Successful endoscopic biopsy for Primary central nervous system lymphoma of the corpus callosum in the splenium with bilateral visuomotor ataxia

Yusuke Morinaga1,*, Kouhei Nii1, Kimiya Sakamoto1, Ritsurou Inoue1, Takafumi Mitsutake1, Hayatsura Hanada1, Seiji Haraoka2

1 Department of Neurosurgery, Fukuoka University Chikushi Hospital, Chikushino, Fukuoka, Japan; 2 Department of Pathology, Fukuoka University Chikushi Hospital, Chikushino, Fukuoka, Japan.

SUMMARY Primary central nervous system lymphoma (PCNSL) is a rare malignant tumor of the central nervous system. It is associated with poor prognosis. Early diagnosis and subsequent planning of adequate treatment strategy are relevant to improve survival and reduce neurological deficit. Specifically, there are no reports of successful endoscopic biopsy for PCNSL of the corpus callosum in the splenium with bilateral visuomotor ataxia. An 74-year-old woman presented to our hospital with anorexia, depression and ataxia beginning six months earlier. Head computed tomography and magnetic resonance imaging showed malignant tumor suspected in the corpus callosum. Endoscopic biopsy was performed via the low parietal approach, suspecting GBM or PCNSL. She had no new postoperative neurological deficits and was pathologically diagnosed with diffuse large B-cell lymphoma (DLBCL). She is currently undergoing radiation chemotherapy with a modified Rankin Scale score of 2. Regarding preoperative symptoms, ataxia was considered to be bilateral visuomotor ataxia caused by damage to the corpus callosum in the splenium, and anorexia and depression were considered symptoms of the surrounding limbic system. Delay in the diagnosis of PCNSL can lead to a poor prognosis. Visuomotor ataxia should also consider the potential for the corpus callosum in the splenium lesion, including PCNSL, and appropriate imaging and pathological diagnosis with endoscopic biopsy can contribute to a good clinical outcome.

Keywords PCNSL, DLBCL, visuomotor ataxia, corpus callosum in the splenium, endoscopic biopsy

1. Introduction

Primary central nervous system lymphoma (PCNSL) is a rare malignant tumor of the central nervous system. It is associated with poor prognosis and accounts for 0.7-0.9% of all lymphomas and only 0.3-1.5% of intracranial tumors. Typically, these lesions are in the cerebral white matter near the corpus callosum, the central gray matter, the basal ganglia-thalamus-hypothalamic region, the posterior fossa and the periventricular region (1). PCNSL and glioblastoma multiforme (GBM) are malignant cerebral neoplasms associated with poor prognosis. Early diagnosis and subsequent planning of adequate treatment strategy are relevant to improve survival and reduce neurological deficit (2). Specifically, there are no reports of successful endoscopic biopsy for PCNSL of the corpus callosum with bilateral visuomotor ataxia. Here, we describe a rare case where PCNSL of the corpus callosum with bilateral visuomotor ataxia was diagnosed early and successfully by endoscopic biopsy.

2. Case Report

A 74-year-old woman presented to our hospital with anorexia, depression, and ataxia beginning six months earlier. She had history of dyslipidemia and had taken atorvastatin. Neurological assessments revealed a Japan Coma Scale grade I-1, ataxia, and no limb paralysis and cerebellar symptoms. Upon hospitalization, his heart rate was 62 bpm and his blood pressure was 120/72 mmHg. Blood sampling revealed normal IL-2 receptor (417 U/mL) and no other abnormalities. Plain head computed tomography (Figure 1A) and magnetic resonance imaging (MRI) (Figure 1B, 1C) showed malignant tumor suspected on the corpus callosum in the splenium with perifocal edema. Neuronavigation-guided endoscopic biopsy was performed using a 10 mm cylinder via the low parietal approach, suspecting
GBM or PCNSL (Figure 2A, 2B). Postoperative MRI revealed good enucleation and no bleeding (Figure 3). She had no new postoperative neurological deficits and was pathologically diagnosed with diffuse large B-cell lymphoma (DLBCL). She is currently undergoing radiation chemotherapy with a modified Rankin Scale score of 2. Regarding preoperative symptoms, ataxia was considered to be visuomotor ataxia caused by damage to the corpus callosum in the splenium, and anorexia and depression were considered symptoms of the surrounding limbic system.

Informed consent was obtained from the patient for publication of this case report and the accompanying images, and the study design was approved by the appropriate ethics review board.
3. Discussion

To the best of our knowledge, reports of successful early diagnosis by endoscopic biopsy for PCNS of the corpus callosum with optic ataxia, such as the present one, are rare (1,2).

Rondot P, et al. (3) reports that visuomotor ataxia (3,4) is a disorder of movement performed under visual control. It can occur in the absence of disturbance of ocular fixation and in the absence of spatial agnosia. This disorder may extend over the whole visual field or it may be localized to one visual half-field, right or left. It may involve both hands or one hand only, so that visuomotor ataxia may be divided into: (1) Unilateral visuomotor ataxia, localized to a single field. In this case it may affect both hands or a single hand. It is direct when the hand is ataxic in the ipsilateral visual field and it is crossed when the hand is ataxic in the contralateral visual field; (2) Bilateral visuomotor ataxia, involving the whole visual field. Each hand may be ataxic only in the contralateral visual field, that is, bilateral crossed visuomotor ataxia; or in the ipsilateral field when it is called bilateral direct visuomotor ataxia. The observed clinical variations which are described here imply the existence of both direct and crossed visuomotor connections, the latter probably crossing the corpus callosum in the splenium (3,4). Callosal disconnection syndrome (5) refers to conduction aphasia, visuomotor ataxia, apraxia of the left hand, agraphia of the left hand, alien hand syndrome (6). In the corpus callosum in the splenium, visual and auditory information integration and memory functions are impaired (3,4). These symptoms may not show any noticeable disability in daily life and are often not recognized without a special callosal function test. In our case, regarding preoperative symptoms, ataxia was considered to be bilateral visuomotor ataxia caused by damage to the corpus callosum in the splenium, and anorexia and depression were considered symptoms of the surrounding limbic system (7).

Marion R, et al. reports that fluorescence-guided endoscopic visualization identified 5-aminolevulinic acid (5-ALA)-positive tissue not sufficiently exposed by conventional microscopic visualization (8). Although 5-ALA was not used in our case, safe endoscopic biopsy could be performed under the neuronavigation, and it was considered to be a minimally invasive treatment.

In conclusion, delay in the diagnosis of PCNSL can lead to a poor prognosis. Bilateral visuomotor ataxia should also consider the potential for the corpus callosum in the splenium lesion, including PCNSL, and appropriate imaging and pathological diagnosis with endoscopic biopsy can contribute to a good clinical outcome.

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*Address correspondence to:*

Yusuke Morinaga, Department of Neurosurgery and Neuroscience, Fukuoka University Chikushi Hospital, 1-1-1 Zokumyoin, Chikushino-shi, Fukuoka 818-8502, Japan.
E-mail: yu_the_morio@yahoo.co.jp

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