Brainstem encephalitis

Bickerstaff brainstem encephalitis is a rare inflammatory disorder of the central nervous system, first described by Edwin Bickerstaff in 1951.

It may also affect the peripheral nervous system, and has featured in common with both Miller Fisher syndrome and Guillain-Barré syndrome.

Rhombencephalitis (RE) is a syndrome of multiple causes and multiple outcomes. Most authors now use the terms "rhombencephalitis" and "brainstem encephalitis" interchangeably even though anatomically they are slightly different. The etiologic categories of RE include infections, autoimmune diseases, and paraneoplastic syndromes (PNS). Listeria is the most common cause of infectious RE. Listeria RE primary occurs in healthy young adults. It usually occurs as a biphasic time course with a flu-like syndrome followed by brainstem dysfunction; 75% of patients have a cerebrospinal fluid (CSF) pleocytosis, and almost 100% have an abnormal brain MRI scan. Positive CSF and blood cultures are the most specific for diagnosis.

Treatment

Treatment primarily is with ampicillin. Enterovirus is probably the second most common infectious cause of RE; however, 95% of cases have occurred in the Asian-Pacific region and there is no specific treatment. Herpes simplex virus (HSV) is the third most common infectious cause of RE, and about 80% of cases are caused by HSV1 and 20% by HSV2. About 50% only had involvement of the brainstem whereas the other 50% also had supratentorial involvement of the temporal and frontal lobes. Mortality with acyclovir treatment was 22% versus those not on acyclovir 75%. Epstein-Barr virus (EBV) and human herpesvirus 6 (HHV6) have caused a few cases. The most common autoimmune etiology is Behçet disease. Over 90% of those with Behçet RE had abnormal MRI scans and 94% had a CSF pleocytosis. Treatment is with corticosteroids and immunosuppressive agents, but only 25% have complete recovery. Paraneoplastic causes are the third category of RE. Brain MRIs are usually normal; there is usually a CSF pleocytosis but the protein is usually normal. Often antineuronal antibodies can be found. Prognosis is poor and treatment is only partially beneficial. Because Listeria and HSV are the most common treatable acute causes of RE, we recommend empiric therapy with ampicillin and acyclovir for all cases after samples have been obtained from CSF and blood for cultures and the polymerase chain reaction (PCR). Antibiotics can be changed based upon MRI, culture results, PCR results, and antibody studies ¹⁾.

Case series

Here, we describe the clinical phenotype of SARS-CoV-2-related CNS disease and evaluate the SARS-CoV-2 antibody index as a tool to differentiate between a direct (viral) and indirect etiology. Out of >4000 hospitalized patients with COVID-19, we included 13 patients with neurological symptoms with suspicion of neuroinflammation. On clinical grounds, eight were classified as having a possible/probable relationship between neurological symptoms and COVID-19. A clinically distinctive

phenotype of brainstem and cerebellar symptoms was seen in 6/8 patients. As we found a positive SARS-CoV-2 antibody index in 3/5 patients, indicating specific intrathecal SARS-CoV-2 IgG production, a direct link with SARS-CoV-2 is likely²⁾

1)

Jubelt B, Mihai C, Li TM, Veerapaneni P. Rhombencephalitis / brainstem encephalitis. Curr Neurol Neurosci Rep. 2011 Dec;11(6):543-52. doi: 10.1007/s11910-011-0228-5. Review. PubMed PMID: 21956758.

Shamier MC, Crijnen YS, Bogers S, IJpelaar JW, de Vries JM, van der Jagt M, Spoor JKH, von der Thüsen JH, Schreurs MWJ, GeurtsvanKessel CH, Titulaer MJ. Brain stem encephalitis is a rare complication of COVID-19. J Neuroimmunol. 2022 Nov 28;374:578007. doi: 10.1016/j.jneuroim.2022.578007. Epub ahead of print. PMID: 36481703.

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