ISSN: 2573-9565

Case Report

Journal of Clinical Review & Case Reports

A Case of Acute Encephalopathy Due to Autoimmune Overload

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Submitted: 08 May 2020; **Accepted:** 15 May 2020; **Published:** 09 Jun 2020

Abstract

Acute disseminated encephalomyelitis, also known as postinfectious encephalomyelitis, is considered an autoimmune disorder that causes inflammation of the brain and spinal cord. It was seen mainly in pediatric population possibly due to vaccination but there have been cases identified in adult [1-9]. Acute disseminated encephalomyelitis can be challenging to diagnose owing to fact that there have been many overlapping symptoms among other demyelinating disorder such multiple sclerosis and Neuromyelitis Optica. In this case report, we will discuss a case about a patient that presented due to acute encephalopathy and was noted to have an atypical MRI of the brain that was not consistent with results of the lumbar puncture [10-12]. Knowledge gained from this case will help bring awareness to the diagnose of acute disseminated encephalomyelitis and how imaging in context with the clinical picture can help us differentiate between the various demyelinating disorders; thereby, giving a better understanding of managing these patients as management can affect prognosis and outcomes.

Introduction

Acute disseminated encephalomyelitis is an immune-mediated, inflammatory, demyelinating disease that predominantly affects the white matter of the brain and spinal cord. The disease manifests as acute encephalopathy associated with multifocal neurological deficits with prodromes such as fever, nausea and vomiting [5]. Clinical presentation of acute disseminated encephalomyelitis may resemble other demyelinating diseases such as a Multiple Sclerosis, Neuromyelitis Optica and Transverse Myelitis.

Case Description

A 38-year-old female with history of lupus and multiple sclerosis presented at an outside hospital due to acute encephalopathy. Patient initially presented to the outside hospital with complaints of headache, severe body aches and numbness that had worsened to the point where she had difficulty walking. Patient was able to protect her airway but after an episode of emesis, patient was intubated for airway protection. Patient was transferred for higher level of care. Upon arrival, patient was noted to be intubated with no sedation on board. She was able to withdraw to noxious stimuli, had a weak cough reflex and a weak gag reflex. Bilateral pupils were noted to be reactive to light but were sluggish with the right pupil not sustaining with light response. MRI of the brain was done which revealed numerous scattered lesions noted throughout the brain and brainstem. MRI of the cervical spine was also done which showed elongated signal abnormalities in the cervical and thoracic cord with at least one enhancing lesion in the cervical cord. Lumbar puncture was done which was positive for HSV, NMO AB at 6.8 with no oligoclonal bands. Patient was initially started with high dose steroids with methylprednisolone 1000mg for presumed multiple sclerosis flare but after lumbar puncture findings were consistent with HSV encephalitis, the steroids were discontinued. Acyclovir 750mg was started; however, after three days of no improvement neurologically, patient was restarted on high dose steroids alongside acyclovir. Patient's neurological status appeared to have improved, as patient was able to open her eyes upon verbal stimuli, follow simple commands and track with her eyes for which she was unable to do initially. As patient neurological status appeared to have plateaued, the decision to start plasmapheresis was made.

Discussion

Acute disseminated encephalomyelitis can be difficult to diagnose and differentiate from other demyelinating diseases such as neuromyelitis Optica and multiple sclerosis due to similar clinical presentation; however, imaging can provide a useful tool in helping to differentiate between the different demyelinating disease.

Although acute disseminated encephalomyelitis has been reported in pediatric population more often, there has been cases that have recorded adult patients. In a significant portion of patient's, between 50-75% of cases, acute disseminated encephalomyelitis is associated with a preceding infection or vaccination [6-9]. One of the early case reports indicated rabies vaccine as a cause of acute disseminated encephalomyelitis [10]. The pathophysiology of acute disseminated encephalomyelitis is not clearly known but it is presumed to be caused by an inflammatory response to a trigger such as an infection [9, 11]. Mainstay of treatment include high dose intravenous glucocorticoids therapy and if patient's do not respond appropriately then plasma exchange, intravenous immunoglobulin or cyclophosphamide can be started [9, 12]. If patients are noted to have concomitant infection, then it is reasonable to start antibiotics with high dose steroids [11, 12].

