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CASE REPORT

# A rare case of Primary Central Nervous System Lymphoma initially diagnosed as demyelinating encephalopathy

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**Abstract**

We report a case of histopathologically-confirmed primary central nervous system lymphoma who was initially diagnosed as demyelinating encephalopathy. A 58-year-old woman was admitted with confusion and left hemiparesis. Head MR showed abnormal flaky hypointense T1 and hyperintense T2 signals at right thalamus, splenium of corpus callosum, bilateral cerebral peduncle, pons, medulla oblongata, basal ganglia and right corona radiata. Her mental status improved a little and she was discharged from hospital after neuroprotective treatment. 10 days after her discharge, her confusion appeared again with hallucination and unsteady walking. Pathological examination revealed non-Hodgkin's lymphoma (WHO classification: DLBCL). The patient continued to deteriorate after the surgery and died 10 days later.

## INTRODUCTION

Primary central nervous system lymphoma (PCNSL) is an aggressive extranodal non-Hodgkin's lymphoma (NHL) involving the brain, spinal cord, meninges and eyes with few metastasis outside the CNS. The reported incidence is 3–4 cases per 100,000 and accounts for 4% of CNS malignancies and 1–2% of NHLs (Ferreri *et al.* 2003). The incidence in immunocompetent individuals has increased by more than 10-fold in the past three decades with an annual current rate of 0.3 per 100,000 person-years (Corn *et al.* 1997). Approximately 90% of PCNSL are diffuse large B-cell lymphoma (DLBCL). Untreated patients survive only for a few months. With the low incidence rate and lack of large-scale clinical randomized controlled research data, the optimal therapy for PCNSL remains to be defined. The differential diagnosis of PCNSL is extensive and it can be challenging to diagnose initially in the absence of pathognomonic clinical features. Herein we report a case of histopathologically-confirmed PCNSL who was initially diagnosed as demyelinating encephalopathy and discuss the current literature.

## CASE REPORT

A 58-year-old woman was admitted with confusion and left hemiparesis. Head MR showed abnormal flaky hypointense T1 and hyperintense T2 signals at right thalamus, splenium of corpus callosum,

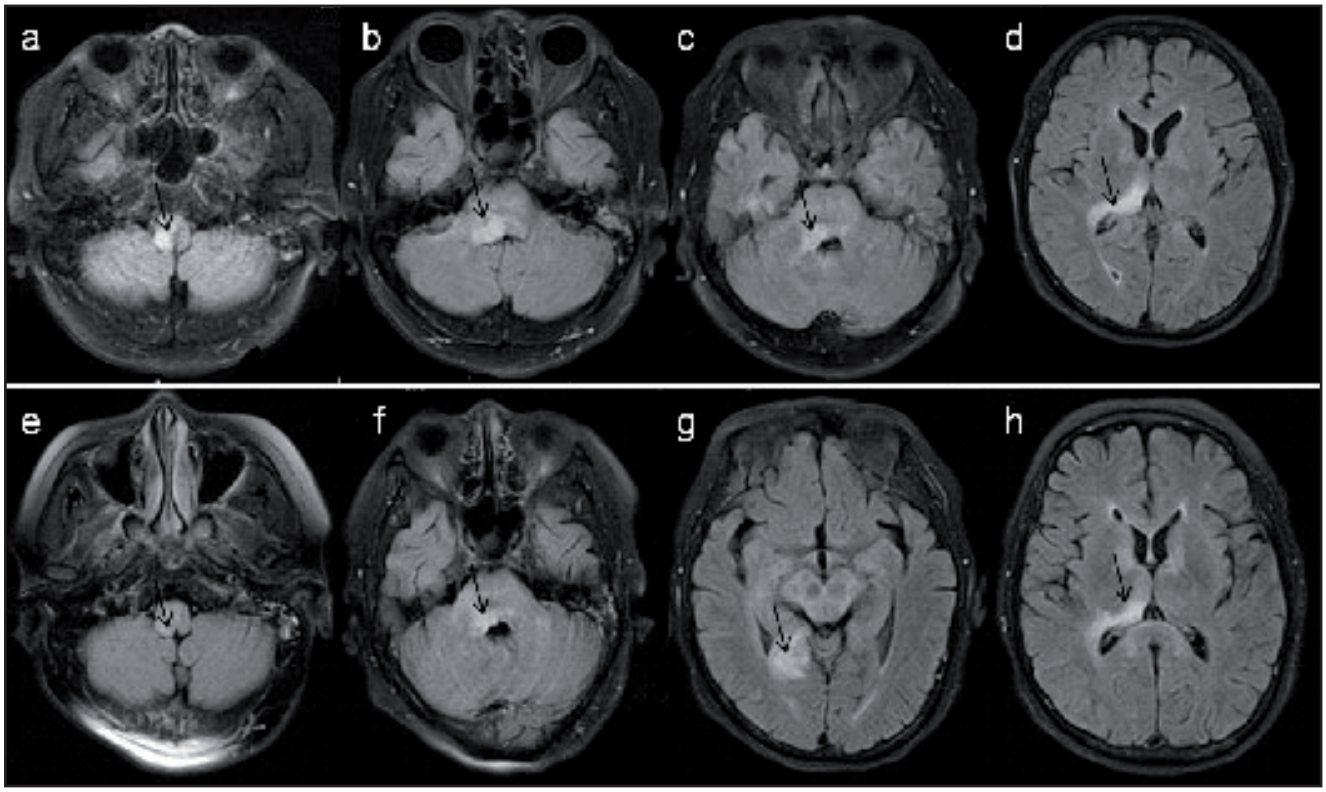


**Fig. 1.** Head MR FLAIR sequence showed abnormal flaky hypointense T1 and hyperintense T2 signals at right thalamus, splenium of corpus callosum, bilateral cerebral peduncle, pons, medulla oblongata, basal ganglia and right corona radiata (a-d arrow indicated); Head MR FLAIR sequence (one months later) showed abnormal flaky hypointense T1 and hyperintense T2 signals were more a lot more than that before (e-h arrow indicated).

