www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Professor of Clinical Neurosurgery, School of Medicine, State U. of NY at Stony Brook.

SNI: Neuro-Oncology

Editor
Mitsutoshi Nakada, MD
Kanazawa University, Ishikawa, Japan

ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

SNI
Open Access

Case Report

Primary extranodal marginal zone mucosa-associated lymphoid tissue-type B-cell lymphoma involving the dura: A case report

Ryo Hamamoto[1], Toshinari Kawasaki[1], Masashi Oda[1], Sosuke Sumiyoshi[1], Kosuke Hayashi[1], Tamaki Kobayashi[1], Yoshihiko Ioroi[1], Tatsuki Uchiyama[2], Motohiro Takayama[1], Masaaki Saiki[1]

Departments of [1]Neurosurgery and [2]Hematology, Japanese Red Cross Otsu Hospital, Otsu, Japan.

E-mail: Ryo Hamamoto - ky107.ryh@gmail.com; *Toshinari Kawasaki - toshi821@kuhp.kyoto-u.ac.jp; Masashi Oda - masashi@kuhp.kyoto-u.ac.jp; Sosuke Sumiyoshi - sosuke.sumi@gmail.com; Kosuke Hayashi - khayashi.neuro@gmail.com; Tamaki Kobayashi - kobayashitamak@gmail.com; Yoshihiko Ioroi - yioroi@kuhp.kyoto-u.ac.jp; Tatsuki Uchiyama - tatsuki@kuhp.kyoto-u.ac.jp; Motohiro Takayama - gara1016@gmail.com; Masaaki Saiki - seijinbyo@hotmail.com

***Corresponding author:**
Toshinari Kawasaki,
Department of Neurosurgery,
Japanese Red Cross Otsu
Hospital, Otsu, Japan.

toshi821@kuhp.kyoto-u.ac.jp

Received: 22 September 2023
Accepted: 09 March 2024
Published: 29 March 2024

DOI
10.25259/SNI_792_2023

Quick Response Code:

ABSTRACT

Background: Primary extranodal marginal zone mucosa-associated lymphoid tissue-type B-cell lymphoma (EMZMBCL), which presents as a dural mass, is a rare intracranial tumor that mimics a subdural hematoma or meningioma.

Case Description: A 49-year-old woman presented to our hospital with transient right upper limb paresis, dysarthria for 10 min, and ongoing right upper-limb numbness. Computed tomography (CT) of the head revealed extra-axial lesions in the left frontal and parietal lobes. Based on the initial CT findings in the emergency room, an acute subdural hematoma was suspected. However, meningiomas and other intracranial tumors were also listed as differential diagnoses because there was no history of head trauma or coagulation abnormalities on blood examination, and further imaging studies were performed. Imaging findings suggested a subdural neoplastic lesion. A partial resection was performed for the lesion. Based on histopathological and immunohistochemical examinations, the patient was diagnosed with EMZMBCL. Whole-brain and intensity-modulated radiation therapies were administered as adjuvant therapies. The patient was discharged without neurological deficits.

Conclusion: EMZMBCL is a rare disease that should be considered in the differential diagnosis of subdural lesions, especially when there is no history of trauma or abnormalities in the coagulation system. The patient had a favorable outcome after selecting radiotherapy as the adjuvant therapy.

Keywords: Intracranial tumor, Meningioma, Primary extranodal marginal zone mucosa-associated lymphoid tissue-type B-cell lymphoma, Radiotherapy, Subdural hematoma

INTRODUCTION

Primary central nervous system lymphoma (PCNSL) accounts for 2–4% of all central nervous system (CNS) tumors and is restricted to the CNS without other lesions.[2,4] Most PCNSLs are non-Hodgkin's lymphomas (NHLs) and diffuse large B-cell lymphomas (DLBCLs), which are high-grade tumors with poor outcomes.[6] On the other hand, an extranodal marginal zone mucosa-associated lymphoid tissue (MALT)-type B-cell lymphoma (EMZMBCL) is a low-

grade NHL that appears to arise from the meninges.[8] Because EMZMBCL proceeds from the dura mater, it can resemble a subdural hematoma or meningioma, which is often misleading in diagnostic imaging.[1,8] Herein, we report a patient with EMZMBCL who initially presented with radiographic findings suggestive of a subdural hematoma or meningioma.

