

## Dural MALT lymphoma with disseminated disease

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### Abstract

Central nervous system (CNS) lymphoma involving the dura mater is very rare and histologically is usually a subtype of non-Hodgkin's lymphoma (NHL) termed mucosa-associated lymphoid tissue (MALT) lymphoma. We present a case of a 46-year old woman with dural MALT lymphoma that was found to also involve a lacrimal gland, inguinal lymph nodes, and bone marrow. Magnetic resonance imaging of the brain showed an extra-axial enhancing mass approximately 6 cm in maximum diameter along the right frontotemporal convexity. Histopathology of the resected dural mass showed MALT lymphoma expressing CD20, CD52, CD19, and CD38. Molecular studies of the B-cell receptor heavy chain demonstrated monoclonality at the involved sites. The patient was treated with four cycles of fludarabine, mitoxantrone, and rituximab with complete remission. She had recurrence in the subcutaneous tissue of the back at 12 months but has remained free of intracranial disease for 31 months. A review of the literature reveals 57 cases of dural MALT lymphoma. Only 4 had extra-CNS involvement at presentation, and only 3 had local recurrence of the dural tumor. Because of the indolent behavior of this tumor, the intracranial portion can be treated conservatively after resection with or without chemotherapy. Deferral of brain radiation can be considered with close clinical and neuroimaging follow up.

### Introduction

Central nervous system (CNS) involvement with non-Hodgkin's lymphoma (NHL) falls into one of three categories: primary CNS lymphomas (PCNSL), disseminated lymphoma with CNS involvement, and primary dural lymphoma. Primary CNS lymphomas are typically diffuse large B-cell lymphomas and are found in the brain parenchyma and other CNS structure.<sup>1</sup> Disseminated lymphoma typically involves the meninges in a leptomeningeal pattern similar to leukemia.<sup>2</sup> It can also be

found intraparenchymally but does not typically occur within the dura.<sup>2-4</sup> A small proportion of CNS lymphomas are extraaxial, intradural lymphomas that typically do not involve the brain parenchyma.<sup>5</sup> The vast majority of these are low grade extranodal marginal zone lymphomas, also called mucosa-associated lymphoid tissue (MALT) lymphomas (primary dural marginal zone lymphoma, PDMZL),<sup>5,6</sup> although a small number of higher grade lymphomas have been reported including diffuse large B-cell and follicular lymphoma.<sup>5</sup> Distinguishing among types of CNS lymphoma can be difficult, and the distinction is sometimes obscured in the literature due to lumping of all *meningeal* lymphomas together. In this review, a dural lymphoma refers to a solid, localized mass, within or indistinguishable from the dura mater, that radiographically may have a dural tail. In contrast, leptomeningeal lymphoma is diffusely spread over the surface of the brain, involving the other meningeal layers but not primarily the dura. When defined in this way, dural lymphoma is relatively rare.

We now report a case of disseminated MALT lymphoma with involvement of the dura and discuss the management of this case in light of the current available literature on such cases.

### Case Report

A 46-year old right handed Caucasian female presented with a small lump in her right upper inner eyelid. This was initially dismissed, but the patient presented again with pressure symptoms in the right frontal region of her head and blurring of vision of the right eye. An MRI of the brain showed a 2×6×6 cm extra-axial enhancing mass along the right frontal convexity, extending anteriorly to the superior ridge of the orbit. There was no significant edema around the lesion, which was initially felt to be consistent with an *en plaque* meningioma (Figure 1). On further review, the lesion was found to be extending across the midline with thickening and enhancement of the left frontal dura mater. There was also increased T2 signal in the adjacent cortical sulci with subtle contrast enhancement. The overlying bone marrow also showed abnormal signal, indicative of an infiltrative process. The patient was referred to our institution for a second opinion regarding diagnosis and management. On physical exam she was neurologically intact and positive findings included a small nodule palpable in the right upper palpebra in a supranasal location. She had no palpable lymphadenopathy except shotty inguinal nodes. The dural mass was resected via a right frontal craniotomy. The pathology was consistent with MALT lymphoma expressing CD20, CD52, CD19, and CD38, but not CD5, with

