



Intracranial Dural Marginal Zone Lymphoma: A Case Report

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Abstract

We present a case of low grade Marginal Zone Lymphoma (MZL) presenting as a dural mass in a 21 year old woman. The patient presented with symptoms of raised intracranial pressure. Imaging findings were suggestive of meningioma. Histological analysis showed proliferation of CD20 positive small lymphoid cells forming nodules that were reminiscent of lymphoid follicles with attenuated germinal centers. In between the follicles, cells with plasmacytic differentiation were seen, which were CD138 positive and showed kappa light chain restriction by immunohistochemistry. Ki 67 proliferation index was 20-30%. The patient's symptoms were alleviated after the surgery and the patient remains disease free more than two years after resection. This case illustrates that although rare, Extra Nodal MZL should be considered in the differential diagnosis of a dura-based mass that fails to show typical meningioma morphology.

Introduction

Intracranial dural Marginal Zone Lymphoma (DMZL) represents a rare group of low-grade B-cell neoplasms with less than 100 cases reported to date. The patient typically present with a few month history of gradually worsening nausea, vomiting, headache and other symptoms of raised intracranial pressure. Radiology reveals an extra-axial contrast enhancing mass without brain involvement. The clinical presentation and radiological appearance is identical to that of the much more common meningioma [1] or sub-dural hematoma [2,3].

Case Presentation

A 21-year-old female presented with several weeks history of dull intermittent headache that was intractable and excruciating and accompanied by nausea and vomiting at presentation. Past medical history was unremarkable. Neurological examination revealed hyper-reflexia and downward drifting of the right upper extremity. Initial head non-contrast Computed Tomography (CT) showed a lenticular mass along the left frontal convexity with mild edema of the adjacent frontal and parietal cortices. These findings initially raised suspicion of epidural or subdural hematoma. In the absence of any history of trauma, vascular malformation was considered; CT angiogram however failed to show any vascular malformation. Magnetic Resonance Imaging (MRI) was then performed, which showed an extra-axial dural based lesion with contrast enhancement and a prominent dural tail (Figure 1a and b). These findings were considered highly suggestive of meningioma.

Complete resection of the mass was planned after discussion with the patient and her family, Intraoperative biopsy showed a dense aggregate of plasmacytoid cells, raising the suspicion for a lymphoid neoplasm. Complete resection of the lesion was performed and the specimen was sent in 10% formalin for pathological examination. Post-operative MRI showed no evidence of residual neoplasm. The patient recovered uneventfully and was symptom free with no residual neurologic deficit soon after the procedure. After the histology (see pathology section below) confirmed the diagnosis of Marginal Zone Lymphoma, extensive clinical and imaging workup was conducted to search for extra-cranial disease. These studies however failed to find any signs of lymphoma. The patient is being periodically followed up by hematology-oncology and neurosurgery teams and remains free of disease two years after resection. Gross inspection showed a tan white firm lenticular mass encapsulated by dura (Figure 1C and D). Microscopic examination revealed a dense lymphoplasmacytic aggregates separated by fibrous septae, giving a vague nodular arrangement (Figure 2A). Cytologically, three types of cells were

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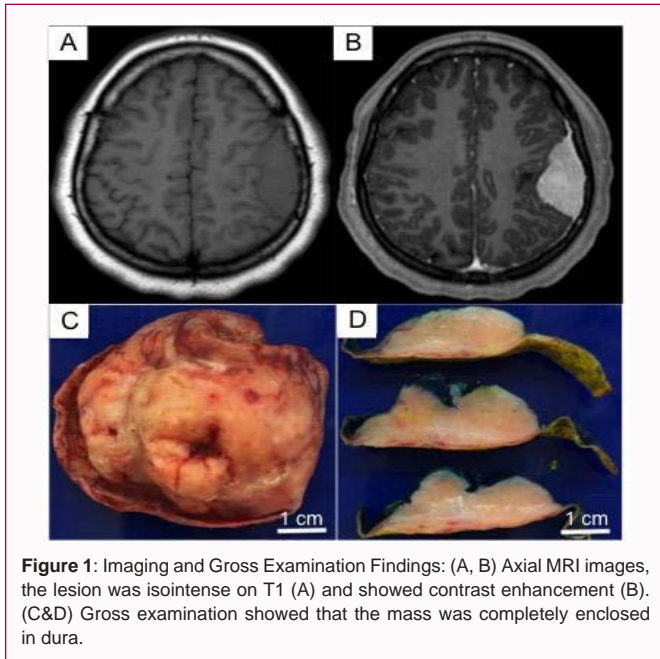


Figure 1: Imaging and Gross Examination Findings: (A, B) Axial MRI images, the lesion was isointense on T1 (A) and showed contrast enhancement (B). (C&D) Gross examination showed that the mass was completely enclosed in dura.