CASE DESCRIPTION

A 49-year-old woman with no significant medical history presented to our hospital with transient paresis in the right upper limb, dysarthria for 10 min, and ongoing numbness in the right upper limb. Although she presented with numbness in the right upper limb, a neurological examination revealed no deficits in the emergency room. Computed tomography (CT) of the head revealed multiple high-density lesions in the left frontal and temporoparietal regions [Figures 1a and b]. Blood tests revealed no abnormalities in the coagulation system. Based on the initial CT findings in the emergency room, an acute subdural hematoma was suspected. As the follow-up CT did not show an increase in the subdural hematoma or worsening of symptoms, the patient was treated conservatively. However, meningiomas and other intracranial tumors were also listed as differential diagnoses because there was no history of head trauma or coagulation abnormalities on blood examination, and further imaging studies were performed. Contrast-enhanced T1-weighted imaging revealed an extra-axial mass with homogeneous contrast enhancement [Figures 1c and d]. Digital subtraction angiography of the external carotid artery revealed a tumor stain consistent with the lesion location [Figures 1e and f]. Blood examination revealed elevated levels of soluble interleukin-2 receptor. ^{18}F-fluorodeoxyglucose (FDG) positron emission tomography with CT revealed no significant high value in FDG uptake. Because the imaging findings were suggestive of a subdural neoplastic lesion, an open biopsy was performed to confirm the diagnosis of a subdural lesion. Intraoperative findings revealed a thickened dura mater and a substantial tumor [Figures 1g and h]. The biopsy lesion was subjected to histopathological examination. The biopsied specimens were confirmed as malignant lymphoma or infiltration of inflammatory lymphocytes in the frozen sections, and additional specimens were biopsied from the same lesion to complete the procedure.

Histopathological and immunohistochemical examination

Histopathological examination revealed small lymphocytic infiltration [Figure 2a]. Immunohistochemical analysis revealed a low MIB-1 Labeling Index (20%). The tumor exhibited positive immunoreactivity to the CD3, CD20, CD27, and insulin-like growth factor 2 messenger RNA-binding protein-3 (IMP3) [Figures 2b and c]. In contrast, CD5, CD10, and Bcl2 showed negative immunoreactivity.

