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Surgical Neurology International

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SNI: Neuro-Oncology

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Case Report

Mucosa-associated lymphoid tissue lymphoma of the dura mimicking meningioma: A case report

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Received: 09 November 2023
Accepted: 05 February 2025
Published: 28 February 2025

DOI
10.25259/SNI_902_2023

Quick Response Code:

ABSTRACT

Background: Primary central nervous system lymphomas (PCNSLs) are relatively infrequent tumors and are usually high-grade and aggressive neoplasms. A small portion of PCNSLs are low-grade lymphomas and can involve the dura. Mucosa-associated lymphoid tissue (MALT) lymphoma of the dura is an extremely rare subtype with only case reports and series documented in the literature.

Case Description: A 65-year-old woman presented with a history of headaches followed by progressive left hemiparesis. Imaging studies showed an extra-axial dural-based tumor causing midline shift. Gross total resection was achieved, and the patient was discharged without postoperative complications. Histopathological examination confirmed the diagnosis of MALT lymphoma of the dura. The patient was evaluated by the oncologist and received adjuvant chemotherapy. At the 10-month follow-up, the patient experienced remission of her symptoms, and the last magnetic resonance imaging showed no evidence of tumor recurrence.

Conclusion: MALT lymphoma of the dura diagnosis requires a high level of suspicion because it can often mimic meningioma. Given its rarity, there is no consensus on the standard treatment strategy. Gross total resection followed by adjuvant therapy is an accepted treatment to manage these cases.

Keywords: Dura, Meningioma, Mucosa-associated lymphoid tissue lymphoma

INTRODUCTION

Primary central nervous system lymphoma (PCNSL) is a rare form of extranodal non-Hodgkin lymphoma that can develop in the brain, spine, cerebrospinal fluid, and eyes.[6] PCNSLs represent approximately 1.9% of all central nervous system tumors.[13] Most PCNSLs are diffuse large B-cell lymphomas (90%), which are high-grade and aggressive neoplasms and, on rare occasions, are Burkitt, T-cell, or low-grade lymphomas.[7]

Marginal zone-B lymphoma (MZBL) is a low-grade subtype that accounts for 7% of all non-Hodgkin lymphomas.[4] There are three subtypes of MZBL: Extranodal MZBL (60.8%) or mucosa-associated lymphoid tissue (MALT), nodal MZBL (30.3%), and splenic MZBL (8.9%).[1] MZBL was initially described as an indolent lymphoma in MALT of the gastrointestinal tract.[15] Other extranodal anatomic sites involved are the stomach, ocular adnexal, skin, lung, and salivary

gland.[4] MZBL developing in the central nervous system (CNS) is an extremely rare entity, and fewer than 130 cases involving the dura have been reported in the literature.[10] Herein, we report the case of MALT lymphoma of the dura misdiagnosed as meningioma in a 65-year-old woman and describe the disease course, histopathology diagnosis, and treatment.

LITERATURE REVIEW

We conducted a literature search of articles written in English using PubMed from inception to 2024. Search terms included: ((dural) OR (meningeal)) AND ((mucosa-associated) OR (marginal zone lymphoma) OR (malt)). This search yielded 48 articles representing 179 cases of MALT lymphomas involving the dura. From this, we only included 151 cases in which intracranial involvement was described. Demographic data, tumor location, and treatment modality were collected. Information about treatment was divided into: only surgery, surgery + radiotherapy (RT), surgery + chemotherapy (CT), surgery + CT + RT, only RT, only CT, and not specified. Statistical analysis was performed using the software Statistical Package for the Social Sciences (version 25.0; IBM Corporation, Armonk, New York, USA).

RESULTS

We found 151 cases of MALT lymphomas involving intracranial dura. There was a prevalence of females (74.8%; *n*: 113) over males (25.1%; *n*: 38), and the mean age was 53.02 (±11.7) years. In order of frequency, the convexity was the most affected area (68%, *n*: 102), followed by tentorium (9.9%; *n*: 15), cavernous sinus (7.2%; *n*: 11), falx (5.2%, *n*: 8), and posterior fossa (3.9%; *n*: 6). There were other uncommon sites such as sellar/suprasellar region, sphenoid wing or clivus (5.9%; *n*: 9).