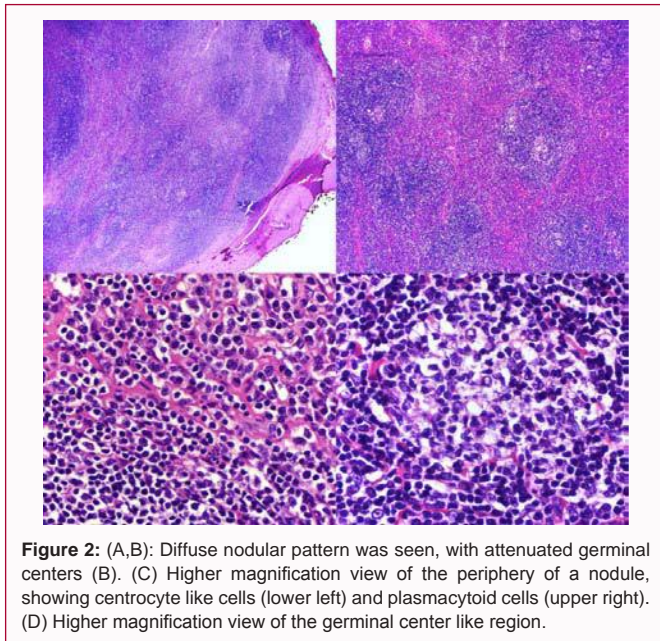


Figure 2: (A,B): Diffuse nodular pattern was seen, with attenuated germinal centers (B). (C) Higher magnification view of the periphery of a nodule, showing centrocyclic cells (lower left) and plasmacytoid cells (upper right). (D) Higher magnification view of the germinal center like region.

present, 1) cells with small centrally placed nuclei and a clear or pale cytoplasm resembling centrocyclics (Figure 2C), 2) Cells with plasmacytic differentiation (Figure 2C), 3) cells of germinal center (Figure 2D). Based on the location and histopathologic findings the differential diagnosis included: a low grade lymphoma such as MALT or Mantle cell lymphoma, plasmacytoma, IgG4 related sclerosing pachymeningitis, and lymphoplasmacyte-rich meningioma. Immunohistochemical (IHC) analysis was performed to rule out these possibilities. The results are summarized in Table 1. In summary, CD20 stain outlined the follicles with attenuated germinal centers, which were positive for CD21 (Figure 3). Between the follicles, the plasmatic cells were positive for CD138. Kappa light chain restriction was demonstrated by both IHC and in situ hybridization (ISH) (Figure 3). Heavy chain studies showed that the plasma cells were weakly positive for IgG, however negative for IgG4. Ki 67 proliferation index was estimated to be 20-30%, consistent with

Table 1: Summary of additional immunohistochemical studies.

| Immunohistochemical maker | Result |
|---------------------------|----------|
| IgG | Positive |
| IgG4 | Negative |
| Kappa | Positive |
| Lambda | Negative |
| IgM | Negative |
| IgA | Negative |
| S100 | Negative |
| EMA | Negative |
| CD56 | Negative |
| CD1a | Negative |
| CD117 | Negative |