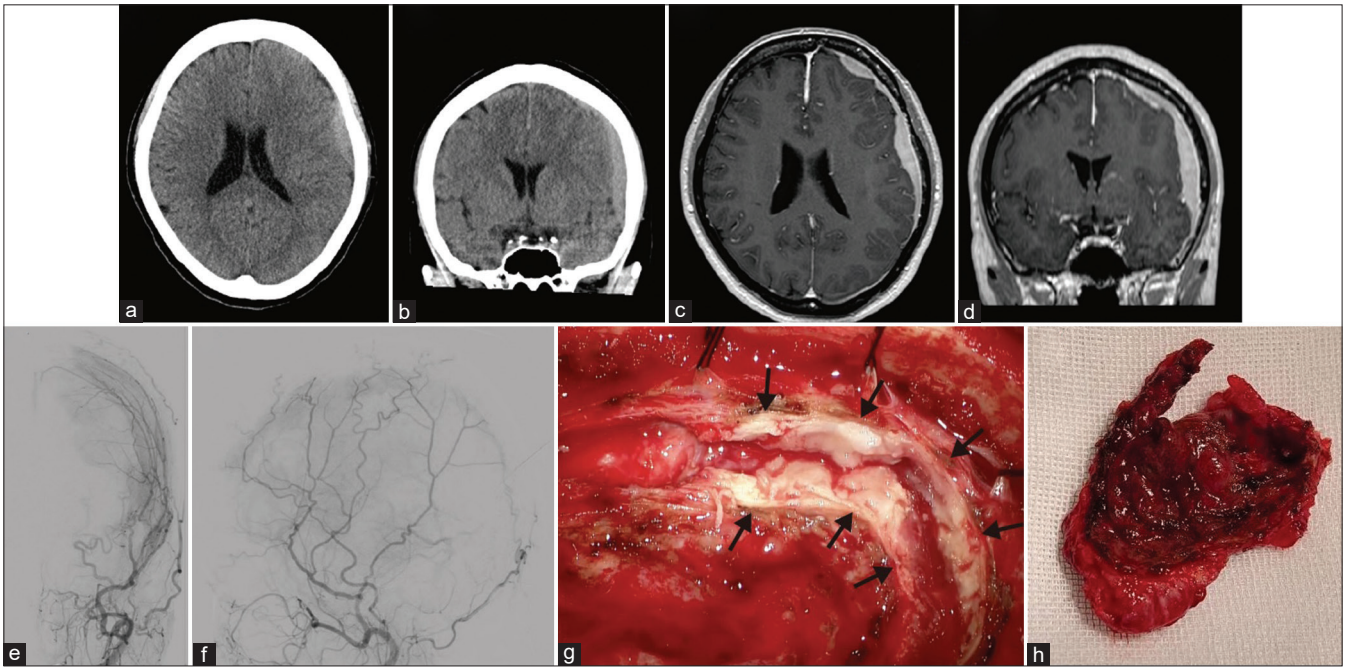

Figure 1: Axial (a) and coronal (b) computed tomography (CT) without contrast demonstrates a high-density subdural lesion. Axial (c) and coronal (d) magnetic resonance imaging with gadolinium contrast show residual contrast-enhancing extra-axial masses in the left frontal, temporal, and parietal lesions. Anteroposterior (e) and lateral (f) view of the left external carotid artery angiogram indicate arterial phase blush within the left convexity lesion. (g and h) Intraoperative findings showed a thickened dura mater (arrows) and a large tumor.

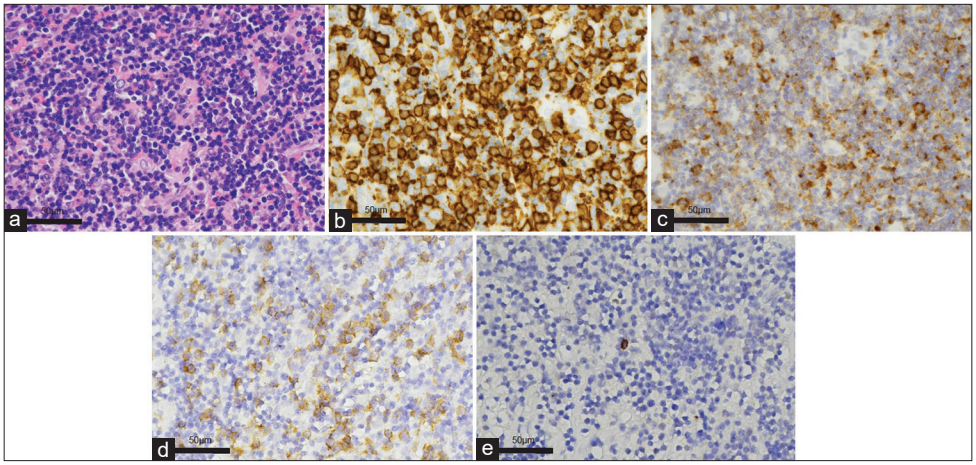

Figure 2: Histopathological and Immunohistochemical features of EMZMBCL. (a) Small lymphocytic infiltration (hematoxylin and eosin). (b) CD20 and (c) Insulin-like growth factor 2 mRNA-binding protein-3 (IMP3) immunohistochemical stains show diffuse positive. Immunohistochemical examination revealed immunoglobulin light-chain restriction (Kappa (d): Lambda (e) = 100:2). EMZMBCL: Extranodal marginal zone mucosa-associated lymphoid tissue-type B-cell lymphoma.

Immunohistochemical examination revealed immunoglobulin light chain restriction (Kappa: Lambda = 100:2) [Figures 2d and e]. Based on a comprehensive review of these findings, the patient was diagnosed with EMZMBCL.

Radiotherapy

Whole-brain (21 Gy in 14 fractions) and intensity-modulated radiation therapy (9 Gy in 6 fractions) were administered as adjuvant therapy. The patient was discharged without neurological deficits. Compared with the pretherapy magnetic resonance imaging (MRI) [Figures 3a and b], MRI performed 3 months after radiation therapy [Figures 3c and d] showed tumour disappearance. MRI performed eight months after radiation therapy revealed no recurrence [Figures 3e and f].

DISCUSSION

PCNSLs, which are extranodal NHLs, are most often DLBCLs and represent approximately 2–4% of intracranial neoplasms, which are high-grade tumors with poor outcomes.[2,4] EMZMBCL, which is a low-grade indolent-type NHL, is a rare subtype of PCNSL.[12] Although its incidence is very low, EMZMBCL cases have been reported.[3,7] There are few MALT lymphomas in the CNS; however, MALT lymphomas, which mostly occur in the stomach, also occur in the mucosa of other organs, including the lungs, salivary glands, bladder, and lacrimal glands, as well as in tissues without mucosa, such as the thyroid gland, breast, and skin.[3] Gastric MALT lymphoma arises from MALT cells acquired as a consequence of chronic *Helicobacter pylori* infection.[13] In other organs, including the skin, ocular adnexa, small intestine, and lungs, other bacterial infections have been suggested to be associated with MALT lymphoma.[13] Although the etiology of EMZMBCL remains unknown, chronic inflammation may contribute to the pathogenesis of EMZMBCL. Furthermore, some biological similarities have been hypothesized between meningothelial cells in the arachnoid membrane and epithelial cells at the sites where EMZMBCL usually originates.[9] Because EMZMBCL most commonly arises at the convexities, falx, tentorium, and cavernous sinus, the radiographic findings of EMZMBCL mimic those of subdural hematoma or meningioma, which show a dural tail sign on MRI. In contrast, the DLBCL variant of primary CNS high-grade lymphoma is typically located in the brain parenchyma.[5]

