Osmangazi Tıp Dergisi
Osmangazi Journal of Medicine 2021;43(4):419-423

Case Report / Olgu Sunumu

A Pediatric Patient with Peripheral Facial Nerve Palsy Due to Acute Disseminated Encephalomyelitis

Akut Dissemine Ensefalomiyelite Bağlı Periferik Fasiyal Sinir Felci Gelişen Bir Çocuk Hasta

[1]Coskun Yarar, [2]Sumeyye Emel Yel, [1]Emre Kaplan, [1]Kursat Bora Carman

[1]Department of Pediatric Neurology, Eskisehir Osmangazi University Faculty of Medicine, Eskisehir, Turkey

[2]Department of Pediatric, Eskisehir Osmangazi University Faculty of Medicine, Eskisehir, Turkey

Correspondence:
Coskun YARAR
Department of Pediatric Neurology, Eskisehir Osmangazi University Faculty of Medicine,
Eskisehir, Turkey
e-mail: coskunyarar@hotmail.com

Abstract

Acute disseminated encephalomyelitis (ADEM) is considered a monophasic acute demyelinating disorder of the central nervous system characterized by diffuse neurologic signs and symptoms coupled with evidence of multifocal lesions of demyelination on neuroimaging. The common presenting symptoms are lethargy, fever, vomiting, headache, meningeal signs, and seizures. Neurological manifestations included altered sensorium, multiple cranial nerve involvement, quadriplegia and paraplegia, dystonia and choreiform movements, ataxia, nystagmus, bladder involvement, speech defect, and double vision. Peripheral facial nerve palsy rarely accompanies. We describe the case of a 7-year-old girl who developed acute disseminated encephalomyelitis with right peripheral facial paralysis. She was admitted to the hospital with visual impairment, depressed mood, gait disturbance, and deterioration of facial expressions. She had horizontal and vertical nystagmus, right peripheral facial paralysis, ataxic gait, and hyperactive deep tendon reflexes. The cerebrospinal fluid (CSF) examination was normal. Cerebral magnetic resonance imaging (MRI) showed multiple white matter lesions. We observed a significant improvement in clinical findings with high dose methylprednisolone pulse therapy.

Keywords: Acute disseminated encephalomyelitis, facial nerve palsy, child, high-dose methylprednisolone pulse therapy

Özet

Akut dissemine ensefalomiyelit (ADEM), nörogörüntülemede multifokal demiyelinasyon lezyonlarının kanıtıyla birlikte yaygın nörolojik belirti ve semptomlarla karakterize, santral sinir sisteminin monofazik akut demiyelinizan bir bozukluğu olarak kabul edilir. Yaygın görülen semptomlar letarji, ateş, kusma, baş ağrısı, meningeal belirtiler ve nöbetlerdir. Nörolojik belirtiler arasında duyusal değişiklikler, çoklu kraniyal sinir tutulumu, kuadripleji ve parapleji, distoni ve koreiform hareketler, ataksi, nistagmus, mesane tutulumu, konuşma bozukluğu ve çift görme vardır. Periferik fasiyal sinir felci nadiren eşlik eder. Akut dissemine ensefalomiyelit ile sağ periferik fasiyal sinir felci gelişen 7 yaşındaki bir kız çocuğu olgusunu sunuyoruz. Hastaneye görme azlığı, depresif ruh hali, yürüme bozukluğu ve yüz ifadelerinde bozulma yakınmaları ile başvurdu. Horizontal ve vertikal nistagmus, sağ periferik yüz felci, ataksik yürüyüş ve derin tendon reflekslerinde artış vardı. Beyin omurilik sıvısı (BOS) incelemesi normaldi. Serebral manyetik rezonans görüntülemede (MRG) çoklu beyaz cevher lezyonları görüldü. Yüksek doz metilprednizolon pulse tedavisi ile klinik bulgularda belirgin bir iyileşme gözlemledik.

Anahtar Kelimeler: Akut dissemine ensefalomiyelit, fasiyal sinir felci, çocuk, yüksek doz metilprednizolon pulse tedavisi

Received 24.08.2020 Accepted 14.12.2020 Online published 15.12.2020

Cite this article as:
Yarar C, Yel SE, Kaplan E, Carman KB, Akut Dissemine Ensefalomiyelite Bağlı Periferik Fasiyal Sinir Felci Gelişen Bir Çocuk Hasta, Osmangazi Journal of Medicine, 2021 43(5) 547-551 Doi: 10.20515/otd.784408

1. Introduction

Acute disseminated encephalomyelitis (ADEM) is an immune-mediated demyelinating disorder of the central nervous system that can follow infections or immunizations (1-3). ADEM is usually a monophasic disease with acute onset characterized by multiple foci of central nervous system damage, predominantly in the cerebral and cerebellar white matter. However, basal ganglia and grey matter may also be involved (3-5).

After the prodromal phase of 1-4 weeks, clinical signs, including altered consciousness and multifocal neurological findings, are appeared (3,6). The common presenting symptoms were lethargy, fever, vomiting, headache, meningeal signs, seizures (1). Neurological manifestations included altered sensorium, multiple cranial nerve involvement, quadriplegia, paraplegia, dystonia, choreiform movements, ataxia, nystagmus, bladder involvement, speech defect, and double vision. The facial nerve is more common than other cranial nerves involved. However, the association with ADEM has rarely been reported. Both peripheric and central facial palsy can be seen (4,7,8).