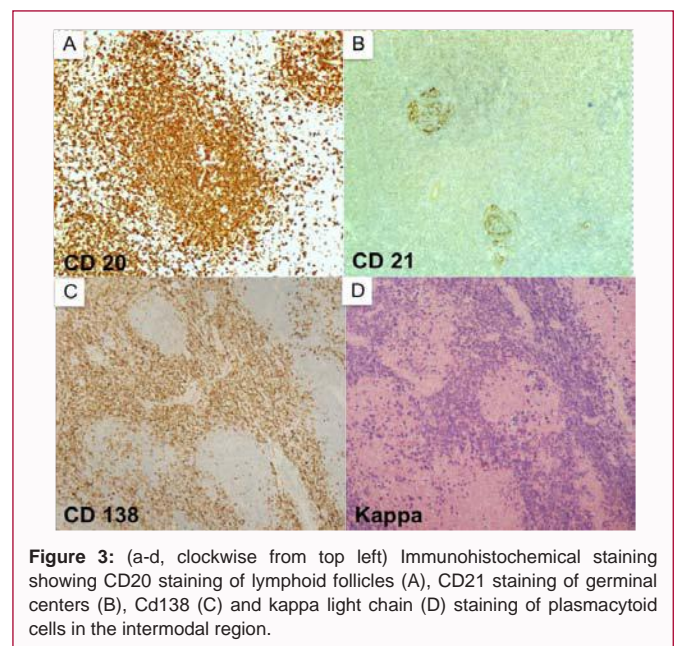


Figure 3: (a-d, clockwise from top left) Immunohistochemical staining showing CD20 staining of lymphoid follicles (A), CD21 staining of germinal centers (B), Cd138 (C) and kappa light chain (D) staining of plasmacytoid cells in the intermodal region.

the indolent feature of this entity. The specimen was received prefixed in formalin; hence flow cytometry was not attempted. These findings are diagnostic for a low grade B cell non-Hodgkin's lymphoma with plasmacytoid differentiation, most consistent with marginal zone lymphoma.

Discussion

Primary CNS lymphoma accounts for approximately 3-4% of primary brain tumors, the majority of these (90%) being large B-cell diffuse lymphomas. These neoplasms usually present as aggressive intra-parenchymal tumors, which may secondarily involve the leptomeninges [4]. Lymphomas primary to the dura mater are much less frequent. Marginal zone lymphoma, designated as 'extranodal marginal zone lymphoma' in the World Health Organization Lymphoma classification, is exceedingly rare [2-8]. This neoplasm was initially discovered in mucosa associated lymphoid tissue (MALT lymphoma) of the stomach, but it can also occur in sites without mucosa such as the orbit, skin, thyroid, thymus, breast, and liver [9].

DMZL most commonly presents in middle aged women with female to male ratio of 4:1 and an average age of presentation of 55 years (range 29 to 70 years) [5,6,10]. This case illustrates that

the diagnosis should also be considered in younger patients. The diagnosis of primary dural MZL requires exclusion of systemic involvement with lymphoma [9]. Diagnostic workup may include: a variety of imaging modalities such as: chest and abdomino-pelvic MRI or CT, Positron Emission Tomography, bone marrow biopsy, ophthalmologic examination (including slit lamp), HIV serology, and lumbar puncture [11].

DMZL typically mimics meningioma in clinical and radiological findings [5]. Typical radiology findings include a single or multiple masses present along the cerebral convexities, falx, tentorium, and sellar/parasellar regions. Rarely, intraventricular and spine masses can occur. The masses are typically lenticular with thickened meninges and frequently show a dural tail. Contrast enhancement with a dural tail has been reported in most cases [11,12]. These features make a radiological distinction from meningioma extremely difficult. However, recently it has been suggested that DMZL shows restricted diffusion behavior on diffusion-weighted images which may help in differentiating these lesions from the typical meningiomas [4].

Although there is no consensus on the treatment of DMZL, surgical resection, chemotherapy and radiation have been tried in combination or stand-alone therapies. Some authors have suggested that radiation or chemotherapy should be reserved for cases in which complete resection is not possible [5,6,11]. Unlike DLBCL, Marginal zone lymphoma has a good prognosis. Most patients attain complete response and have good local disease control. In a series of 8 cases, Iwamoto et al. [11] reported complete response in all cases, however 3 patient developed systemic relapse.

Other rare entities that should be considered as differentials for a dural based mass includes: solitary fibrous tumor/hemangiopericytoma, metastatic tumor, lympho-plasmacyte-rich meningioma, IgG4 related sclerosing pachymeningitis, and Rosai-Dorfman disease. These entities differ from DMZL in histologic and immunohistochemical features. Lymphoplasmacyte-rich meningioma features dense infiltrates of lymphocytes and plasma cells mixed with regions of EMA positive meningotheial cells. Idiopathic hypertrophic pachymeningitis typically shows a polymorphic infiltrate of lymphocytes, plasma cells and multinucleated giant cells. IgG4 related sclerosing disease can be picked up by IgG4 immunohistochemistry. Langerhans cell histiocytosis shows CD1a and Langerin positivity and Rosai-Dorfman disease is usually S100 and CD68 positive and showsemperiopoies is [13]. These features were not seen in the current case, favoring the diagnosis of extra-nodal marginal zone lymphoma, arising from dura (DMZL).

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