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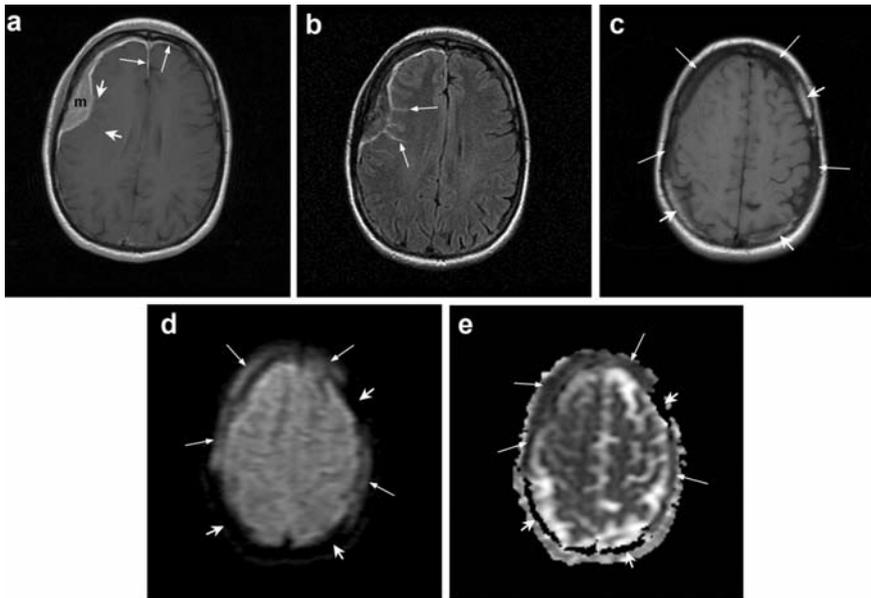
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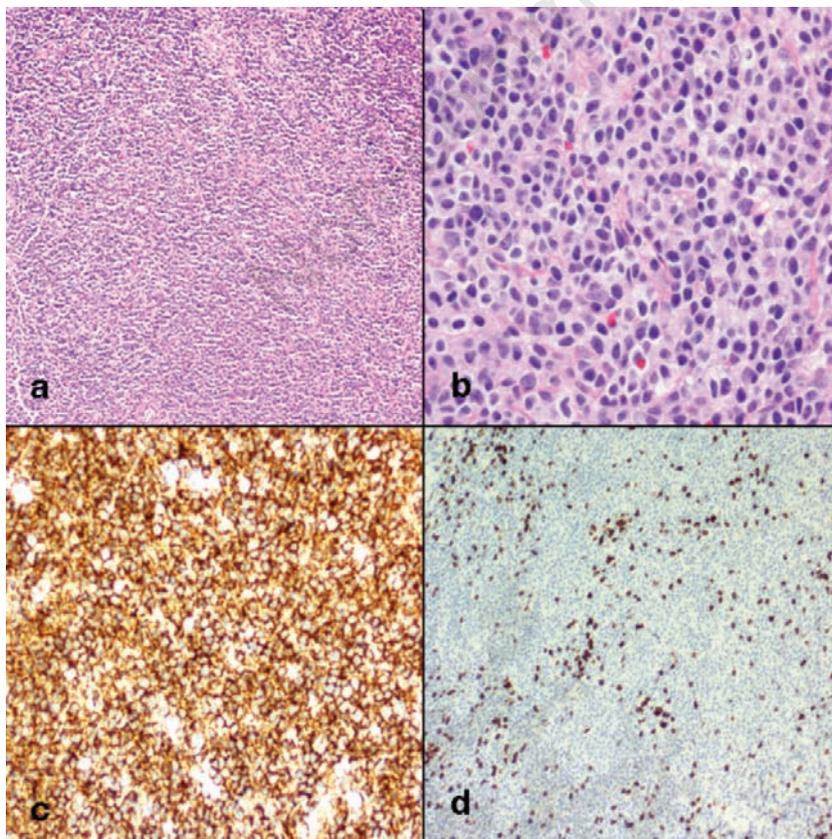
kappa light chain restriction (Figure 2).

