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CASE REPORT OF PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA MISDIAGNOSED AS TUMEFACTIVE MULTIPLE SCLEROSIS BY BRAIN BIOPSY

Neurology

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ABSTRACT

Primary central nervous system lymphoma (PCNSL) can be misdiagnosed as tumefactive multiple sclerosis (MS). Both entities share common presentation of focal neurological deficit , radiological enhancement, response to steroid initially and some time relapsing remitting course. Further more , biopsy can be misleading as it may show demyelination features. This is making the diagnosis of PCNSL challenging . So, high index of suspicion and awareness that brain biopsy might show demyelinating changes is needed for earlier diagnosis & management.

KEYWORDS

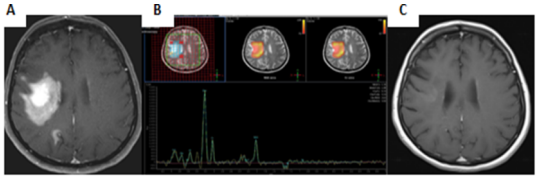
primary central nervous system lymphoma , tumefactive , multiple sclerosis , demyelination

INTRODUCTION

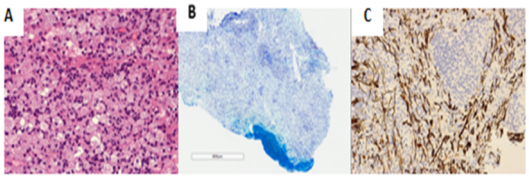
Primary central nervous system lymphoma (PCNSL) can be misdiagnosed as tumefactive multiple sclerosis (MS) in patients whom present with focal neurological deficit , atypical enhancing lesions and have relapsing , remitting course . Clinical deterioration despite disease modifying therapy in MS patients should raise the suspicion of PCNSL.

CASE STUDY

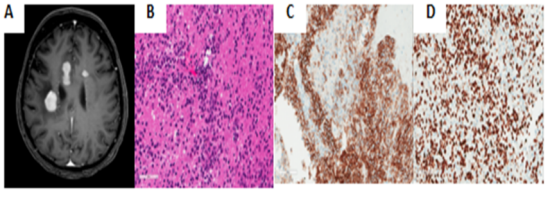
A 48-year-old woman who present with left sided weakness & seizures , found to have multi-focal enhancing white matter lesions that thought to be primary CNS Lymphoma initially. However, brain biopsy showed no evidence of lymphoma with ongoing inflammatory demyelinating process. Despite of good initial response to 3 days only of methylprednison & interferon beta 1A for two years. Later on , patient's condition deteriorated and a repeated brain biopsy was consistent with diffuse large B-cell lymphoma.



A, MRI Brain T1W axial post contrast image in July 2015 demonstrated right frontal lobe enhancing lesion with mild mass effect.
B, Multivoxel MR spectroscopy demonstrated decrease NAA, high Choline and lactate consistent with aggressive lesion.
C, Follow-up MRI in November 2015 demonstrated improvement with decrease in size & enhancement..



A, H & E stained –section showing sheets of foamy macrophages admixed with lymphocytes.
B, Loss of myelin stain confirmed with luxol fast blue stain with residual rim of intact myelin is observed in lower portion.
C, Relative sparing of axons is shown with the neurofilament immunostain confirming demyelination



A, MRI Brain T1W post contrast image in March 2017 demonstrated disease progression with multiple avidly enhancing lesions.
B, Lymphoma cells exhibiting both angiocentricity & diffusely infiltrative patterns.
C. Positive immune reaction for B cell marker CD20.
D. Very high Ki-67 proliferation index.

DISCUSSION:

Literature review showed several cases were misdiagnosed initially as tumefactive MS & turned to to be PCNSL. Both entities share common presentation of focal neurological deficit , radiological enhancement, response to steroid initially and some time relapsing remitting course. Our case is an example of a diagnostic dilemma. Despite of the high suspicion of lymphoma, the initial biopsy was in favor of demyelinating process. Furthermore, the long course of the disease and the response to interferon beta in the begging are atypical for PCNSL. There is at least one case published similar to our patient where patient was treated as demyelinating condition based on brain biopsy but turned to to be diffuse large B-cell lymphoma. One theory that PCNSL may rarely be preceded by “sentinel demyelination,” a pathologic entity characterized by histologically confirmed demyelinating inflammatory brain lesions that mimic multiple sclerosis (MS). Other theory that the initial biopsy was taken from the surface of the lesion rather than the core.

CONCLUSION:

Diagnosis of PCNSL can be very challenging despite of doing brain biopsy. High index of suspicion and awareness that brain biopsy might show demyelinating changes in some cases are clues for early diagnosis

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