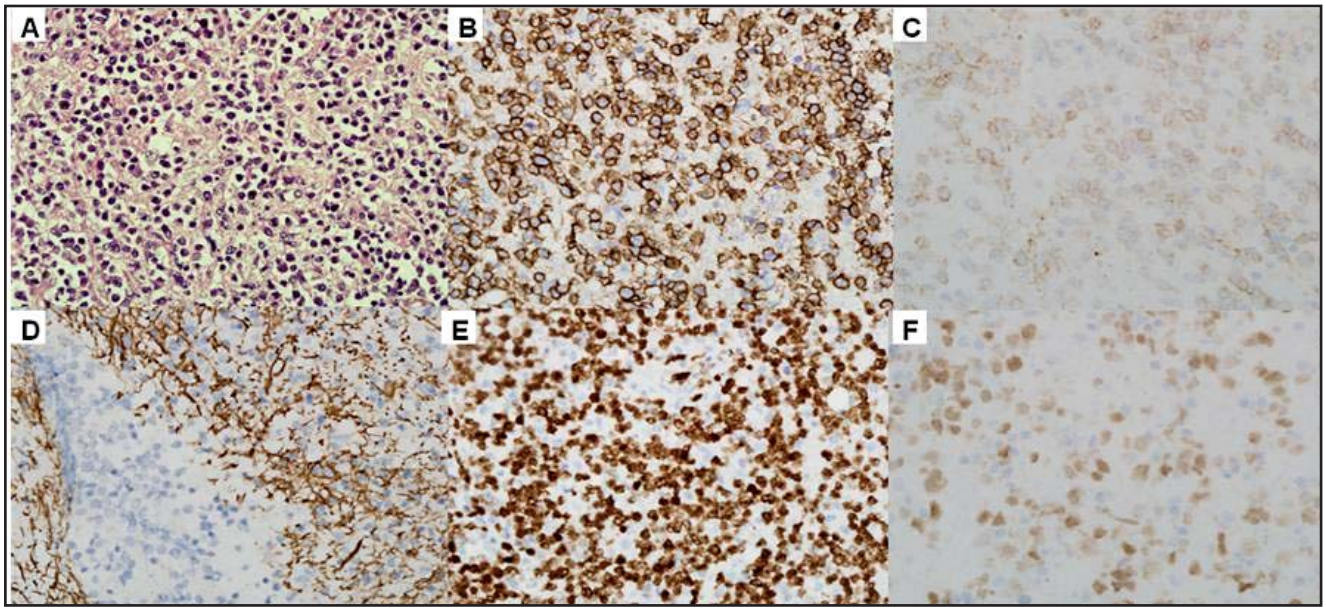


**Fig. 2.** Pathological examination revealed NHL (WHO classification: DLBCL) (A: HE staining revealed abnormal lymphocyte proliferation; B: CD20 positive mostly oringinated from B cell; C: CD79a positive; D: GFAP negative; E: Ki-67 positive; F :MUM1 positive).

bilateral cerebral peduncle, pons, medulla oblongata, basal ganglia and right corona radiate (Figure 1). She was diagnosed as "demyelinating encephalopathy" whereas methylprednisolone was not administered on account of diabetes. Her mental status improved a little and she was discharged from hospital after neuroprotective treatment. 10 days after her discharge, her confusion appeared again with hallucination and unsteady walking. Head MR showed multiple abnormal signals and these signals were more than that MR aforemen-

tioned (Figure 1). Enhancement scan showed right occipital lobe was abnormally enhanced. Stereotactic biopsy was administered and the pathological examination revealed NHL (WHO classification: DLBCL) (Figure 2). The patient continued to deteriorate after the surgery and died 10 days later.

## DISCUSSION

PCNSL was first described by Bailey in 1929. It comprises highly malignant tumors which exhibit a preference for basal ganglions, corpus callosum, thalamus, peri-ventricular white matter and subependymary region. It is considered as an AIDS-marking neoplasia. These lymphomae course without fever, have a high mortality rate with rapid evolution toward clinical and neurological deterioration. Image tests reveal a central area or necrosis and ring contrast capture at the level of the injury, which gives rise to a differential diagnostic with demyelinating encephalopathy (Haldorsen *et al.* 2007). Our patient had no history of AIDS immunodeficiency or use of immunosuppressive drugs and therefore PCNSL was not initially considered.

Demyelinating encephalopathy is a common cause of space-occupying injuries. The treatment of choice is empirically associated with high dexamethasone dosages to reduce the brain edema with the ensuing clinical and radiological improvement in 1–3 weeks; if clinical improvement is absent, it is recommended to carry out a stereotactic biopsy of the space-occupying lesion (Sparano *et al.* 2001).

The presence of a single large sized lesion (>3 cm) and localized at the level of the periventricular white matter suggests the diagnostic of PCNSL (Corti *et al.* 2001). In our case, pathological examination finally confirmed the diagnosis.

PCNSL, a rare form of extranodal NHLs, is typically a DLBCL that is confined to the nervous system. The diagnosis of PCNSL is supported by CT and MRI studies, but is ultimately confirmed on the basis of stereotactic biopsy in most patients. In general, the diagnostic of these neoplasiae is usually carried out by the internist physician as in most of cases they debut with neurological focal signs and only 10% is admitted to neurosurgery department as space-occupying lesions. Therefore, we emphasize the crucial role the neurologist can play in the early diagnostic of CNS neoplasiae.

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