A PET-CT scan of the whole body showed shotty cervical, axillary, and inguinal lymph nodes that demonstrated mildly increased metabolic activity -standardized uptake value (SUV) in the 2-4 range with the highest values in inguinal regions. A biopsy of a left inguinal node showed lymphoid proliferation and flow cytometry studies were consistent with a kappa-restricted B-cell neoplastic proliferation. The patient also had an anterior orbitotomy and biopsy of the orbital mass that demonstrated lacrimal gland involvement by MALT lymphoma with the same immunophenotype as the lymph node and the dural mass. Bone marrow biopsy showed a small population (2.6% of nucleated cells) of kappa-restricted B-lymphocytes by flow cytometry, but spinal fluid was negative.

For the tissue taken from the orbit, the dura and the lymph nodes, PCR amplification of the variable region of the B-cell receptor heavy chain (according to published methods,<sup>7</sup>) resulted in a single band of about 250 base pairs on gel electrophoresis (Figure 3). PCR of the bone marrow failed to yield a defined single band. The PCR products were sent for sequencing, and sequence analysis indicated that the sequences from the orbit and the dura



**Figure 1.** Brain MR imaging. (a) Post-contrast axial T1-weighted image shows a homogeneously enhancing extra-axial dural-based mass (m) in the right frontal region. The thickening and enhancement of the dura extends anteriorly along the falx and along the left frontal lobe (long arrows). There is also subtle enhancement extending into a few of the adjacent cortical sulci (short arrows). (b) Corresponding axial fluid-attenuated inversion recovery (FLAIR) image reveals abnormal hyperintensity within the cortical sulci (arrows) adjacent to the lesion, corresponding to subtle enhancement in A). (c) Non-enhanced axial T1-weighted image at a slightly higher level demonstrates loss of normal bone marrow hyperintensity in the calvarium adjacent to the dural mass and in the left parietal bone (long arrows). Note normal bright bone marrow (short arrows). (d) Corresponding diffusion-weighted image (DWI) reveals signal intensity similar to the dural mass within the affected bone marrow (long arrows). Note normal bone marrow without any signal (short arrows). (e) Corresponding apparent diffusion coefficient (ADC) map shows that the diffusion of water molecules within the affected bone marrow is similar to the dural lesion (long arrows). Normal bone marrow remains black without any signal (short arrows).



had 99.2% identity. The sequence from the lymph nodes was of lesser quality, but also had a very high identity with the dural sequence (80.1%), indicating that the lymphoid cells in these tissues were clonally related.

The patient was treated with intravenous fludarabine, mitoxantrone, and rituximab every four weeks for a total of four cycles. The patient did not receive radiation treatment to the dural lesion; the decision was made to follow her closely with serial MRIs to monitor for intracranial recurrence.

The patient relapsed with biopsy-proven MALT lymphoma in subcutaneous tissues of the back and buttock 12 months after diagnosis. A bone marrow biopsy at that time was negative for malignancy. She was treated with additional rituximab and then maintained on rituximab. At 33 months since diagnosis she is again in complete remission and has remained free of CNS disease.

## Materials and Methods

Several thick slices of paraffin embedded tissue were provided by the pathology laboratory. Genomic DNA was extracted using the Qiagen DNeasy Blood & Tissue kit. Semi-nested PCR was performed as previously described<sup>7,8</sup> using the following degenerate primers:

FR2a: TGG(A/G)TCCG(C/A)CAG(G/C)C(T/C)(T/C)CNGG

LJH: TGAGGAGACGGTGACC

VLJH: GTGACCAGGGTNCCTTGGCCCCAG

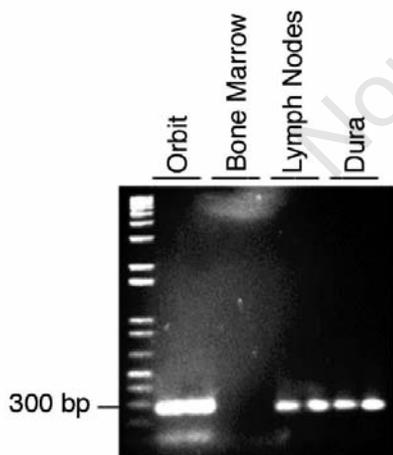
Primers were ordered from Integrated DNA Technologies (IDT®). PCR conditions were as follows: denaturation at 94°C for 5 min, followed by 40 cycles of 94°C for 2 min, 50°C for

**Figure 2.** Morphology and immunohistochemical staining of dural mucosa-associated lymphoid tissue (MALT lymphoma) (a) At low magnification there is a diffuse lymphocytic infiltration of the dura with characteristic perivascular pattern. Follicular structures were not identified. (original magnification x100). (b) At higher magnification the neoplastic cells can be seen to exhibit a morphologic spectrum with abundant pale agranular cytoplasm and small to medium-sized nuclei with round to irregular nuclear borders. Rare large centroblast-like cells and plasma cells are present (original magnification x400). (c) CD20 immunohistochemical stain shows diffuse positivity (original magnification x50). (d) CD3 immunohistochemical stain shows only scattered positive cells (original magnification x50).

