Coexistence of intracranial marginal zone B-cell lymphoma and meningioma: Case report and review of the literature

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ABSTRACT

Synchronous presentation of multiple primary central nervous system tumors is extremely rare. Meningioma is the most commonly reported tumor in association with other intracranial neoplasms. Review of the literature revealed only 17 cases of meningiomas co-existing with intracranial lymphomas. A 44-year-old woman presented with headache and facial paresis. Magnetic resonance imaging revealed intracranial dural-epidural right frontal mass with tumor extension into the overlying calvarium. A right frontotemporal craniotomy and tumor resection was performed. Histopathologic examination showed a composite meningioma and marginal zone B-cell lymphoma. Radiographic, laboratory, and bone marrow examinations failed to reveal any evidence of systemic disease. Of the 17 lymphoma patients associated with meningioma, two were mucosa-associated lymphoid tissue type but both of these tumors were systemic metastases to intracranial meningiomas. Present report is the first case of a primary intracranial marginal zone B-cell lymphoma interdigitated with a fibroblastic type meningioma.

Key words: Brain synchronous tumors, lymphoma, meningioma

INTRODUCTION

In the absence of predisposing factors, such as phacomatoses or prior radiotherapy synchronous presentation of multiple intracranial tumors is exceedingly rare. Meningiomas are the tumor type more often found in the case of multiple intracranial tumors of different histology.[1] Meningiomas have been reported to occur in conjunction with brain metastasis, gliomas, pituitary adenomas, craniopharyngiomas.[1,2]

Although meningiomas are among the most frequent intracranial tumors, primary central nervous system lymphomas are rare that account for approximately 1% of all intracranial tumors. Most primary intracranial lymphomas are high-grade diffuse large B-cell lymphomas. Low-grade lymphomas are less common, show dural attachment and mimic a menengioma radiographically.[3,4] Immunodeficiency and Epstein–Barr virus has been implicated in the pathogenesis of intracranial lymphoma. The association with other malignancies is extremely rare. A detailed review of the literature revealed 17 cases of meningiomas coexistence with intracranial lymphoma.[2-10]

We report a rare case of the fibroblastic meningioma associated with extranodal marginal zone B-cell lymphoma at the same dural site.

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CASE REPORT

A 44-year-old woman was admitted to the neurosurgical service with a 2–3 years history of headache and 3 months history of right-sided facial paresis. Her medical history was negative. Laboratory investigations and neurological examination were unremarkable. Magnetic resonance imaging (MRI) of the brain revealed intracranial dural-epidural right frontal mass with tumor extension into the overlying calvarium [Figure 1]. She underwent a right frontotemporal craniotomy and resection. Intraoperatively, the tumor was showing erosion entirely through dura and invading adjacent bony structures. The tumor was completely resected.

Histopathologic examination revealed two different types of neoplasms. The vast majority of the resected

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specimen was composed of sheets of small to medium sized, mildly atypical uniform lymphocytes [Figure 2]. Mitoses and apoptosis were rare. Immunohistochemically, the lymphoid cells were strongly and diffuse positive for leukocyte common antigen, CD20, CD79a, and bcl-2 [Figure 3]. Focal and patchy CD5 and CD43 immunoreactivity were observed. The Ki-67 staining was 5–10%. CD10, bcl-6, CD23, CD3, cyclin D1, tdt, CD34 were all negative. These findings were consistent with an extranodal marginal zone B-cell lymphoma. The second and less extensive component of the tumor mass was juxtaposed to the above-described lymphoma and focally infiltrating it [Figure 4]. This lesion was a predominantly fibrotic neoplasm composed of fascicles and sheets of bland cells with oval to spindle shaped nuclei and a moderate amount of eosinophilic cytoplasm. There were scattered psammoma bodies, and the mitotic index was 0–1 per 10 high power fields. There were no atypical or anaplastic features and brain invasion. Immunohistochemical analysis of this component showed strong epithelial membrane antigen, weak S-100 and 5% progesteron positivity [Figure 5]. The diagnosis was consistent with fibroblastic meningioma WHO grade 1. These two tumors were appeared juxtaposed, but rarely infiltration of lymphoma cells into the meningioma was observed.

