

Case Report

Hydrocephalus as the sole presentation of primary diffuse large B-cell lymphoma of the brain: Report of a case and review of literature

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Abstract

Background: The most common lymphoid malignancy in adults is diffuse large B-cell lymphoma (DLBCL). The median age of occurrence of DLBCL is between 6th and 7th decade of life, although some other types of aggressive non-Hodgkin's lymphomas (NHL) are present in younger age. Primary central nervous system lymphoma (PCNSL) is an uncommon type of extranodal NHL, which is either more prevalent or is diagnosed more often than before.

Case Description: A 22-year-old man with ventriculoperitoneal shunt (VPS) performed at another center was referred with manifestations of shunt malfunction, unusual behavior, dysphasia, and hallucination. The shunt malfunction was handled appropriately several times and exploration of the enhancing cystic temporal lesion confirmed the diagnosis of DLBCL.

Conclusion: In this communication, we intend to highlight the issue that hydrocephalus refractory to several interventions can be the sole manifestation of PCNSL and needs special clinical considerations.

Key Words: Diffuse large B-cell lymphoma, hydrocephalus, shunt malfunction

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INTRODUCTION

The most common lymphoid malignancy in adults is diffuse large B-cell lymphoma (DLBCL). The incidence of non-Hodgkin's lymphomas (NHL) is estimated to be 15–20 cases/100,000 annually.^[1] The median age of occurrence of DLBCL is between the 6th and 7th decade, and some types of aggressive NHL are present at a lower age.^[1] DLBCLs are highly aggressive, even though they are acceptably curable illnesses. DLBCL is composed of 4% of central nervous system (CNS) tumors, and 1% of all NHL.^[1] In patients with limited disease, 5-year

progression-free survival is expected in 80–85% of the cases, whereas this survival rate drops to 50% in patients

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with advanced disease. The occurrence of DLBCLs in some extranodal regions such as brain^[15] or testis^[19] needs special treatment strategies, and the prognosis of the illness in such cases is very poor. Primary central nervous system lymphoma (PCNSL) is an uncommon type of extranodal NHL. It originates from a cell type that is not present in the CNS in normal condition. Complete regression of tumor by corticosteroids or cranial radiation, and tumor recurrence followed by a fatal course is common.^[1,5,17]

PCNL can involve brain parenchyma, spinal cord, leptomeninges, and eyes.^[2-3,4-6,11-13,16] The clinical presentation of the patients is variable. Cognitive problems, headache, psychiatric symptoms, focal neurological deficits including aphasia, hemiparesis, and ataxia are the major clinical manifestations.^[14]

The purpose of this communication is to highlight a possible strange clinical presentation of PCNL with hydrocephalus resistant to current therapies.

CASE REPORT

A 22-year-old man was referred to this hospital with manifestations of shunt malfunction and history of ventriculoperitoneal shunt (VPS) at another center. The symptoms of increased intracranial pressure appeared the previous year with unusual behavior, speech disturbance, and hallucination before admission in the previous center. His symptoms improved with shunting but recurred after 4 months. Shunt revision was performed and he was discharged with modified Rankin Scale (mRS) 2. The symptoms recurred 2 months later. He had no adenopathy and bone marrow aspiration was negative. Full paraclinical evaluations and all laboratory tests were highly suspicious of histiocytosis even though CD1 test was negative. Cerebrospinal fluid (CSF) analysis was normal (WBC: 4, RBC: 668, Glc: 5, Pro: 54), and CSF culture and cytology was negative. He was treated with corticosteroids and another shunt was placed in the contralateral ventricle under antibiotic coverage. The reports of magnetic resonance imaging (MRI) provided from the previous hospital denoted a communicating hydrocephalus and no space occupying lesion all along the brain and spinal cord. After 3 months, he was admitted in our hospital. The mentioned information was all the data that were delivered from the previous center to our hospital.

In physical examination, he was conscious and awake but hardly cooperative and sometimes even unresponsive to painful stimuli. Brain computed tomography (CT) scan showed cystic dilatation of left temporal horn surrounded by perilesional edema [Figure 1]. Brain MRI showed hydrocephalus with periventricular changes without focal enhancement. There was cystic expansion

of the left temporal horn with fine wall enhancement and perilesional edema [Figure 2]. Whole spine contrast-enhanced MRI was normal.

