are somewhat similar to those of primary lymphomas of the dura mater, because the

Table 1: Clinical data on primary choroid plexus lymphoma as described in the literature

Reference	Age (years)	Sex	Presentation	Location	Treatment	Follow-up
Kelley <i>et al.</i> ^[7]	53	Male	Headache, seizure	Right atrium	Resection, intrathecal liposomal cytarabine	14 months (alive)
Jung <i>et al.</i> ^[6]	63	Male	Seizure, transient monoparesis of the left leg, disoriented mentality	Right atrium	Resection, MTX	NM
Terasaki <i>et al.</i> ^[17]	56	Male	Headache (2 days)	Right inferior horn	MTX, 40 Gy WB RT, 10 Gy local RT	19 months (alive)
Cecchi <i>et al.</i> ^[3]	71	Female	Confusion, motor speech disturbance, left hemiparesis (2 weeks)	Right atrium	Resection, MTX	26 months (alive)
Cheatle <i>et al.</i> ^[4]	27	Male	Nausea, vomiting, headache (10 days)	Body of left LV	EVD, CSF reservoir, HD-MTX, rituximab, intraventricular liposomal cytarabine	12 months (alive)
Sebastian <i>et al.</i> ^[15]	65	Female	Incidental, no symptom	Left atrium	Resection. Adjuvant therapy not mentioned	NM
Present case	66	Male	Short-term memory disturbance	Right atrium and inferior horn	Resection, HD-MTX, carboplatin and etoposide, 22 × 1.8 Gy WB RT	8 months (alive)

CSF: Cerebrospinal fluid, EVD: External ventricular drainage, HD: High-dose, LV: Lateral ventricle, MTX: Methotrexate, NM: Not mentioned, RT: Radiation therapy, WB: Whole brain

Table 2: Pathologic data on primary choroid plexus lymphoma as described in the literature

Reference	Histology	Markers
Kelley <i>et al.</i> ^[7]	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue	Monotypic B-cell: CD19, CD20, CD45, Bcl-2, Bcl-6 (+); CD5, CD10, cyclin D1 (-). Follicular dendritic cell: CD21 (+). Hyperplastic follicles: CD10, Bcl-6 (+)
Jung <i>et al.</i> ^[6]	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue	B-cell: CD20, CD43, CD79a (+); CD45RO, CD3, CD5, Cyclin D1, CD10, CD23, Bcl-6 (-).
Terasaki <i>et al.</i> ^[17]	T-cell lymphoma	T-cell marker (+)
Cecchi <i>et al.</i> ^[3]	High-grade diffuse large B-cell lymphoma	CD20 (+)
Cheatle <i>et al.</i> ^[4]	Diffuse large B-cell lymphoma	NM
Sebastian <i>et al.</i> ^[15]	Mucosa-associated lymphoid tissue lymphoma	CD20, CD79a, Bcl-2 (+)
Present case	Diffuse large B-cell lymphoma	LCA, CD20, CD30, CD79a, Bcl-2, Bcl-6, MUM-1 (+); CD3, CD10, CD56, EBER (-)

NM: Not mentioned

most common dural lymphomas are MALT lymphomas. These data suggest that the phenotypes of intracranial lymphomas depend on the site of origin. Like PCNSLs, primary choroid plexus lymphomas were shown as enhancing mass lesions on MRI, without hydrocephalus in most cases [Table 3].

Our patient was initially diagnosed as suffering from ependymoma according to the biopsy material obtained at another hospital, possibly because of the intraventricular location and the tightly packed proliferation of tumor cells which may mimic the epithelioid arrangement characteristic of ependymoma. Although the tumor was already disseminated in the subarachnoid space, we performed subtotal removal for mass reduction prior to radiation therapy of the lesion. However, because the final diagnosis was re-interpreted as diffuse large B-cell lymphoma, we subsequently performed systemic chemotherapy as well as whole-brain radiation therapy.

Differentiation of choroid plexus tumors by imaging characteristics alone remains difficult. Calcification is frequently observed in meningiomas and choroid plexus papillomas as well as in native choroid plexus,

and hydrocephalus can be seen as a frequent presenting feature in choroid plexus papilloma.^[15] Hyperintensity on diffusion-weighted imaging may be helpful for differentiation between lymphomas and meningiomas.^[16] Although we did not perform fluorodeoxyglucose/positron emission tomography (FDG-PET) of the brain, these scans may be valuable in establishing a diagnosis as PCNSLs demonstrate high FDG uptake and can be diagnosed by FDG-PET with high sensitivity, whereas ependymomas show low uptake throughout the tumor.^[13,14,19] Furthermore, FDG-PET is helpful in determining whether or not patients have systemic lesions.^[18]

Given the rare scenario of this lymphoma presentation, the best algorithm for the establishment of the correct diagnosis remains a tissue-based diagnosis, until specific biomarkers can be identified.

CONCLUSION

Here, we present a rare case of choroid plexus lymphoma, which should be considered in the differential diagnosis of choroid plexus tumors.

Table 3: Radiologic data on primary choroid plexus lymphoma as described in the literature

Reference	Plain CT	T1WI	T1WIC+	Hydrocephalus	Additional
Kelley <i>et al.</i> ^[7]	NM	NM	NM	-	2-cm oval mass
Jung <i>et al.</i> ^[6]	Hyperdense mass	NM	Homogeneously enhancing mass (3.5 × 1.7 cm)	-	Perilesional edema, dilated subependymal deep veins
Terasaki <i>et al.</i> ^[17]	Homogeneous mass	Homogeneous mass	Rim enhancing mass	-	
Cecchi <i>et al.</i> ^[3]	NM	NM	Homogeneously enhancing mass (2.5 cm at its major axis)	-	Grossly round mass, perilesional edema, no midline shift
Cheatle <i>et al.</i> ^[4]	Hyperdense mass	Homogeneous mass	Homogeneously enhancing mass with strongly enhancing rim	+	
Sebastian <i>et al.</i> ^[15]	NM	NM	Homogeneously enhancing mass	-	
Present case	Hyperdense mass	Heterogeneously hypointense mass	Heterogeneously enhancing mass	-	Perilesional edema

CT: Computed tomography, NM: Not mentioned, T1WI: T1-weighted imaging, T1WIC+: T1-weighted contrast imaging

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

The authors declare no conflicts of interest in association with this study.

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