We found a wide spectrum of treatment modalities involving one or more procedures. The most frequent treatment was surgery + RT (49.6%; *n*: 75), followed by surgery + CT (13.9%; *n*: 21), only surgery (13.9%; *n*: 21), surgery + RT + CT (8.6%; *n*: 13), only RT (3.3%; *n*: 5), and only CT (2.6%; *n*: 3). In 12 cases, the modality of treatment was not specified (7.9%) [Figure 1]. The mean time of follow-up was 38.2 months (*n*: 119; ±42.4).

CASE PRESENTATION

A 65-year-old female with a medical history of hypertension and diabetes presented with a 3-month history of asthenia, adynamia, and disorientation. She also experienced progressive left hemiparesis and worsening headaches during the past month. On clinical examination, dense left-sided hemiparesis was revealed, and pyramidal signs were present.

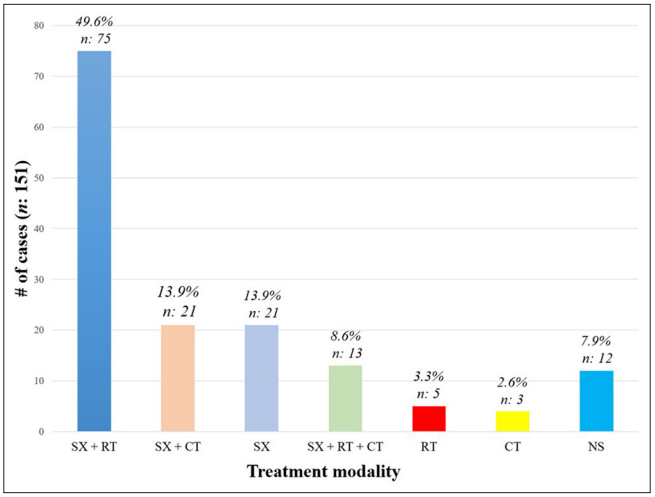

Figure 1: This graph shows the number and percentage of different treatment modalities used for patients with dural MALT lymphomas. SX: surgery, RT: Radiotherapy, CT: Chemotherapy, NS: Not specified.

Cranial magnetic resonance imaging (MRI) demonstrated a right extra-axial dural-based lesion arising from frontal convexity with perilesional edema and mass effect on the adjacent brain parenchyma [Figures 2a and b]. After the administration of gadolinium, the lesion showed avid and homogeneous enhancement, measuring 5.7 × 3.8 × 4.9 cm [Figures 2c and d]. These findings suggested the diagnosis of a convexity meningioma, and a regimen of 4 mg of intravenous dexamethasone 4 times a day was started before surgery.

Surgical resection

The patient underwent surgery in a supine position with the head rotated to the left in three-point fixation in the Mayfield clamp. A Falconer incision was performed, followed by a right frontoparietotemporal craniotomy. A soft brownish tumor attached to the dura was resected. Hemostasis was secured, and the dura was closed with a dural graft. Bony infiltration was observed, and a cranioplasty with methyl methacrylate was performed. The postoperative course was unremarkable, and the patient was discharged on the 3rd postoperative day.

The neuropathology team received a specimen characterized by a nodular tumor attached to the dura that showed a brown-tan external surface with small vessels [Figure 3a]. The gross section was solid, soft, and discreetly spongy with heterogeneous shades of brown color [Figure 3b]. The histological analysis showed a low-grade lymphoid tumor with a diffuse pattern and some secondary follicles. The neoplasm was composed of small, rounded lymphocytes, plasma cells, and some dendritic cells in the germinal centers [Figures 3c and d]. The neoplastic cells expressed CD20 and CD43 antigens by immunohistochemistry. Furthermore, there was expression of CD138 in plasma cells and cytoplasmic lambda light chain restriction [Figure 4]. These

