Endoscopic biopsy of a B-cell lymphoma involving the entire ventricular system: A case report

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Abstract. A 62-year-old male suffering from vomiting and mild preceding nausea for 15 days was examined in the present case report. Magnetic resonance imaging revealed a homogeneously enhancing cluster-like lesion involving the lateral, third and fourth ventricles. An endoscopic biopsy was performed, and histopathological examination led to the diagnosis of a high-grade diffuse large B-cell lymphoma. To the best of our knowledge, the present study reports the first case of a primary lymphoma involving the entire ventricular system. Therefore, primary lymphomas should be considered in the list of ventricular tumors. An endoscopic biopsy requires minimal invasion to obtain an adequate tissue sample, and frequently leads to the correct diagnosis and subsequent treatment protocols.

Introduction

Primary central nervous system lymphomas (PCNSLs) are a class of non-Hodgkin's lymphomas, which are primarily of diffuse large B-cell origin (90-95%), with the remaining being T-cell lymphomas (5-10%). The incidence of PCNSL has markedly increased over the last three decades (1). Primary ventricular lymphomas are extremely rare and may present as a solid mass or as diffuse ventriculitis. The clinical presentations of PCNSL are nonspecific and include headaches, vomiting, focal neurological deficits or global neurological deterioration (2). Since lymphomas are tumors with diffuse infiltration, surgical resection is not an efficient treatment; however, the disease may be controlled by chemotherapy and radiotherapy. Therefore, only a limited biopsy is required to confirm the diagnosis of a lymphoma, thereby preventing serious after-effects (1). Endoscopic neurosurgery for the biopsy of intraventricular brain tumors has been established in recent years. The efficacy of endoscopic tumor management is based on the principle that the intraventricular cerebrospinal fluid (CSF) serves as an excellent natural medium for imaging and light transmission. To date, only a few cases of intraventricular PCNSL have been reported in the literature (1-9). To the best of our knowledge, the present study reports the first case of PCNSL involving the entire ventricular system, as demonstrated by an endoscopic biopsy.

Case report

Written informed patient consent was obtained from the patient's family. In August 2012, a 62-year-old male was admitted to the Affiliated Bayi Brain Hospital of the Military General Hospital of Beijing PLA (Beijing, China), suffering from spontaneous, unrelenting vomiting and mild preceding nausea for 15 days, without the presence of a headache. The patient did not have previous medical history, general and neurological examinations were found to be normal and initial blood tests were unremarkable. A magnetic resonance imaging scan was performed, which revealed a homogeneously enhancing cluster-like lesion involving the lateral, third and fourth ventricles (Fig. 1A-C). To exclude the possibility of metastases, a whole-body positron emission tomography-computed tomography examination was performed, which produced negative results. No ventricular obstruction, other mass lesions or gross edemas were observed; therefore, the administration of steroids was withheld. An endoscopic biopsy was performed with neuronavigation under general anesthesia. The endoscopic biopsy revealed a white-red, solid mass with a rough surface on the occipital horn of the lateral, third and fourth ventricles, covering the foramen of Monro. Histopathological examination identified a highly cellular tumor comprising large cells with small decondensed nuclei and basophilic cytoplasm, scanty stroma and brisk mitotic activity, with the B-cell markers, CD20 and CD79a. In addition, >80% of the cells exhibited

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Table I. Summary of primary fourth ventricular schwannoma

Author/year	Age (yr)/Gender	Location	Clinical Signs	Treatment
Werneck/1977 (5)	17/F	4th V	Meningitis	Not available
Bogdahn/1986 (9)	51/M	Lateral V	Seizures	Biopsy, chemotherapy, radiotherapy
	71/F	Lateral V	Confusion, motor and speech disturbances, left hemiparesis	Biopsy, chemotherapy, radiotherapy
Haegelen/2001 (3)	33/F	4th V	Headaches, vertigo, static cerebellar syndrome	Chemotherapy, stem cell transplantation, radiotherapy
Kelley/2005 (6)	53/M	3rd V	Headache, seizures	Gross total resection
Jung/2006 (7)	63/M	3rd V	Confusion, left leg monoparesis	Radical resection
Terasaki/2006 (8)	56/M	Lateral V	Headache	Chemotherapy, radiotherapy
Cecchi/2008 (2)	71/F	3rd V	Confusion, motor speech disturbances, left hemiparesis	Chemotherapy, radiotherapy
Hill/2009 (4)	69/M	4th V	Vomiting, mild preceding nausea	Biopsy, chemotherapy
Brar/2012 (1)	65/F	Lateral V, 4th V	Normal	Biopsy, chemotherapy
Present/2014	62/M	Lateral V, 3rd V, 4th V	Vomiting, with mild preceding nausea, no headache	Biopsy, chemotherapy, radiotherapy

M, male; F, female; V, ventricle.