Postoperatively, the patient did well with complete neurological recovery. Radiographic, laboratory, and bone marrow examinations failed to reveal any evidence of systemic disease.

DISCUSSION

Multiple primary intracranial tumors in the absence of predisposing factors such as phacomatoses or prior radiotherapy are rare. Most of these associations are thought to be coincidental.[1] Some hypotheses have

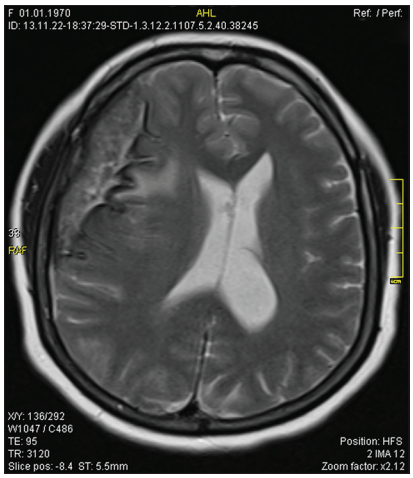

Figure 1: Right frontal dural-epidural mass with tumor extension into the overlying calvarium

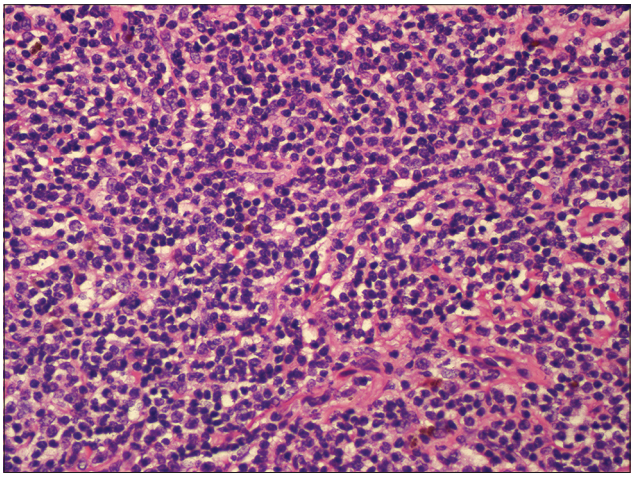

Figure 2: Hematoxylin and Eosin stained sections of the lymphoma component showed sheets of small to medium sized, mildly atypical uniform lymphocytes (H and E, ×200)

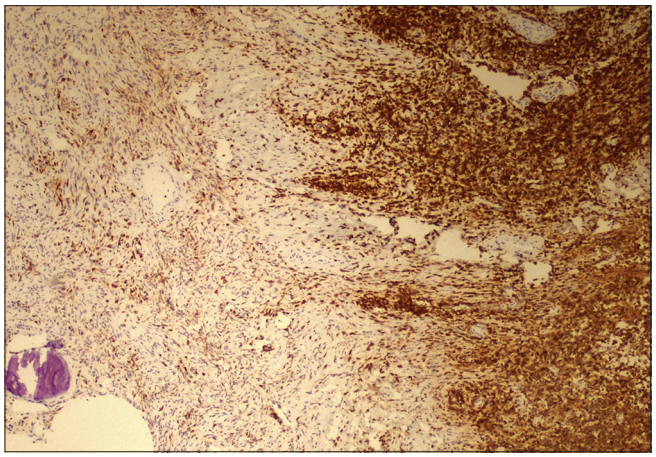

Figure 3: Immunohistochemically diffuse staining with leukocyte common antigen in the lymphoma component