He underwent craniotomy to explore the presumably dilated temporal horn. There was no abnormality in the extradural and subdural region in the exposed area of the temporal region. The arachnoid was shiny and no granulomatous change or focal thickening of the arachnoid was visible. Trans sulcal approach in between the middle and low temporal gyri preformed to enter the cystic/expanded lesion. Entering the dilated/isolated temporal horn, nests of shiny debris infiltrating the ependymal layer of the dilated horn could be stripped off and removed as much as possible [Figure 3]. The choroid plexus was coagulated enough, and washing the collapsed cystic component could maintain slow CSF flow. Pathology report confirmed the diagnosis of DLBCL [Figure 4]. The patient was referred to neuro-oncologist with discharge mRS = 2. He improved significantly after two doses of chemotherapy using Methotrexate, but deteriorated and died 2 months later with tumor seeding in the 4th ventricle, brain stem, and cervical spine.

DISCUSSION

PCNSL has a highly fulminant course and misdiagnosis or late diagnosis may lead to early mortality.^[14] The most common type of PCNSL is DLBCL.^[10] This type of PCNSL is composed of immunoblasts or centroblasts resulting in lymphoid clustering around small cerebral vessels.^[9] Most believe that CNS neither has a lymphatic circulation nor any endogenous accumulation of lymphoid tissue.^[19] Accordingly, the suggested source for development of intraventricular lymphoma is activation of lymphocytes located in the stroma of choroid plexus seeding to the wall of the ventricles as in this case.^[19] DLBCL includes a group of lymphoid malignancies characterized by large cells with vesicular nuclei, prominent nucleoli, basophilic cytoplasm, and a high proliferation index as seen in the pathology report of our case [Figure 3]. Most cases of DLBCL have unknown cause, but there are some differences between *de novo* (primary) cases and those arising from progression/transformation (secondary) of a less aggressive lymphoma, e.g. chronic lymphocytic leukemia, small lymphocytic lymphoma, follicular lymphoma, marginal zone lymphoma, or nodular lymphocyte predominant Hodgkin lymphoma.^[7]

The predisposing and etiological factors for DLBCL include agents producing molecular aberrations,^[18] several chemical substances, such as pesticides, fertilizers,^[4] and alkylating agents used in the treatment of solid tumors and hematological malignancies. The prevalence of lymphoma (secondary) rises following

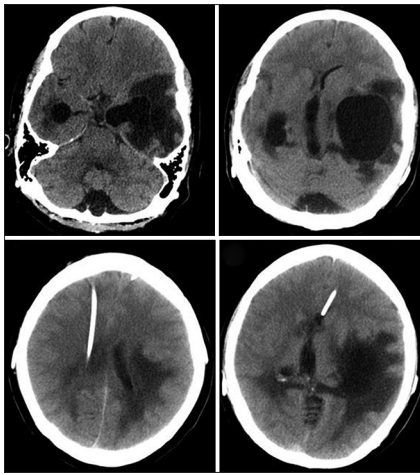


Figure 1: No contrast CT scan showing two ventricular catheters, each in one of the frontal horns collapsing both ventricles. The left temporal horn is isolated and dilated remarkably with perilesional edema

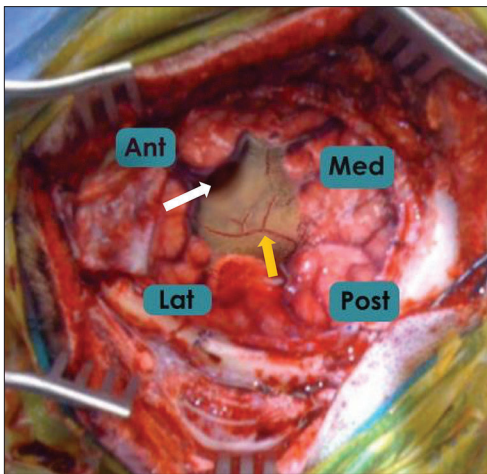


Figure 3: Surgical view of left temporal lobe with the exposed temporal horn (yellow arrow) containing the cystic lesion at superior border (white arrow) and velvety debris along the ependymal layer

the combination between alkylating agents and ionizing radiation.^[17] The patients suffering from immune-compromised situation after organ transplants are of high risk for diffuse aggressive lymphoma of the brain.^[9,17] No one of these predisposing elements was present in this case.