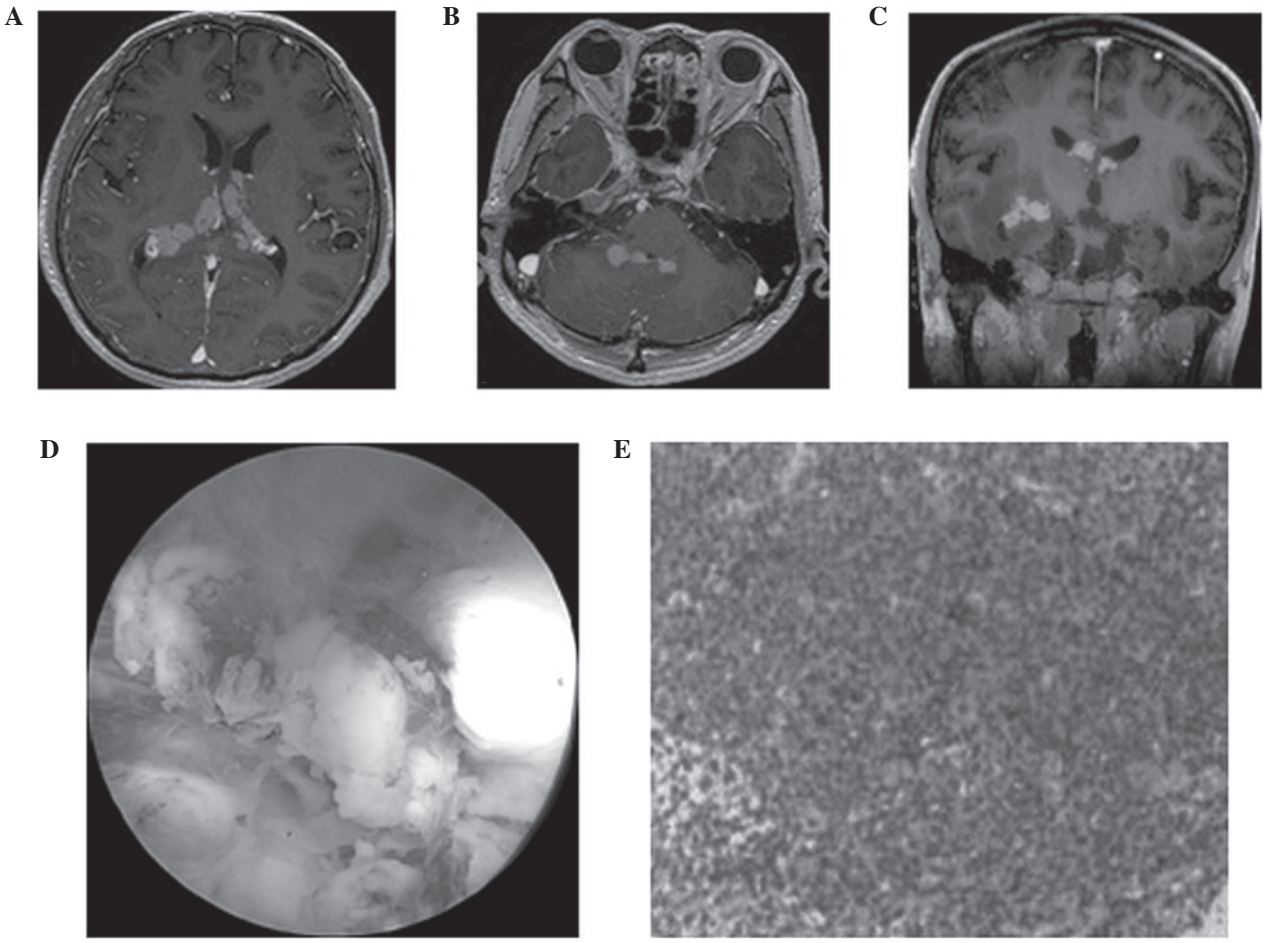

Figure 1. (A and B) Axial T1 and (C) coronal-weighted magnetic resonance imaging scans with gadolinium contrast agent, demonstrating the homogeneous enhancing activity of the tumor, which is located in the lateral, third and fourth ventricles. (D) An endoscopic biopsy revealed a white-red, solid mass with a rough surface, found on the occipital horn of the ventricles. (E) Histopathological examination confirmed the diagnosis of a high-grade diffuse large B-cell lymphoma.

nuclear positivity for the proliferation marker, Ki67, while the cells were found to be negative for Epstein-Barr virus latent membrane protein 1. Thus, the patient was diagnosed with a high-grade diffuse large B-cell lymphoma.

The patient commenced chemotherapy, which consisted of cyclophosphamide (750 mg/m^2), doxorubicin (50 mg/m^2) and vincristine (1.4 mg/m^2), with a maximum single dose of 2 mg. The treatment was adminstered intravenously on day 1, and etoposide was administered at a dose of 100 mg/m^2 intravenously on days 1-3 alongside 100 mg oral prednisone on days 1-5. This cycle was repeated every three weeks for six courses. The patient then underwent entire brain radiotherapy at a dose of 30 Gy. The patient showed good recovery without any neurological deficits. Following the treatment, the levels of plasmatic markers (lactate dehydrogenase and β_2 microglobulin) were determined by protein electrophoresis, slit-lamp evaluation, a bone marrow biopsy and computed tomography scans of the chest and abdomen appeared to be normal. In addition, serological tests for human immunodeficiency virus, cytomegalovirus and Epstein-Barr virus were negative. At the seven-month follow-up examination, the patient was healthy, with no evidence of recurrence.

Discussion

Only a few cases of intraventricular PCNSL have been reported in the literature, of which three cases were in the fourth ventricle (2-5), four cases were in the third ventricle (2,6,7) and three cases were in the lateral ventricles (8,9), while only one case was identified with simultaneous involvement of the lateral and fourth ventricles (1). To the best of our knowledge, the present study reports the first case of a lesion involving the entire ventricular system. Table I shows the characteristics of the previously reported cases, along with the findings of the present study.

Although the occurrence of PCNSL is extremely rare, including the possibility of such tumor in the differential diagnosis of intraventricular neoplasms is required, particularly in cases where the lesion is located in one or multiple ventricles and exhibits a cluster-like appearance (10). Surgical resection is not an efficient treatment method for PCNSL; however, the disease may be controlled by chemotherapy and radiotherapy. Therefore, only a limited biopsy is required to confirm the diagnosis of a lymphoma, thereby preventing serious after-effects.

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