2 min, and 72°C for 2 min, followed by elongation at 72°C for 20 min. Genomic DNA was first amplified with the FR2a and LJH primers, then a 1:5000 dilution of the PCR product was amplified with the FR2a and VLJH primers. Products were analyzed by electrophoresis on a 2% agarose gel and then extracted using Qiaquick Gel Extraction Kit®. PCR products were sent for sequencing at the MUSC Biotechnology Resource Laboratory. Some sequences were corrected by visually calling nucleotides from the chromatogram using ChromasLite 2.01 from Technelysium Pty Ltd.® (<http://www.technelysium.com.au>). Sequences were analyzed using Biology Workbench 3.2 (<http://seqtool.sdsc.edu>).

## Discussion

This report describes a patient with disseminated low grade MALT lymphoma that involved the dura. The histological evidence showed that a B-cell malignancy was disseminated at presentation, involving the lacrimal gland, lymph nodes, bone marrow and intracranial dura. Examination of all of these tissues revealed a MALT lymphoma. This tissue distribution suggests that the patient had a subclinical MALT lymphoma that became disseminated. Laboratory studies proved that the lymphoid proliferations were neoplastic and not reactive or inflammatory processes. PCR amplification of the IGH variable region in the samples from dura, lymph node and lacrimal gland demonstrated a single band of the same size, indicating monoclonality. In addition, sequencing of this segment demonstrated fur-



**Figure 3.** PCR amplification of the variable region of the B-cell receptor heavy chain resulted in a single band for tissue taken from the orbit, lymph node and dura, demonstrating that rearrangement has occurred and the B-cells are monoclonal. PCR of the bone marrow failed to yield a defined single band.

ther that the gene rearrangement was identical in this region.

Dural lymphomas pose special diagnostic and therapeutic challenges. Because they are uncommon, they are often initially misdiagnosed as meningiomas or other lesions based on imaging.<sup>3,9-42</sup> Intracranial lymphomas may be epidural, dural, subdural, subarachnoid, and combinations of these.<sup>43</sup> A dural mass with associated leptomeningeal and/or bone marrow imaging abnormalities is highly unusual for meningioma and is suggestive of lymphoma or metastatic malignancy. Since the therapy for meningioma and dural lymphoma are different, several authors have stressed the importance of getting a tissue diagnosis before treatment of a dural lesion identified on MRI.<sup>15,31,33,44</sup>

The majority of dural lymphomas are MALT lymphomas.<sup>5</sup> MALT lymphoma is a type of B-cell lymphoma that has the characteristics of marginal zone cells and typically arises from nests of lymphoid cells in mucosal tissues. Occasionally it is found to arise in non-mucosal sites such as skin, orbit or dura.<sup>45,46</sup> MALT lymphoma was originally described by Isaacson and Wright in 1983.<sup>47</sup> Since that time, clinical experience with this entity has steadily grown, as has the understanding of its etiology and pathogenesis. According to the Revised European – American Classification of Lymphoid Neoplasms (REAL) and the subsequent update by the World Health Organization (WHO), Classification of Neoplasms of the Hematopoietic and Lymphoid Tissues, marginal zone B-cell lymphomas are a histologic category of the mature peripheral B-cell neoplasms.<sup>48,49</sup> Marginal zone B-cell lymphomas (MZLs) are further divided clinically into nodal, extranodal, and splenic types. Extranodal marginal zone B-cell lymphoma is equivalent to MALT lymphoma. While the earlier REAL classification considered extranodal and nodal MZLs to be histologically the same, only distinguishable by their clinical presentation, the newer WHO classification regards nodal MZLs to be a distinct entity with a worse prognosis than extranodal/MALT MZLs with nodal involvement.<sup>48,49</sup> MALT lymphomas usually arise in the gastrointestinal tract, especially the stomach, but are also found in the head and neck region (orbital tissues, salivary glands, and thyroid), skin, and lung.

