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Case Report

Primary central nervous system lymphoma (PCNSL) mimicking a falx meningioma with increasingly massive intracerebral hemorrhage ☆

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ABSTRACT

When using imaging to diagnose brain tumors, it is important to determine whether the tumor is intra- or extra-parenchymal. An 80-year-old man was found on magnetic resonance imaging to have an enhanced mass that appeared to be in the falx and a massive hematoma in the left frontal lobe; the provisional diagnosis was falx meningioma. However, the tumor and hematoma were found intraoperatively to be completely intraparenchymal. Additionally, the falx was intact and not adherent to brain tissue. Malignant lymphoma was diagnosed histologically on the basis of abnormal proliferation of atypical CD20-positive lymphocytes. Cerebral hemorrhage is an extremely rare presentation of primary central nervous system lymphoma. To the best of our knowledge, only 7 cases have been reported. All the reported cases had enhancement in the hematoma; however, in our case, there was definite enhancement outside the hematoma, making the correct diagnosis of lymphoma difficult.

Introduction

It is not difficult to distinguish between meningioma and primary central nervous system lymphoma (PCNSL) on imaging studies. Generally, the former is an extra-parenchymal and the latter is an intra-parenchymal tumor, showing a clear difference on magnetic resonance imaging (MRI). However, malignant lymphoma with a large subcortical hematoma can make a definite diagnosis difficult.

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Case presentation

A healthy 80-year-old man was admitted to our hospital because of sudden onset of cognitive dysfunction and mild right hemiparesis. His Glasgow coma scale (GCS) score was 15 (E4V5M6) and the patient could feed himself and walk with help. Computed tomography without contrast medium demonstrated an intracerebral hemorrhage (ICH) in the left frontal lobe (Fig. 1A Day 0) with extensive brain edema, suggesting a malignant tumor. No antiplatelets or anticoagulants were prescribed to the patient, and his platelet count was 23.9 $\times$ 10^4 /μL (normal range: 15.8-34.8), activated partial thromboplastin time (APTT) 30.6 seconds (24.0-39.0), prothrombin (PT) time 13.1 seconds (8-13), and prothrombin international normalized ratio (PT-INR) 1.18. Malignant lymphoma was included in the initial differential diagnoses because his interleukin-2 receptor (IL-2R) concentration was 1047 U/mL (normal range: 122-496 U/mL) on admission. However, the hematoma gradually enlarged (Fig. 1A; Day 5) and MRI showed gadolinium–dimeglumine enhancement (Fig. 1B; white arrows) and the dural tail sign (Fig. 1B; white dotted arrows) in the falx, suggesting meningioma. Although very rare, findings on fluid-attenuated inversion recovery (FLAIR) and T2W images led to the diagnosis of large ICH caused by falx meningioma. On the 10th hospital day, the patient's consciousness level suddenly deteriorated to GCS 6 (E2M2M2). Computed tomography showed further enlargement of the ICH (Fig. 1A; Day 10). Emergency surgery was performed to remove the hematoma and enable histological examination.

Intraoperative findings

The brain was tight and bulged out as soon as the dura mater was opened. There was no hematoma outside the brain. The ICH was aspirated through a small corticotomy in the left