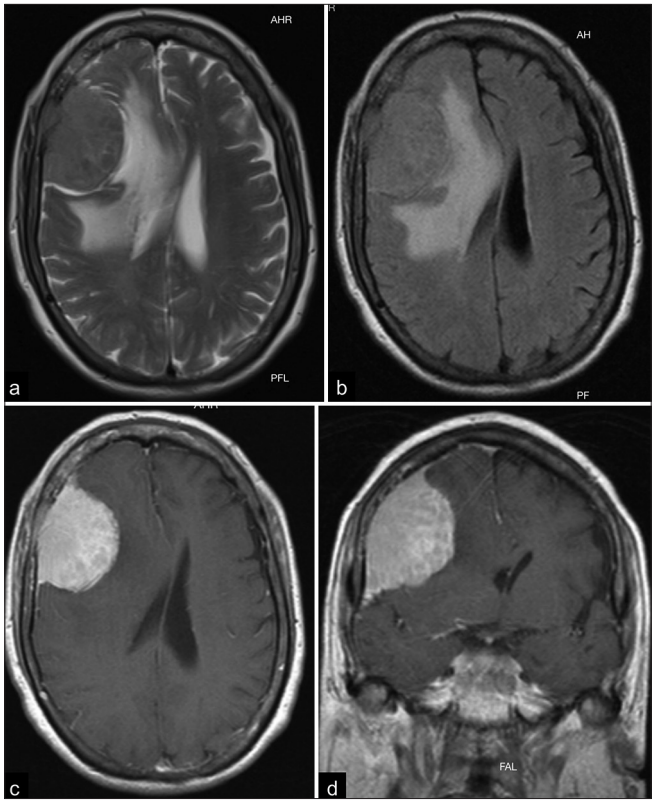

Figure 2: Preoperative magnetic resonance imaging. (a and b) Both T2-weighted imaging and T2 fluid-attenuated inversion recovery (FLAIR) images demonstrate a remarkable mass effect on the surrounding brain and vasogenic edema. (c and d) Axial T1-weighted and coronal T1-weighted images showing a right frontal extra-axial lesion with homogeneous and avid enhancement.

findings were consistent with MALT lymphoma of the dura with plasmacytic differentiation.

The patient was evaluated by the oncologist for potential adjuvant treatment. Four courses (every 3 weeks) of intravenous methotrexate-cytarabine combination therapy were administered: methotrexate 3.5 g/m² on day 1 and cytarabine 2 g/m² on days 2 and 3; repeated treatment. The patient underwent a whole-body positron emission tomography–computed tomography scan without evidence of an extracranial tumor location. At 10 months after surgery, she experienced improvement of hemiparesis, and the last MRI demonstrated no evidence of tumor recurrence [Figure 5].

DISCUSSION

PCNSLs represent 4-6% of extranodal lymphomas.[7] Among low-grade PCNSLs, the MALT subtype is the most common, and it is usually limited to the meninges with no systemic disease at the time of presentation.[3] The pathogenesis of primary MALT-type MZBL of the CNS is not well understood. In the CNS, there is no MALT tissue, but it has been hypothesized that meningothelial cells are analogous to epithelial cells at sites where extranodal MZBL arises.[3,8] MZBL has been associated with multiple genetic abnormalities, such as trisomy 3, which results in overexpression of the proto-oncogene BCL-6; inactivation of TNFAIP3 in cases with plasmacytic differentiation; or activating NOTCH2 mutations accompanied by inactivating TBL1XR1 mutations in cases with monocytoid morphology.[16]

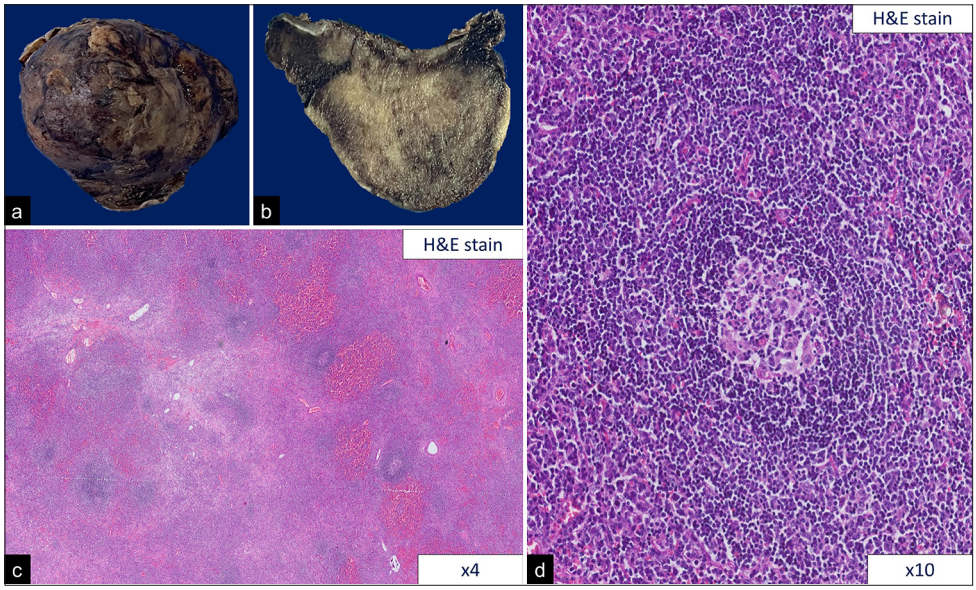

Figure 3: (a and b) The external surface and gross section are shown, and the tumor was solid, dark brown and dural attached. (c and d) Hematoxylin and eosin (H&E) staining showed a diffuse pattern with secondary follicle formation, x4 and x10.

