



BÖLÜM 10

AKUT DISSEMINÉ ENSEFALOMİYELİT (ADEM)

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GİRİŞ

Akut dissemine ensefalomyelit (ADEM), nadir görülen, daha çok subkortikal yerleşimli, enflamatuar, demiyelinizan bir hastaliktır. Çocuk ve genç erişkinlerde daha sık görülmektedir. Öncesinde aşılama ve enfeksiyon sık izlenmektedir.Çoğu zaman klinik seyri monofaziktir; bununla birlikte, bazen multifazik ADEM meydana gelir ve bu hastalığı multipl sklerozdan (MS) ayırt etmekte güçlük çekilebilir. Hastalık en sık olarak tanımlanamayan viral veya bazen bakteriyel enfeksiyoz bir hastalığı takip edebilir (1). ADEM tüm dünyada görülmektedir. ADEM'in aksine çocuklarda MS tanısı daha seyrek konulur. ADEM'in прогнозu genellikle iyidir, ancak bazen ciddi nörolojik sekeller görülür. Bu bölümde ADEM'in etiyolojisi, klinik, laboratuvar, radyolojik bulguları, tedavi ve прогнозu tartışılmaktadır.

PATOGENEZ

ADEM'in patogenezi, hayvanların miyelin protein ürünleri ile bağışıklaştırılmışıyla indüklenen akut demiyelinizan bir hastalık olan hayvan modeli deneysel otoimmün ensefalomyelit (EAE) ile benzerliğe sahiptir (2).

EAE'ye benzer şekilde, insanlarda, geçmişte tavşan veya keçi merkezi sinir sistemi (MSS) dokusu ile kontamine olmuş canlı, zayıflatılmış bir aşısı olan Semp le kuduz aşısı ile bağışıklamadan sonra ADEM vakaları gözlemlenmiştir (3).

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de en azından kısmi olarak, genellikle tam çözümürlük gösterir. ADEM tedavisi, IVIG ve plazma değişimi olarak tanımlanmış olmasına rağmen, çoğunlukla üç ila beş günlük yüksek doz intravenöz glukokortikoid ile tedavi edilir.

KAYNAKLAR

1. Stonehouse B, Gupte G, Wassmer E, et al. Acute disseminated encephalomyelitis: recognition in the hands of general paediatricians. *Arch Dis Child* 2003; 88: 122- 4.
2. Rivers TM, Sprunt DH, Berry GP. Observations on attempts to produce acute disseminated encephalomyelitis in monkeys. *J Exp Med*. 1933;58:39–53.
3. Hemachudha T, Griffin DE, Giffels JJ. Myelin basic protein as an encephalitogen in encephalomyelitis and polyneuritis following rabies vaccination. *N Engl J Med*. 1987;316:369–374.
4. Fujinami RS, Oldstone MB. Amino acid homology between the encephalitogenic site of myelin basic protein and virus: mechanism for autoimmunity. *Science*. 1985;230:1043–1045.
5. Jorens PG, VanderBorgh A., Ceulemans B. Encephalomyelitis-associated antimyelin autoreactivity induced by streptococcal exotoxins. *Neurology*. 2000;54:1433–1441.
6. Garg RK. Acute disseminated encephalomyelitis. *Postgrad Med J*. 2003;79:11–17.)
7. Behan PO, Kies MW, Lisak RP. Immunologic mechanisms in experimental encephalomyelitis in nonhuman primates. *Arch Neurol*. 1973;29:4–9.
8. Levine S. Hyperacute, neutrophilic, and localized forms of experimental allergic encephalomyelitis: a review. *Acta Neuropathol*. 1974;28:179–189.
9. Cole J, Evans E, Mwangi M, et al. Acute disseminated encephalomyelitis in children: an updated review based on current diagnostic criteria. *Pediatr Neurol*. 2019;100:26–34.
10. Hynson JL, Kornberg AJ, Coleman LT. Clinical and neuroradiologic features of acute disseminated encephalomyelitis in children. *Neurology*. 2001;56:1308–1312.
11. Dale RC, de Sousa C, Chong WK, et al. Acute disseminated encephalomyelitis, multiphasic disseminated encephalomyelitis and multiple sclerosis in children. *Brain*. 2000;12:2407–2422.
12. López-Chiriboga AS, Majed M, Fryer J, et al. Association of MOG-IgG serostatus with relapse after acute disseminated encephalomyelitis and proposed diagnostic criteria for MOG-IgG-associated disorders. *JAMA Neurol*. 2018; 75 :1355-1363.
13. Lotze ET, Chadwick DJ. Acute disseminated encephalomyelitis (ADEM) in children: pathogenesis, clinical features, and diagnosis. 2020.
14. Yıldız O, Pul R, Raab P, et al. Acute hemorrhagic leukoencephalitis (Weston-Hurst syndrome) in a patient with relapse-remitting multiple sclerosis. *J Neuroinflammation*. 2015; 17 :175.