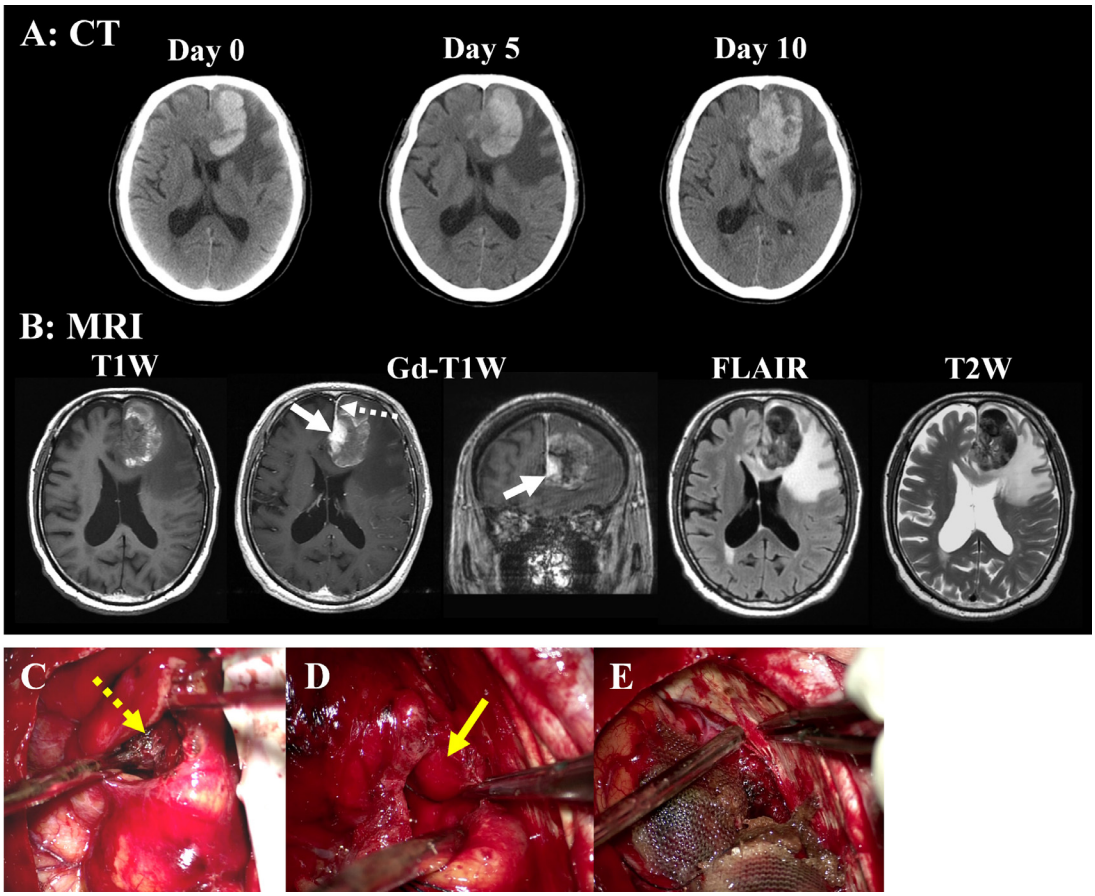

Fig. 1 – (A) Computed tomography images on admission (Day 0) demonstrating a high-density mass suggesting hemorrhage in the left frontal lobe with large low-density area. On Day 5, the high-density area has spread to the right frontal lobe. The hematoma has enlarged further by Day 10, severely compressing the lateral ventricle. All CT images were performed without contrast media. (B) Magnetic resonance imaging on Day 5 displaying high-signal lesions on T1W, suggesting acute hemorrhage. Gd-T1W demonstrates definite enhancement in or adjacent to the falx (white arrows) with a dural tail sign (dotted arrow), suggesting that the enhanced mass is attached to the falx. FLAIR and T2W demonstrate a massive intraparenchymal low-signal mass in the left frontal lobe, implying a large hematoma caused by the tumor at the falx. (C-E) Intraoperatively, no hemorrhage was found outside the brain. The hematoma was identified in the frontal lobe by corticotomy (yellow dotted arrow). An elastic mass that bled easily was identified medical to the left frontal lobe (yellow arrow). The falx was found to be intact and not adherent to the tumor mass.