La Rocca *et al*. and Bustoros *et al*. have reported 126 and 104 cases of EMZMBCL, respectively.[3,7] They described EMZMBCL as a rare condition with a relatively good prognosis, and radiation therapy, chemotherapy, and combination therapy were the treatments of choice after the surgery procedure. EMZMBCL exhibits a marked female predilection in middle age (median: approximately 50 years, range: 22–78). However, the reasons for this predominance remain unknown. Various therapeutic options have been developed due to the lack of high-volume clinical trials and definite treatment protocols. Most patients in the reported cases underwent major surgical procedures (77.8%) or biopsies (16.7%) to obtain tissues and establish a diagnosis or perform a resection of the lesion. The reason for the high number of major surgical procedures may be that EMZMBCL was not initially suspected; rather, meningioma or subdural hematoma was suspected, and the procedure was intended for total removal.

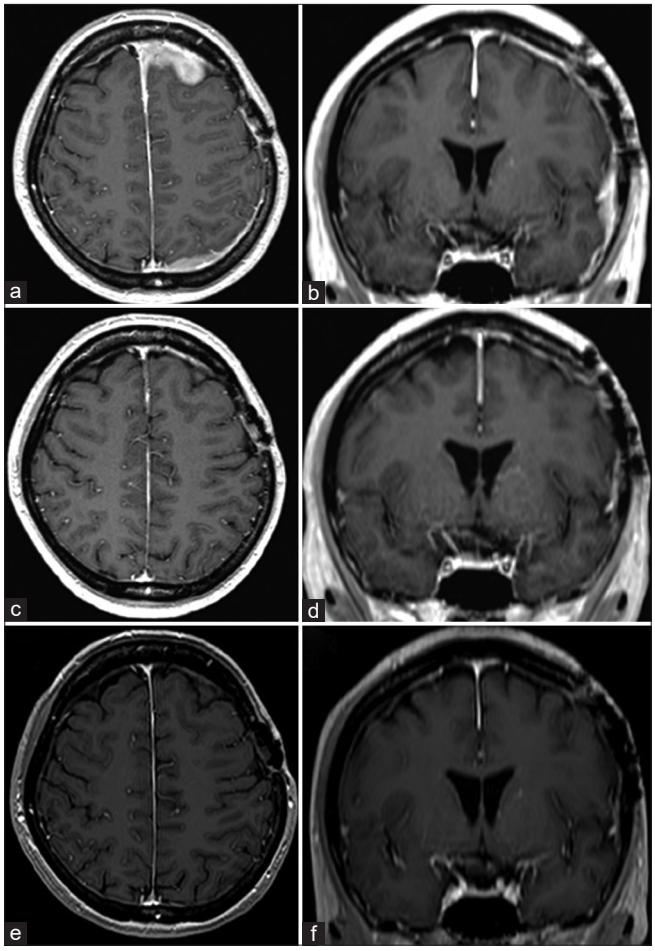

Figure 3: Preradiotherapy magnetic resonance imaging (MRI). Axial (a) and coronal (b) MRI with gadolinium contrast show residual contrast-enhancing extra-axial masses in the left frontal, temporal, and parietal lesions. The thickening and enhancement of the dura extend along the falx and the left frontal and parietal lobes. MRI at three months after radiation therapy. Axial (c) and coronal (d) MRI with gadolinium contrast indicate the disappearance of residual contrast-enhancing extra-axial masses in the left frontal, temporal, and parietal lesions and normalization of the dural thickening and enhancement. MRI at eight months after radiation therapy. Axial (e) and coronal (f) MRI with gadolinium contrast reveal no recurrence.

MALT lymphoma shows expression of pan-B cell markers (CD19, CD20, and CD79a) but does not express CD5 and CD10.[3] MIB-1 Labeling Index is typically low (10–30%).[3] To confirm the diagnosis, the predominance of CD20 and kappa light chain restriction is crucial.[6] In the present case, tumor was consistent with the findings of the immunohistochemical examination, leading to a diagnosis of EMZMBCL.