PCNSL is characterized by nonspecific neurologic symptoms which is in contrary to the clinical presentation of the systemic “B” type of lymphoma usually presenting with fever, weight loss, and night sweats. The incidence of focal neurological deficits and global neurological deterioration due to PCNSL are the same; that is why the authors have not been able to define typical clinical manifestations for it.^[16] Neuropsychiatric symptoms have been reported in 43% of cases of PCNSL.^[10] However, some have reported

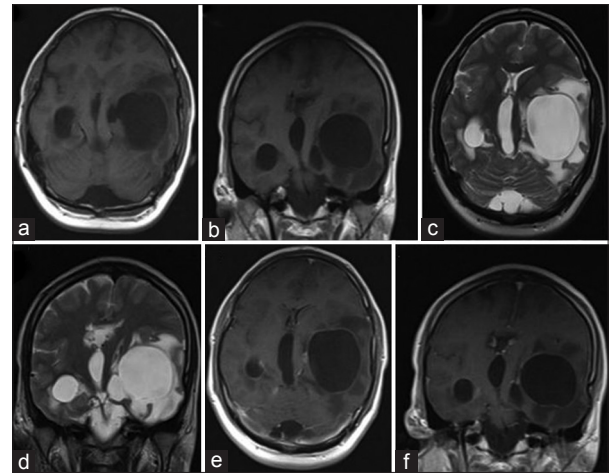


Figure 2: Hypointense cystic lesion in axial and coronal T1 weighted MRI (a and b); hyperintense cystic lesion with massive edema in axial and coronal T2 weighted MRI (c and d); rim enhancing lesion in axial and coronal contrast-T1 weighted MRI (e and f). (Sagittal views not included due to bad quality)

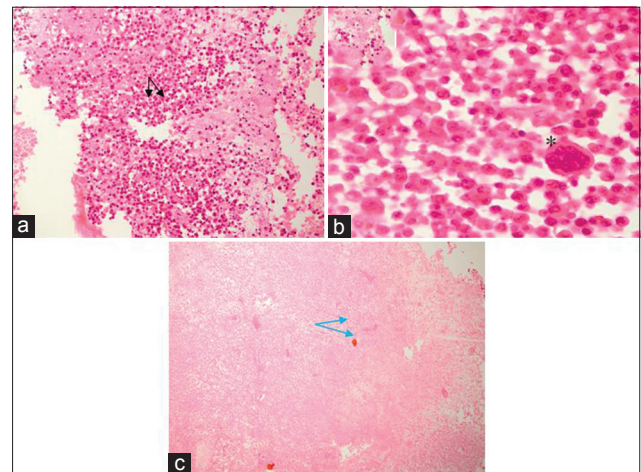


Figure 4: (a and b) Large lymphoid cells with variation in size and shape (black arrows), multilobulated nuclei, large atypical lymphoid cell with prominent multilobulated nuclei (asterisk), (c) necrotic tissue with ghost cells (blue arrows). CD10 and CD20 was positive in immunohistochemistry

psychiatric symptoms such as mania, depression, and intermittent vomiting as the prominent manifestations of PCNSL.^[8,15] These were the main reasons for referring our patient to the hospital and was added to the manifestations of hydrocephalus.

The neuropsychological features are highly associated with involvement of the periventricular white matter or the corpus callosum by the tumor.^[14,16] This correlation was present in our patient and the images. A superficial location appearing in MRI, location in the basal ganglia, corpus callosum or fornix, infiltration of the periventricular ependyma, contrast enhancement, and absence of necrosis are some of the imaging characteristic features of PCNSL.^[6-8] We highlighted such findings in this case with infiltration of the ependyma of the ventricles leading to hydrocephalus

as the unique imaging character of lymphomatous origin of hydrocephalus. Patients with PCNSL recurrence have poor prognosis, and median survival time is 2–5 months.^[4]

The present case demonstrates that PCNSL can involve ventricular system and lead to hydrocephalus. A refractory or recurrent case of hydrocephalus after shunting needs to be evaluated for an underlying disease such as DLBCL.

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Conflicts of interest

There are no conflicts of interest.

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