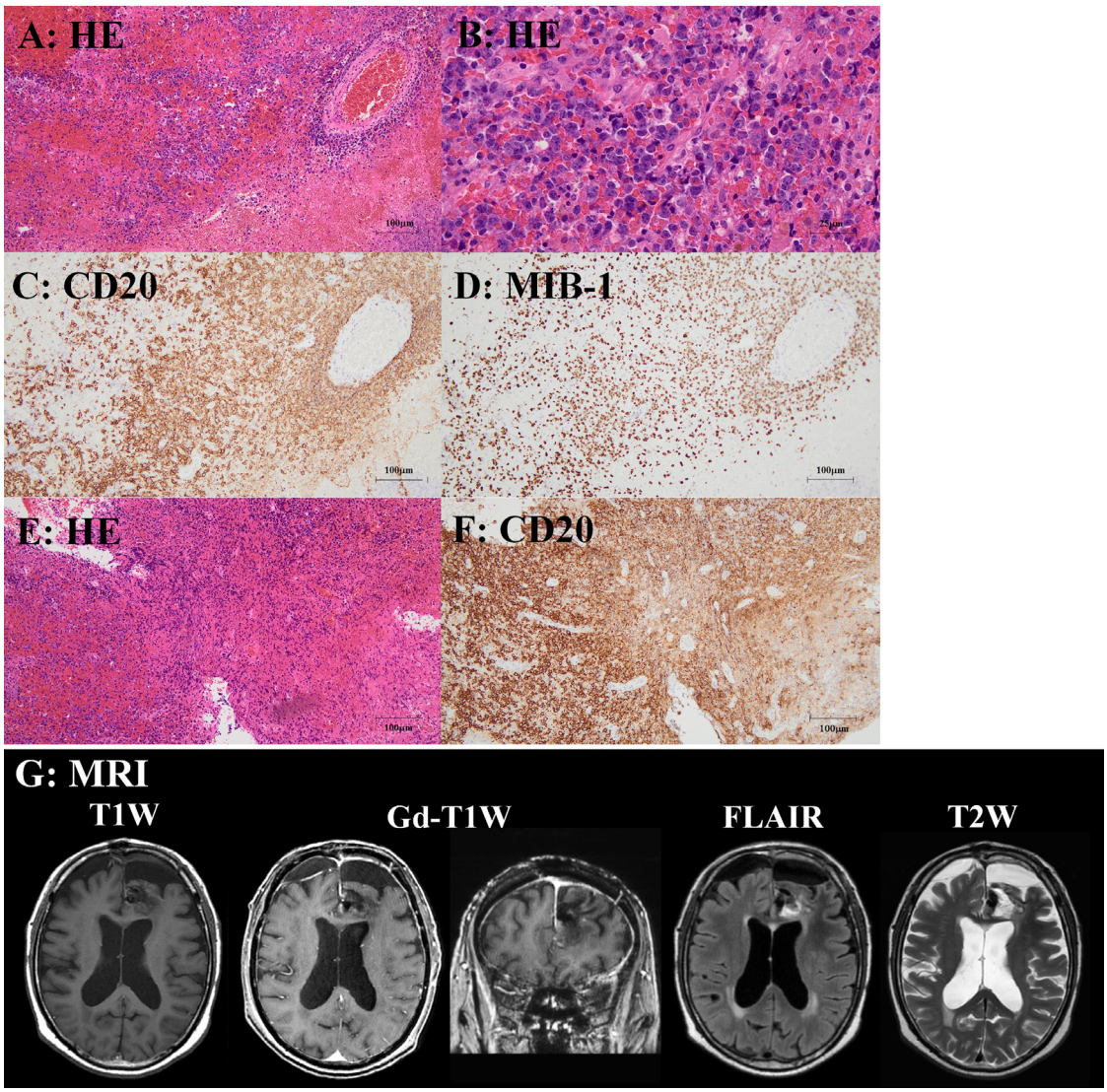

Fig. 2 – (A, B) Histological examination showed clusters of atypical lymphocytes. These atypical lymphocytes are CD20 positive (C) and the MIB-1 index is 80%-90% (D). (E, F) CD20-positive, atypical lymphocytes are also present in the hematoma-affected brain tissue. (G) Magnetic resonance imaging 3 weeks after surgery demonstrates that the tumor mass has been removed, there is no mass lesion in the falx, and the brain edema has resolved according to FLAIR. A high intensity is in the left frontal lobe on FLAIR and T2W, which is not enhanced, is the reaction after tumor resection.

frontal lobe (Fig. 1C; yellow dotted arrow). After achieving decompression, an elastic tumor mass was identified medial to the hematoma (Fig. 1D; yellow arrow). The tumor bled profusely on resection and no clear border was found between it and normal brain tissue. The tumor was resected as completely as possible, after which the dura mater was found to be normal (Fig. 1E).