Radiotherapy significantly prolongs overall survival in both gastric and nongastric extranodal marginal zone lymphomas.[3] EMZMBCLs are highly radiosensitive, and surgery followed by focal low-dose radiotherapy has shown good outcomes.[10] In the present case, whole-brain (21 Gy in 14 fractions) and intensity-modulated radiation therapy (9 Gy in 6 fractions) were administered as adjuvant therapies, resulting in favorable tumor control with no recurrence of EMZMBCL. Although chemotherapy may expose patients to various risks, it may be necessary in cases of secondary or recurrent EMZMBCL.[11] Therefore, radiation therapy alone may be sufficient when tumor is well-controlled.

CONCLUSION

Here, we reported the case of a patient with EMZMBCL who initially presented with radiographic findings suggestive of a subdural hematoma. EMZMBCL is a rare disease that should be considered in the differential diagnosis of subdural lesions, especially when there is no history of trauma or coagulation system abnormalities. In this case report, the patient had a favorable outcome following partial tumor resection and adjuvant radiotherapy.

Ethical approval

This study was approval by the hospital ethics committee (No. 807, August 16, 2023).

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

REFERENCES

1. Ariani R, Ballas L. Primary CNS extranodal marginal zone B-cell lymphoma: A case series of 2 patients treated with external beam radiation therapy. Case Rep Oncol 2021;14:725-32.
2. Batchelor TT. Primary central nervous system lymphoma: A curable disease. Hematol Oncol 2019;37(Supplement 1):15-8.
3. Bustoros M, Liechty B, Zagzag D, Liu C, Shepherd T, Gruber D, *et al*. A rare case of composite Dural extranodal marginal zone lymphoma and chronic lymphocytic leukemia/small lymphocytic lymphoma. Front Neurol 2018;9:267.
4. Ferreri AJ, Marturano E. Primary CNS lymphoma. Best Pract Res Clin Haematol 2012;25:119-30.
5. Haldorsen IS, Espeland A, Larsson EM. Central nervous system lymphoma: Characteristic findings on traditional and advanced imaging. Am J Neuroradiol 2011;32:984-92.
6. Kim MY, Kim SM, Chung SY, Park MS. Dural marginal zone lymphoma confused with meningioma en plaque. J Korean Neurosurg Soc 2007;42:220-3.
7. La Rocca G, Auricchio AM, Mazzucchi E, Ius T, Della Pepa GM, Altieri R, *et al*. Intracranial Dural based marginal zone MALT-type B-cell lymphoma: A case - based update and literature review. Br J Neurosurg 2021;26:1-7.
8. Lopetegui-Lia N, Delasos L, Asad SD, Kumar M, Harrison JS. Primary central nervous system marginal zone B-cell

lymphoma arising from the Dural meninges: A case report and review of literature. Clin Case Rep 2020;8:491-7.
9. Park I, Huh J, Kim JH, Lee SW, Ryu MH, Kang YK. Primary central nervous system marginal zone B-cell lymphoma of the basal ganglia mimicking low-grade glioma: A case report and review of the literature. Clin Lymphoma Myeloma 2008;8:305-8.
10. Takeuchi I, Tanei T, Kuwabara K, Kato T, Naito T, Koketsu Y, *et al*. Primary Dural mucosa-associated lymphoid tissue lymphoma mimicking falx meningioma: A case report. NMC Case Rep J 2022;9:123-8.
11. Tsutsumi Y, Ito S, Nagai J, Tateno T, Teshima T. Patients with marginal zone Dural lymphoma successfully treated with rituximab and bendamustine: A report of two cases. Mol Clin Oncol 2021;15:208.
12. Tu PH, Giannini C, Judkins AR, Schwalb JM, Burack R, O'Neill BP, *et al*. Clinicopathologic and genetic profile of intracranial marginal zone lymphoma : A primary low-grade CNS lymphoma that mimics meningioma. J Clin Oncol 2005;23:5718-27.
13. Zucca E, Bertoni F. The spectrum of MALT lymphoma at different sites: Biological and therapeutic relevance. Blood 2016;127:2082-92.

How to cite this article: Hamamoto R, Kawasaki T, Oda M, Sumiyoshi S, Hayashi K, Kobayashi T, *et al*. Primary extranodal marginal zone mucosa-associated lymphoid tissue-type B-cell lymphoma involving the dura: A case report. Surg Neurol Int. 2024;15:113. doi: 10.25259/SNI_792_2023