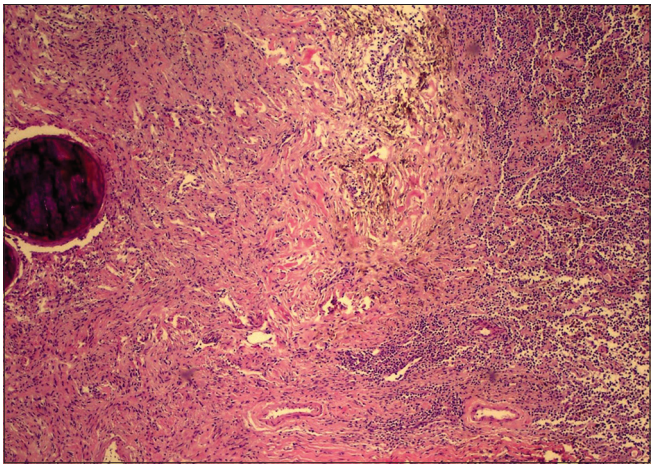

Figure 4: A fibrous meningioma with psammoma body (left side) juxtaposed to the atypical lymphoid proliferation (H and E, ×200)

Table 1: Combined intracranial meningioma and lymphoma reported in English literature

Authors and year	Age	Sex	Location of meningioma	Location of lymphoma	Interval between diagnosis	Lymphoma type	Treatment	Outcome
Kuroiwa *et al.* 1990	62	Female	Posterior fossa	Right temporal	Synchronous	B-cell	Resection, RT	6 months survival
Slowik *et al.* 1990	56	Female	Parasagittal	Left frontal	Autopsy	Non-Hodgkin lymphoma	None	-
Slowik *et al.* 1990	80	Female	Right frontoparietal	Right occipital horn, 4th vent	Autopsy	Non-Hodgkin lymphoma	None	-
Ildan *et al.* 1995	38	Female	Right paraventricular	Left tempoparietal	Synchronous	-	Resection	1-year survival
Buccoliero *et al.* 2004	67	Female	Frontobasal	Frontobasal	2 months	B-cell	Resection, steroid, KT	6 months survival
Maiuri *et al.*	64	Male	Sağ parasagittal	Right posterior parietal	Synchronous	Diffuse large B-cell	Resection, RT	15 months survival
Mori *et al.* 2006	70	Female	Parasagittal	Parasagittal	2 years	Diffuse large B-cell	Resection, RT, KT	NA
George *et al.* 2007	71	Male	Right greater wing of sphenoid	Right sphenoid wing	Synchronous	Diffuse large B-cell	Resection	NA
Riccioni *et al.*	66	Female	Left frontal parasagittal	Frontoparietal	5 years	Follicular	Resection, KT	1-year survival
Colen *et al.*	65	Male	Left temporal	Caverneus sinus	2 years	Anaplastic large cell	Resection, RT, KT	NA
Widdel *et al.*	-	-	Dural based	Dural based	5 years	Marginal zone B-cell	Resection	NA
Gordon *et al.*	65	Female	Frontal	Frontal	Synchronous	B-cell	Resection, KT	18 months survival
Jankowski *et al.*	63	Female	T6-T7 spinal	T6-T7 spinal	Synchronous	B-cell	Resection, KT	1-year survival
Muftah *et al.*	73	Female	Left parietal parasagittal	Left parietal parasagittal	Synchronous	Intravascular large B-cell	Resection	1 month survival
Martin *et al.*	62	Female	Left parieto occipital	Left parieto occipital	14 years	Marginal zone B-cell	Resection, KT	1-year survival
Lapa ve ark 2013	73	Male	Left posterior-parietal	Right frontal	Synchronous	Diffuse large B-cell	NA	NA
Vrotsos ve ark 2013	48	Female	Left mid temporal	Left mid temporal	Synchronous	Burkitt	Resection, KT	NA

