Case Report

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Tumefactive Multiple sclerosis

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Introduction

Tumefactive multiple sclerosis (MS) is a rare variant of MS. This form of MS can masquerade as neoplasm or infectious etiology. Understanding of the disease is limited to case report but it is associated with high morbidity and mortality.

Case report

A 44 year old man presented with a 2-month history of progressive right upper extremity weakness, confusion and visual change. Physical exam revealed weakness, hyperreflexia on the right side and right homonymous hemianopia. MRI of the brain showed multiple ring-enhancing lesions located in both cerebral hemispheres. CSF analysis disclosed elevated protein with positive oligoclonal bands and myelin basic protein. Stains and cultures for bacteria and mycobacteria were negative. Serologies including HIV, Toxoplasmosis, and Lyme were all negative. Patient was treated with high-dose IV corticosteroid and clinically improved. One month later, he presented with increasing confusion, aphasia and progressive weakness. Repeat MRI of the brain revealed worsening multiple ring-enhancing lesions with surrounding vasogenic edema in most lesions. High-dose corticosteroid was promptly started. There was also concern about infection, especially brain abscess; hence, intravenous ceftriaxone, vancomycin, and metronidazole were empirically given. Due to uncertainty of diagnosis, first brain biopsy at right frontal lobe lesion yielded non-specific gliosis. Repeat MRI brain showed increasing number of ring-enhancing lesions in both cerebral hemispheres. As a result, a second brain biopsy was performed, which showed an active demyelinating process consistent with multiple sclerosis. Patient experienced severe disability and was discharged to long-term facility with slowly tapered schedule of corticosteroid. He was readmitted several times and eventually family decided hospice care.

Discussion

Multiple sclerosis is diagnosed by demonstrating clinical and/or radiographic evidence of dissemination of disease in time and space[1]. Tumefactive MS is a term used when the clinical presentation and/or MRI findings are indistinguishable from a brain tumor[2]. Not all case of tumefactive MS are fulminant. Marburg variant MS is an acute rare variant of MS which has a rapidly progressive course with frequent, severe relapses leading to death or severe disability within weeks to months[3]. The tumefactive demyelinating lesions are defined as large (>2 cm.) white matter lesions with little mass-like effect or vasogenic edema, and post-gadolinium magnetic resonance imaging (MRI) typically showing an incomplete ring enhancement[2,4]. The clinical and imaging characteristics of these demyelinating lesions may mimic primary and secondary brain tumors, brain abscess, tuberculoma, and other inflammatory disorders e.g. sarcoidosis, primary sjogren's syndrome[5]. As a result, tumefactive MS is frequently misdiagnosed. There are some MRI characteristics that are more suggestive of tumefactive demyelinating lesions than of other etiologies. These include incomplete ring enhancement, mixed T2-weighted iso-and hyperintensity of enhanced regions, absence of a mass effect and absence of cortical involvement[2,6]. Differential diagnosis of rapidly progressive neurological deficit with ring-enhancing lesions include brain abscess, primary brain neoplasm or brain metastasis, acute disseminated encephalomyelitis (ADEM) and tumefactive multiple sclerosis. Careful clinical history, CSF study, serial MRI evaluation and follow-up are usually sufficient to make a diagnosis. Some cases pose considerable diagnostic difficulty owing to clinical and radiographical resemblance to brain tumor, for which biopsy may be warranted. Pathologically, the lesions are characterized by massive macrophage infiltration, acute axonal injury, and necrosis. No specific histological features distinguished specimens derived from patients developing classic multiple sclerosis from those who had tumefactive form[7]. A limited number of cases of Marburg's variant MS have been reported in the literature whereby most patients died within a period of weeks to months. Only two cases survived after one year[7,8]. There is no current standard treatment for this condition. Plasma exchange and Mitoxantrone are reportedly showed some promising options[9,10]

Our patient presented somewhat like a stroke with visual field defect and right hemiparesis which is unusual in MS, but MRI

and CSF exam yielded a diagnosis of probable MS. Because of his abrupt clinical deterioration and impressive worsening of his MRI, concern was raised about possibility of infection or neoplasm. Hence, he received two brain biopsies, the second of which showed active demyelination, confirming the diagnosis of severe tumefactive multiple sclerosis and can be consider as a Marburg variant multiple sclerosis.

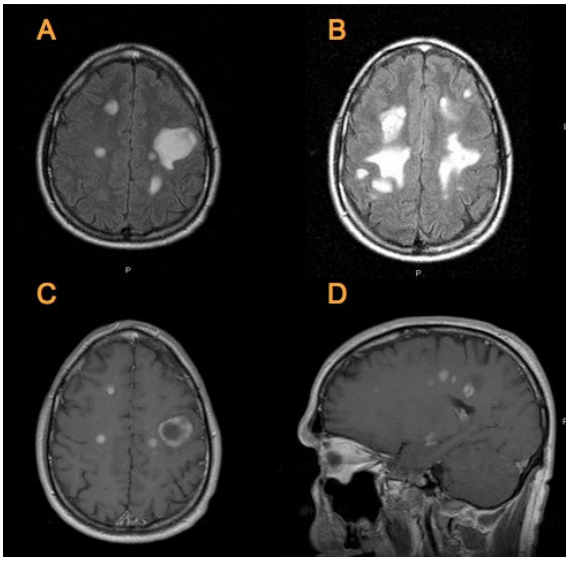

Figure A: FLAIR imaging at first presentation showed lesion in both hemisphere. **Figure B:** FLAIR imaging at one month later showed progression of multiple lesion in both hemisphere. **Figure C:** T1 Post contrast imaging showed intense ring enhancement pattern in almost all lesions with mild edema and minimal mass effect. **Figure D:** Showed lesion view as sagittal section.

Conclusion

Marburg variant multiple sclerosis carries a high morbidity and mortality. This disease notoriously mimics other conditions leading to delay diagnosis and treatment. Absence of definitive diagnosis test apart from brain biopsy makes diagnosis, prognosis and treatment decisions difficult.

Competing Interests
None declared
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