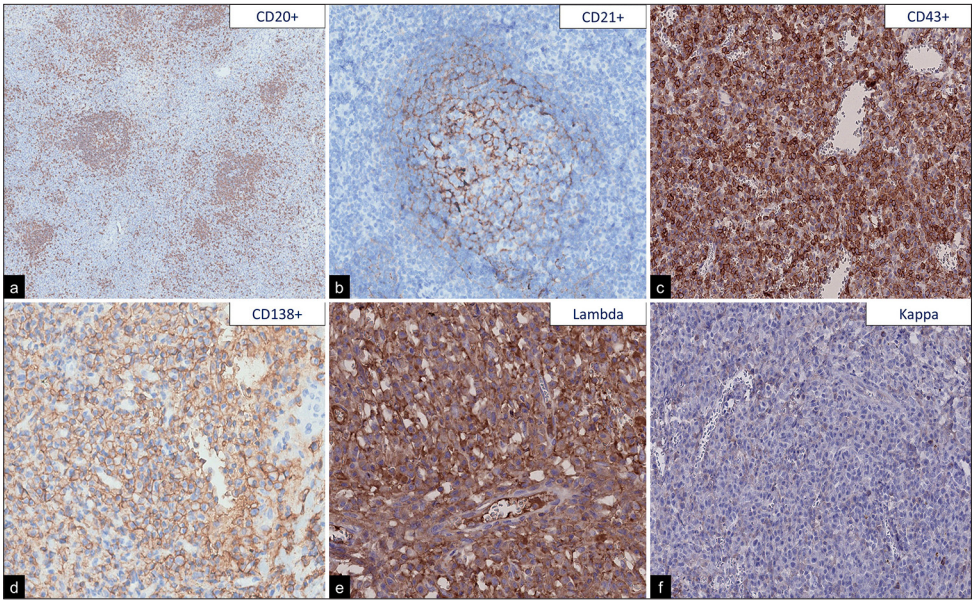

Figure 4: Immunohistochemistry. (a) Diffuse and follicular CD20-positive B cells, ×4. (b) Dendritic cells in the germinal center CD21 positive, ×10. (c) CD43-positive neoplastic cells, ×10. (d) CD138-positive plasma cells, ×10. (e and f) Lambda light chain restriction with negativity for Kappa, ×10.

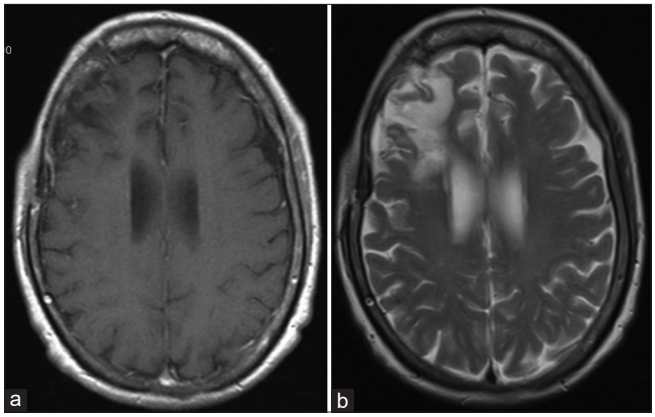

Figure 5: Postoperative magnetic resonance imaging. (a and b) Axial T1-weighted with gadolinium and axial T2-weighted images showing postsurgical changes and gross total resection.