Neuromyelitis Optica, also known as Devic disease, is also an inflammatory condition that is considered a relapsing syndrome with presentation of optic neuritis or transverse myelitis [13, 14]. The epidemiology can be differing in different literature as it is presumed the rate may be higher as many of the cases were misdiagnosed as multiple sclerosis [14, 15]. Mainstay of treatment in the acute setting is similar to acute disseminated encephalomyelitis with high dose intravenous glucocorticoids; however, long-term treatment include treatment with azathioprine, rituximab, mycophenolate or methotrexate [14, 16].

Multiple sclerosis is a chronic inflammatory disease that affects the central nervous system. Most patients present with a relapsing-remitting episode presentation followed by a secondary progressive episode. Some patients on presentation may present with primary progressive disease instead of the more common clinical subtypes [17, 18]. Common symptoms that patient's present with include sensory loss, muscle weakness, ataxia, double vision and impaired balance [19]. Diagnosis of multiple sclerosis is mainly through clinical presentation with emphasis on the need to show dissemination of lesions in space and time as well as to exclude alternative diagnosis [20, 21]. The Mcdonald criteria discusses how imaging can help further aid and supplement the clinical presentation in diagnosing multiple sclerosis, which can result in earlier diagnosis [21]. Mainstay of treatment for acute exacerbations for multiple sclerosis includes glucocorticoids, similar to other various demyelinating diseases; however long term management can vary depending on the clinical subtypes in multiple sclerosis.

When comparing MRI findings between acute disseminated encephalomyelitis, multiple sclerosis and Neuromyelitis Optica, different areas are noted to be affected. In Neuromyelitis Optica, the areas that appear to be affected are the areas of the brain that have the aquaporin channels that get attacked by the neuromyelitis Optica antibodies which include periventricular areas, medulla oblongata, optic nerve, and corpus collasum [1, 2, 3]. Acute disseminated encephalomyelitis is noted to affect the cortex, basal nuclei, midbrain, and pons [1, 2]. In Multiple sclerosis, area of the brain most notably affected include periventricular, more commonly known as Dawson's finger, optic nerve, cortex, u-fibers, corpus collasum, and pons [2].

Our case report is unique in that based on MRI finding and clinical presentation, patient appeared to have not one demyelinating disorders but in fact appeared to have two autoimmune disorders occurring, making patient's prognosis poor. Patient's presentation of acute encephalopathy with diagnosis of HSV and MRI finding revealing basal ganglia and cortex involvement is more consistent with acute disseminated encephalomyelitis but patient's neuromyelitis Optica antibody being positive on lumbar puncture and having corpus collosum and optic nerve involvement is more consistent with Neuromyelitis Optica. The patient on initial presentation was, due to having a history of multiple sclerosis and not being on any disease modifying therapies to control her multiples sclerosis symptoms, was presumed to have an acute exacerbation of her multiple sclerosis. However, due to her lumbar puncture being negative for oligoclonal bands and her MRI findings not consistently fitting the typical distribution of multiple sclerosis, other etiologies are believed to be playing a part in her disease. Thus, the importance of understanding the characteristics of the various demyelinating disorders is important as it affects prognosis and future management.

There is various infections and vaccinations being implicated as leading to acute disseminated encephalomyelitis, which is not consistently seen in Neuromyelitis Optica with some literature stating that the disease being more sporadic [7-10, 13-16]. Multiple sclerosis etiology and pathogenesis remains unclear, with literature stating the cause is more likely to be multifactorial [22]. Therefore, identifying an infectious process can aid not only in the management of acute disseminated encephalomyelitis but can help to differentiate between the other demyelinating disorders. It has also been stated that interferons which have been utilized in long term management in multiple sclerosis can in reality worsen clinical outcomes in patients with neuromyelitis Optica, which further emphasizes the importance of educating oneself on how acquired demyelinating disorders treatments contrast from one another [17]. With this case report, significance of identifying the diagnosis and treating the patient in a timely manner is crucial for patient's outcome and understanding future outcomes can help with prognosis as it has been noted that about 25 percent of acute disseminated encephalomyelitis can progress to multiple sclerosis [4].

Conclusion

As more information is explored regarding acquired demyelinating disorders, it is important to continue to research the unique characteristics among them to help clinicians in treating patient's more adequately. Furthermore, importance of multidisciplinary care should be incorporated such as consulting neurology and infectious disease specialists to optimize timely care with patients presenting themselves with concern of an autoimmune disease to help with prognosis and future outcomes.

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