Acute disseminated encephalomyelitis

Acute disseminated encephalomyelitis (ADEM), or acute demyelinating encephalomyelitis, is a rare autoimmune disease marked by a sudden, widespread attack of inflammation in the brain and spinal cord. As well as causing the brain and spinal cord to become inflamed, ADEM also attacks the nerves of the central nervous system and damages their myelin insulation, which, as a result, destroys the white matter. It is often triggered after the patient has received a viral infection or, perhaps exceedingly rarely specific non-routine vaccinations.

Case reports

A 17-year-old girl underwent emergency bifrontal decompressive craniectomy for severely raised intracranial pressure with brainstem compression, having developed acute disseminated encephalomyelitis (ADEM) following Epstein-Barr virus infection.

Bourke et al discussed the current evidence for craniectomy in both ADEM and infective encephalitis and propose an approach to management [1].

Ceronie et al., described a novel presentation following urological surgery. Using illustrative features from our case study, we describe some of the clinical features, aetiologies, diagnostic uncertainties and pathogenic mechanisms of the disease. A 69 year old gentleman underwent transurethral resection of the prostate. He then developed confusion, unsteadiness, behavioural disturbance and left-sided hemiparesis. On admission he was febrile with left hemiplegia and ataxia. Neuroimaging showed multifocal, posterior-predominant semi-confluent lesions. Autoimmune serology and virology were negative. Cerebrospinal fluid revealed mildly elevated protein. Brain biopsy confirmed a diagnosis of ADEM. ADEM is a predominantly a childhood disorder and rare in older adults. It is precipitated by vaccinations, viral, bacterial or parasitic infections. It is rarely described after surgical intervention. Differential diagnosis is wide and includes multiple sclerosis (MS), encephalitides and encephalopathies. Treatment is with corticosteroids, plasma exchange, intravenous immunoglobulin or cyclophosphamide. Up to a quarter will experience recurrence and 10% progress to MS. Further study is needed to determine its pathogenic and immunological characteristics [2].

A patient who died of a fatal meningoencephalitis after removal of a third ventricle colloid cyst. Postoperative clinical and iconographic evolution let us think about acute disseminated encephalomyelitis probably due to cerebrospinal fluid contamination by inflammatory proteins contained in the colloid cyst. This case raises the question of the possibility of colloid cyst content spraying while using an ultrasonic aspiration device [3].

References

1)
Bourke D, Woon K. Craniectomy for acute disseminated encephalomyelitis. Pract Neurol. 2020 Sep

25:practneurol-2020-002705. doi: 10.1136/practneurol-2020-002705. Epub ahead of print. PMID: 32978272.

2)

Ceronie B, Cockerell OC. Acute disseminated encephalomyelitis in an older adult following prostate resection. eNeurologicalSci. 2018 Nov 22;14:40-42. doi: 10.1016/j.ensci.2018.11.006. eCollection 2019 Mar. PubMed PMID: 30619950; PubMed Central PMCID: PMC6304343.

3)

Desse N, Sellier A, Bernard C, Dagain A. Fatal acute disseminated encephalomyelitis (ADEM) after third ventricle colloid cyst resection with ultrasonic aspirator during neuroendoscopic procedure. Acta Neurochir (Wien). 2018 Sep;160(9):1789-1792. doi: 10.1007/s00701-018-3631-8. Epub 2018 Jul 29. PMID: 30058026.