15. Schwarz S, Mohr A, Knauth M. Acute disseminated encephalomyelitis: a follow-up study of 40 adult patients. *Neurology*. 2001;56:1313–1318.
16. Menge T, Kieseier BC, Nessler S. Acute disseminated encephalomyelitis: an acute hit against the brain. *Curr Opin Neurol*. 2007;20:247–254.
17. Lim KE., Hsu YY, Hsu WC. Multiple complete ring-shaped enhanced MRI lesions in acute disseminated encephalomyelitis. *Clin Imaging*. 2003;27:281–284.
18. Tenembaum S, Chamois N, Fejerman N. Acute disseminated encephalomyelitis: a long-term follow-up study of 84 pediatric patients. *Neurology*. 2002;59:1224–1231.
19. Caldemeyer KS., Smith RR., Harris TM. MRI in acute disseminated encephalomyelitis. *Neuroradiology*. 1994;36:216–220.
20. Hung PC, Wang HS, Chou ML, et al. Acute disseminated encephalomyelitis in children: a single institution experience of 28 patients *Neuropediatrics*, 43 (2012), pp. 64–71) *Dev Med Child Neurol*, 60 (2018), pp. 1123–1131)
21. Fridinger SE, Gulay A. “Defining encephalopathy in acute disseminated encephalomyelitis.” *Journal of child neurology* 29.6 (2014): 751–755.
22. Suppiej A, Vittorini R, Fontanini M, ET et al. Acute disseminated encephalomyelitis in children: focus on relapsing patients. *Pediatric neurology*, 39(1), 12–17.
23. Rossor T, Benetou C, Wright S, et al. Early predictors of epilepsy and subsequent relapse in children with acute disseminated encephalomyelitis. *Multiple Sclerosis Journal*, 26(3), 333–342.
24. Lennon VA, Wingerchuk DM, Kryzer TJ. A serum autoantibody marker of neuromyelitis optica: distinction from multiple sclerosis. *Lancet*. 2004;364:2106–2112.
25. Cusmai R., Bertini E, Di Capua M. Bilateral, reversible, selective thalamic involvement demonstrated by brain MR and acute severe neurological dysfunction with favorable outcome. *Neuropediatrics*. 1994;25:44–47.
26. Hartfield DS, Loewy JA, Yager JY. Transient thalamic changes on MRI in a child with hypernatremia. *Pediatr Neurol*. 1999;20:60–62.
27. Sahlas DJ, Miller SP, Guerin M. Treatment of acute disseminated encephalomyelitis with intravenous immunoglobulin. *Neurology*. 2000;54:1370–1372.
28. Keegan M, Pineda AA., McClelland RL. Plasma exchange for severe attacks of CNS demyelination: predictors of response. *Neurology*. 2002;58:143–146.
29. Borlot F, da Paz JA., Casella EB. Acute hemorrhagic encephalomyelitis in childhood: case report and literature review. *J Pediatr Neurosci*. 2011;6:48–51.
30. Idrissova ZhR., Boldyreva MN., Dekonenko EP. Acute disseminated encephalomyelitis in children: clinical features and HLA-DR linkage. *Eur J Neurol*. 2003;10:537–546.
31. Murthy SN., Faden HS., Cohen ME. Acute disseminated encephalomyelitis in children. *Pediatrics*. 2002;110:e21) (Kimura S., Nezu A., Ohtsuki N. Serial magnetic resonance imaging in children with postinfectious encephalitis. *Brain Dev*. 1996;18:461–465.

▲ Demyelinizan Hastalıklar

32. Hahn CD, Miles BS, MacGregor DL. Neurocognitive outcome after acute disseminated encephalomyelitis. *Pediatr Neurol.* 2003;29:117–123.
33. Jacobs RK., Anderson VA, Neale JL. Neuropsychological outcome after acute disseminated encephalomyelitis: impact of age at illness onset. *Pediatr Neurol.* 2004;31:191–197.