Histological findings

A diffuse-large B-cell type malignant lymphoma was diagnosed on the basis of clusters of atypical lymphocytes (Figs. 2A and B) with CD20 positivity (Fig. 2C) and a high MIB-1 index of 80%-90% (Fig. 2D). Atypical CD20 positive lymphocytes were also identified in the hematoma-affected brain tissue (Figs. 2E and F).

By 3 weeks after the surgery, the patient's consciousness had improved to GCS 15 (E4V5M6) and he was able to walk with help. A follow-up MRI demonstrated no definite tumor enhancement and FLAIR showed that the brain edema had resolved (Fig. 2G). He was discharged our hospital with a mild disorientation after 3 courses of high-dose methotrexiate treatment.

Discussion

The radiological features of unusual ICH location, heterogeneity of the hematoma, and extremely extensive brain edema

at the onset of hemorrhage suggested a malignant tumor, possibilities including metastasis, glioblastoma, and malignant lymphoma. However, a PCNSL was excluded because his IL-2R concentration of 1000 U/mL was not high enough to reliably indicate malignant lymphoma, the enhanced mass adjacent to the falx with a dural tail sign on MRI implied an extraparenchymal tumor such as a meningioma, and no enhancement was identified in the hematoma. Because massive ICH is an extremely rare presentation of PCNSL [1–7], a large ICH on presentation is generally considered to exclude PCNSL from the differential diagnosis [6]. To the best of our knowledge, only 7 cases (5 males and 2 females) of PCNSL with ICH have been reported. Enhancement was detected in the hematomas in all of these cases, suggesting intratumoral bleeding [1–7]. Average age of these cases was 51.6 ± 14.1 (29-80) years old, and locations of the hematomas included midbrain in one case, left basal ganglia in 1, and left frontal lobe in 5. Our case is an 80-year-old man with hematoma in the left frontal lobe. Probably it is a coincidence, but it is a very interesting fact that 6 out of 8 patients, including our case, had bleeding in the left frontal lobe.

In those reports, brain metastases, malignant gliomas, and malignant lymphoma are radiologically listed as differential diagnoses, but it is very difficult to make a definitive diagnosis of PCNSL by imaging studies. The tumor in our case was located in the left cingulate gyrus and histological examination showed that the hemorrhage was within the tumor. Extensive brain edema and the massive ICH caused cingulate herniation that displaced the tumor toward the falx as if it was attached to it. The dural tail sign may have been caused by the reaction of the falx to the impinging lymphoma. These unusual factors made it difficult to correctly identify the tumor's location. However, it is also important to make full use of all imaging techniques for diagnosing tumors. Compared to meningioma, malignant glioma, and metastasis, blood flow in malignant lymphoma is low [8,9]. Therefore, if perfusion MRI had been performed, meningioma could have been ruled out, leading to the radiological diagnosis of malignant lymphoma.

It is controversial for PCNSL to achieve resection by open surgery or to perform biopsy. It was hard to reach to the definite diagnosis of malignant lymphoma during surgery in our case, and the authors resected the tumor as much as possible to prevent rebleeding from the tumor. It might have contributed to good results in our case, and Schellekes et al. [10] reported that overall survival time was significantly longer for superficial PCNSL by resection in open surgery than by diagnostic biopsy.

Patient consent

Written consent to publishing details of this patient was obtained from his family.

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