MALT lymphoma generally has a good prognosis and can be treated with minimally toxic methods.<sup>5,50,51</sup> While MALT lymphoma usually remains localized for a long time, retrospective studies have shown that one third to one fourth of non-gastrointestinal cases had disseminated disease at diagnosis.<sup>46,50,51</sup> One report indicates that MALT lymphomas arising in non-gastrointestinal tract locations have a higher rate of dissemination than those arising in the GI tract.<sup>52</sup> MALT lymphomas have a ten-

dency to spread to non-contiguous mucosa associated sites.<sup>52</sup> Whether disseminated MALT lymphoma carries a poorer prognosis than localized disease is a subject of debate. In a series of patients studied by Arcaini *et al.*,<sup>45</sup> dissemination to lymph nodes or bone marrow was associated with shorter overall survival, while Thieblemont *et al.* found no difference in outcome between patients with disseminated versus localized disease.<sup>51,53</sup>

While MALT lymphomas remain indolent, there is a risk of transformation to higher grade lymphomas. In most cases, MALT lymphomas have been reported to transform into diffuse large B-cell lymphoma,<sup>54-57</sup> which has a much worse prognosis. Indolent lymphomas transform into higher grade lymphomas at a rate of 3% per year.<sup>58</sup>

No evidence based guidelines exist for treating MALT and the other types of marginal zone lymphoma, but current treatment strategies have been reviewed by Thieblemont and Coiffier.<sup>45</sup> Localized disease that is clearly linked to an infectious stimulus (such as *Helicobacter pylori* in gastric MALT lymphoma) has been treated with antibiotics as the sole initial treatment, resulting in complete remission, although it is uncertain if this completely eradicates neoplastic cells. For localized MALT that is not linked to an infectious stimulus, localized treatment with surgery or radiation has been successful. For disseminated disease, a number of chemotherapeutic agents have been tried, including corticosteroids, alkylating agents (cyclophosphamide and chlorambucil), nucleoside analogues (fludarabine and cladribine), doxorubicin, mitoxantrone, and rituximab, alone or in combination.<sup>45,59-61</sup>

The clinician encountering a patient with dural MALT lymphoma has little guidance on what therapies might be effective in controlling the disease and if, in fact, radiation or cytotoxic treatment is warranted after resection, given the indolent nature of the disease and the potential for CNS toxicity. Radiation has been considered a preferred therapy for localized dural MALT lymphoma because it is a very radiosensitive tumor, and a local modality of treatment is appropriate for a tumor that generally remains localized. However, there is no consensus on dose and schedule, and irradiation of the brain has significant side effects and must be used judiciously, especially in young to middle aged patients who are at increased risk of early dementia. In the case of a dural MALT lymphoma that is not localized, radiation may be less preferred because the patient must receive systemic therapy for the disseminated disease and additional therapy may not be necessary.

An interesting feature of the lymphoma in this case is the unusual tissue distribution. As described above, MALT lymphoma is usually indolent and rarely becomes disseminated.

**Table 1. Dural based lymphoma in the literature by histological type (see text for references).**

Histology	Number of cases	Extra-CNS involvement presentation
MALT/Marginal zone lymphoma	67	4
Diffuse large B-cell lymphoma	9	1
Follicular lymphoma	6	4
CLL/SLL	6	0
Other	5	1
Total	93	10

Dural MALT lymphoma is almost always localized, without systemic involvement. Only 4 cases have been reported with involvement outside of the CNS on presentation;<sup>10,35,62</sup> this case is the fifth. In two of these cases, the only extra-CNS involvement was in the ocular adnexa, with presumed contiguous spread to the globe, the optic nerve, cavernous sinus and the dura.<sup>10</sup>