NA – Not available; RT – Radiation therapy; KT – Chemotherapy

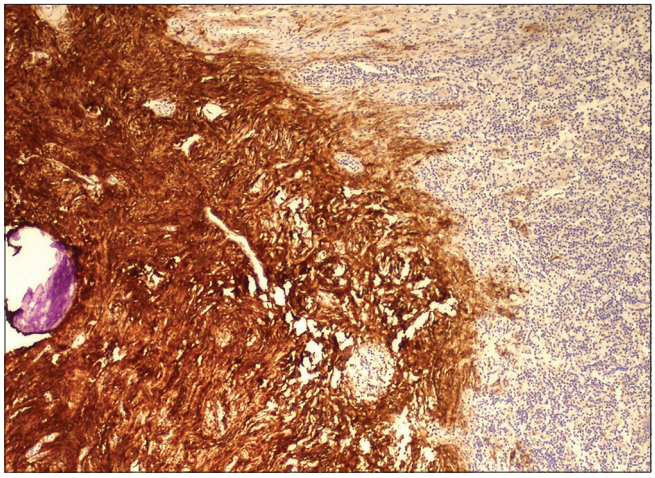

Figure 5: Immunohistochemically diffuse staining with epithelial membrane antigen in the meningioma component

been suggested such as locally acting oncogenic factor or an irritative effect of a tumor inducing the growth of other neoplasm, to explain the presence of different multiple primary intracranial tumors in patients without predisposing factors. Meningioma, one of the most common intracranial tumors, is the most commonly reported tumor in association with other intracranial neoplasms due to its slow growth.[1,2] Brain metastases are the intracranial tumors more often associated with meningiomas.[1]

The combined presence of intracranial meningioma and lymphoma is exceptionally rare with only 17 cases reported in the literature[1-10] [Table 1]. Most of the patients are woman with a mean age of 64.2 (38–80 years). The predominance of these tumors occurring at late decades may support the theory of coincidence and female predominance may suggest the possibility of hormonal pathways. Two of the cases were discovered at autopsy, 10 were simultaneous presentation, four had diagnosis of lymphoma after a meningioma, and one had a history of lymphoma before development of meningioma (minimum 2 months–maximum 5 years delayed).[2-10] Present case is a 44-year-old woman, with contiguous tumors at the same dural site. She had no previous imaging studies to determine whether one lesion was present before the other, but her clinical symptoms were headache and paresis, not systemic lymphoma-related symptoms.

Intracranial lymphoma is a relatively rare disease accounts for approximately 1% of all intracranial tumors.[3,4] Most primary intracranial lymphomas are high-grade diffuse large B-cell lymphomas, but most

primary dural lymphomas are low-grade lymphoma and are in mucosa-associated lyphoid tissue (MALT) type.[4] Of the 17 lymphoma patients associated with meningioma, two were MALT type but both of these tumors were systemic metastases to intracranial meningiomas.[4,5] Present case is a unique primary MALT type lymphoma associated with meningioma. Other histopathologic types of lymphomas combined with meningioma are: Five diffuse B large cell, 4 B-cell non-Hodgkin lymphoma, 1 follicular lymphoma, 1 anaplastic large cell lymphoma, 1 intravascular large B-cell lymphoma, 1 systemic metastasis of Burkitt lymphoma and 1 lymphoma with intermediate features between large B-cell and Burkitt lymphoma.[2,3,6-10]

There have been several theories for the association of lymphoma and meningioma. Meningiomas are highly vascular tumors and create an ideal environment for tumor growth. Paracrine effects of growth factors from the meningioma microenvironment may promote lymphogenic growth. It has also proposed that meningiomas has a relatively low metabolic rate and provide a noncompetetive environment. Furthermore, as meningiomas are common and slow growing, they have an increased risk of concurrence with a second neoplasm.[1,3,4]

The recognition of multiple intracranial tumors might depend on the sensitivity of the diagnostic test used. MRI, due to its sensitivity, can be decisive in recognizing smaller lesions or lesions that are not visible on CT.[6] However, when the lesions are adjacent as in the present case, it may be problematic to achieve a correct preoperative radiologic diagnosis.

We report here, a unique case of a synchronous primary marginal zone B-cell lymphoma associated with meningioma at the same dural site. Although the pathogenesis of the association between primary lymphoma and meningioma remains unclear, hormonal, oncogenic, or paracrine factors produced by meningiomas may play a role in this association. While meningiomas are the most commonly reported tumor combined with other intracranial neoplasms, care must be taken for the adequate sampling of meningiomas.

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