The pathological features of MZBL arising in the CNS are similar to those of MZBL in other extranodal sites. They are composed of small or medium size lymphocytes and marginal zone cells, sometimes with remnants of reactive follicles with follicular colonization. Neoplastic cells show expression of pan-B cell markers (CD20, CD79a) and lack expression of CD5, CD10, CD23, BCL6, and cyclin D1.[16] Often, MALT lymphomas have plasmacytic differentiation and show restricted Ig light chains with a preponderance of κ light chain restriction.[3,16]

MALT lymphomas of the dura have a female predilection with a female-to-male ratio of 3:1. The median age at presentation was approximately 51 years (range: 28–77 years).[10] The clinical presentation of PCNSL varies depending on the CNS compartment involved. In 70% of patients, a focal neurological deficit is observed when parenchyma or leptomeninges are involved, which leads to prompt imaging diagnosis.[7] The patients may have symptoms such as headache, fatigue, seizures, visual loss, dizziness, or hemiparesis. Some cases have underlying autoimmune or infectious diseases, including hepatitis C, Sjogren's syndrome, Graves' disease, and multiple sclerosis.[3,10] Our patient's presentation was progressive over a few months, causing a motor deficit and altered mental status. The patient had no active infection or associated autoimmune diseases.

In approximately 2% of cases, the imaging finding of an extra-axial dural-based lesion with homogeneous contrast enhancement on MRI is not a meningioma. The main differential diagnoses are metastases, solitary fibrous tumors, or lymphoproliferative diseases.[12] Radiologically, MALT lymphoma of the dura usually presents as a meningioma-like mass at cerebral convexities, but other less common sites of involvement are the parenchyma, cavernous sinus, choroid plexus, and cerebellopontine angle.[2] Dural lymphoma should be suspected when imaging demonstrates an extensive soft-tissue mass without relevant bone destruction.[14] Because radiological features are not sufficient to diagnose MZBL of the dura, other data, such as patient age, personal medical history, and risk factors for lymphomas, are useful to rule out other differential diagnoses.[11] In our patient, preoperative clinical and radiological features resembled a convexity meningioma, which caused a neurological deficit. Therefore, surgical treatment was chosen, and gross total

resection was achieved. Macroscopic evaluation of the gross section suggested an angiomatous meningioma subtype; however, the histopathological examination unexpectedly reported MZBL.

The standard management for dural MALT lymphomas is not clearly defined. Due to their rarity, they are not usually included in most of the guidelines for non-gastric MZBL treatment.[17] Modalities of treatment include surgical tumor resection, followed by RT, CT, or a combination which have been used.[9] In different series, the majority of patients underwent some form of surgical intervention. Most of the patients were treated with two modalities (64%): Resection plus RT, resection plus CT, or RT plus CT. The rest were treated with a single modality (27%), and in minor cases (9%), they were treated with all modalities combined.[3] Furthermore, histologic subtype is a major determinant of prognosis among patients with PCNSL. Patients with MALT lymphoma have favorable outcomes independent of treatment modality.[5] In selected cases, the extent of surgical resection has been demonstrated to be a positive prognostic marker for overall survival. Nevertheless, when maximal resection was chosen, lymphoma was not suspected in preoperative imaging.[9] In our case, after the surgical procedure, the patient was evaluated by the oncologist, and the adjuvant therapeutic modality was based on systemic CT.

Patients with MALT lymphomas of the dura have a better prognosis compared to other extranodal sites. In a recent review, encompassing 93 cases reported that the 5-year overall survival and progression free-survival were 96.7% and 81.2%, respectively. Strict clinical and radiological follow-up is necessary because disease recurrence has been reported between 12 and 40 months after surgery.[3] At the last follow-up, this patient's survival rate was reported to be 10 months with no evidence of relapse.

CONCLUSION

MALT lymphoma of the dura is a rare variant of PCNSL and usually presents as a dural-based lesion misdiagnosed as meningioma. The pathological diagnosis is made with the finding of small lymphocytes with diffuse patterns and immunohistochemical expression of B-cell markers. There is no standard management defined for this entity, but surgical treatment should be the first therapeutic step when a mass effect occurs, followed by adjuvant treatment.

Ethical approval: The Institutional Review Board approval is not required.
Declaration of patient consent: Patient's consent is not required as patients identity is not disclosed or compromised.
Financial support and sponsorship: Nil.
Conflicts of interest: There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation: The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript, and no images were manipulated using AI.

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How to cite this article: Falcón Molina JE, Lozano Guzmán I, Rodríguez Florido MA, Maciel Ramos E, Castillejo Adalid LA, Ascencio Montiel MA. Mucosa-associated lymphoid tissue lymphoma of the dura mimicking meningioma: A case report. Surg Neurol Int. 2025;16:63. doi: 10.25259/SNI_902_2023