Dural lymphoma is relatively rare, and most of the reported cases are of dural MALT, the most common type. Here we attempt to summarize the available literature on dural lymphoma. A literature search found 93 cases of dural based lymphomas (Table 1). Of these, 67 (72%) were described as MALT or marginal zone lymphoma,<sup>6,9,10,12,15,16,20-25,28-35,41,44,62-70</sup> 9 were diffuse large B-cell lymphoma,<sup>11,14,18,27,36,37,42,71</sup> and 6 were follicular lymphoma.<sup>3,17,26,38,40,72</sup> Six were described as small lymphocytic lymphoma<sup>13,14,73,74</sup> or B-cell lymphoproliferative disorder of CLL type,<sup>75</sup> but the four of these that were described prior to the establishment of the REAL-WHO criteria may have actually been MALT/Marginal zone lymphoma since immunophenotypical analysis was not performed.<sup>15,74</sup> The remaining 5 cases were described in various ways, including large-cell immunoblastic T-cell lymphoma,<sup>14</sup> monocytoid malignant B-cell lymphoma,<sup>76</sup> centroblastic/centrocytic lymphoma,<sup>19</sup> precursor B-cell lymphoblastic lymphoma,<sup>77</sup> and diffuse large cell non-Hodgkin lymphoma due to Richter's transformation.<sup>39</sup> Of the cases of dural MALT lymphoma/MZL, 63 (94%) were isolated to the CNS, while only 4 (6%) were associated with extra-CNS disease at the time of diagnosis. An additional 5 cases subsequently became disseminated. Of the non-MALT dural lymphomas, 20 (77%) were isolated to the dura, and 6 (23%) were associated with disseminated disease. While these numbers are small, they suggest that dural MALT lymphomas are highly likely to remain as primary dural tumors and not spread to other tissues. Other dural lymphomas appear to have a higher incidence of either arising in the context of a disseminated

**Table 2. Outcomes of dural MALT lymphoma cases in the literature by treatment modality (see text for references).**

Treatment post surgery/biopsy	Number of cases (# with outcomes reported)	Local recurrence	Systemic recurrence
Chemotherapy only	5 (4)	2	0
Radiation only	30 (30)	0	2
Radiation + chemo	18 (17)	1	4
None	5 (4)	0	0
Unknown	9 (2)	0	0

lymphoma, or arising in the dura and subsequently disseminating.

Of the 57 cases of dural MALT lymphoma that have outcomes reported after partial or total surgical resection, only 3 had local recurrence in the dura, and only 6 had systemic recurrence (Table 2). Of those receiving radiation, 1 out of 47 had local recurrence. Of those receiving chemotherapy, 3 out of 21 had local recurrence (17 patients received both chemotherapy and radiation). Of those receiving chemotherapy alone, 2 out of 4 had local recurrence. None of the 4 patients who received neither chemotherapy nor radiation had local or systemic recurrence. Follow-up was highly variable, but average progression free survival for dural MALT lymphoma was greater than 29 months. Of the four patients who had extra-CNS disease at presentation, 1 received chemotherapy only, 1 received radiation only and the other two received radiation and chemotherapy.

These numbers, while small, suggest that local recurrence is uniformly rare after surgical resection. Local recurrence occurred slightly more frequently with chemotherapy alone than with radiation alone, however the significance of this is unclear since there were only 3 instances of local recurrence in the entire series. In two of the three cases of local recurrence, the patients were salvaged with additional chemotherapy with complete response.<sup>10,31</sup> Puri *et al.* propose involved field radiation (IFRT) as an effective means to locally control dural MALT lymphoma, however it is unclear whether even the reduced toxicity of IFRT is justified in a disease that so rarely has local recurrence.<sup>6</sup> In the current case, the patient received systemic chemotherapy for her disseminated disease, which may have also served to control her dural tumor since the dura is not protected by the blood-brain barrier. Based on the published experience, the clinician encountering a dural MALT lymphoma, with or without dissemination, must carefully weigh the need for radiotherapy, but may consider deferring radiotherapy, with close follow-

up. This is especially true in the setting of complete resection or systemic chemotherapy.

## Conclusions

This case of disseminated MALT lymphoma with involvement of the dura demonstrates the importance of maintaining a wide differential diagnosis for dural lesions with imaging characteristics that appear typical for a meningioma. Careful analysis of the imaging studies, primarily MRI, frequently offers additional information, detecting the imaging findings that are suggestive of other disease processes, such as lymphoma. A biopsy and tissue diagnosis should be obtained and complete staging should be done. Dural MALT lymphomas appear to have a very low incidence of local recurrence and therefore irradiation may not be necessary; close surveillance is a valid option for managing this indolent tumor.

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