ADEM diagnosis is based on the acute onset of neurologic signs and symptoms, along with evidence of multifocal lesions of demyelination on neuroimaging (7,9).

In this case report, we describe a patient who presented with peripheral facial palsy and diagnosed with ADEM.

2. Case

A 7-year-old girl without a known illness was brought to our hospital with visual impairment complaints, gait disturbance, depressed mood, and unilateral deterioration of facial expressions. One week before admission, she had an upper respiratory tract infection and received oral amoxicillin-clavulanate treatment. Two days before admission, her complaints showed up. On admission, she was in good general condition and stable vital findings. She had on physical examination oropharyngeal hyperemia, 2/6 systolic murmur, hyperactive deep tendon reflexes, horizontal nystagmus that spontaneously hit the right side, vertical nystagmus that hits up the examination, right peripheral facial paralysis, ataxic gait. The fundoscopic examination was normal.

Complete blood count, electrolytes, liver function tests, renal function tests, and hemostasis tests were within normal limits. Erythrocyte sedimentation rate was 7 mm/hr, C reactive protein was negative. On serologic tests, Herpes simplex virus type 1 and 2, Epstein Barr virus, Parvovirus, Measles, Chickenpox, Mumps, Hepatitis B virus, Hepatitis C virus, Human immunodeficiency virus, Toxoplasma, Rubella, Cytomegalovirus, and Borrelia burgdorferi antibodies were negative. Immunological tests for autoimmune diseases were not studied.

The cerebrospinal fluid examination was normal [normal opening pressure, showed no cells, normal protein (24,8 mg/dl), and normal glucose (66 mg/dl)], the oligoclonal band of IgG was negative, IgG index was increased (0.84). Anti-MOG antibody could not be studied. The cerebrospinal fluid culture was sterile, and the viral meningitis panel was negative.

Cranial computerized tomography was normal. Cerebral MRI showed multiple lesions in the bilateral cerebellar hemispheres, brain stem, middle cerebellar peduncles, basal ganglia, and corpus callosum (Figure 1,2 and 3).

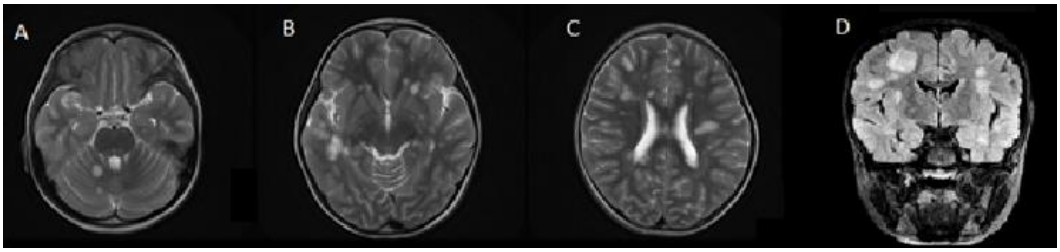

Figure 1. A, B, C, Axial T2-weighted image of brain magnetic resonance imaging (MRI), showing multiple hyperintensities. D, T2-FLAIR MRI shows multiple round lesions, seen as an increased signal.

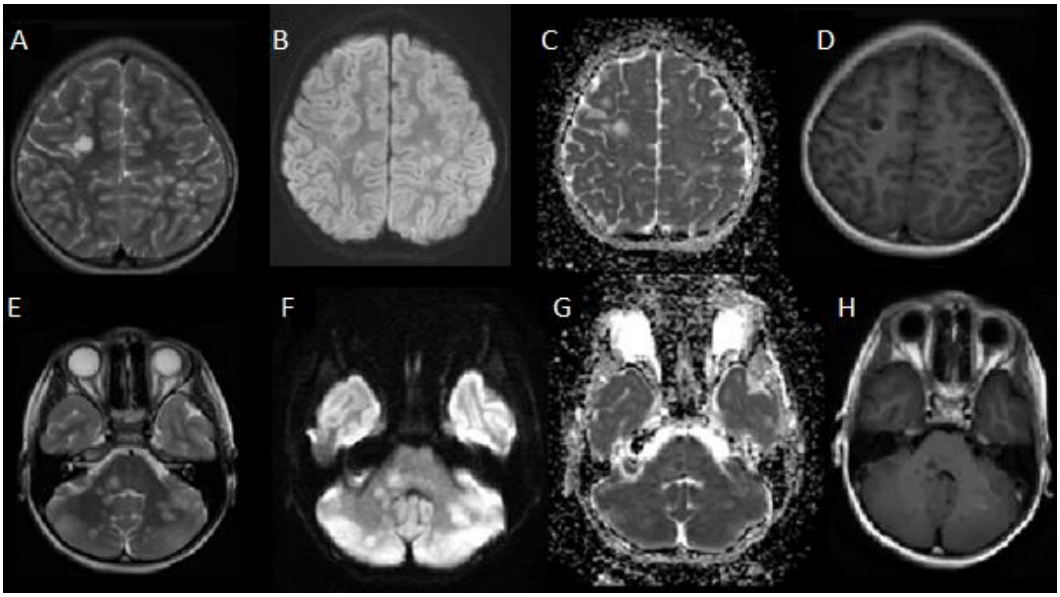

Figure 2. Distribution of lesions on MRI A, Axial T2-weighted image of supratentorial area. B, Axial Diffusion-weighted image of supratentorial area. C, Apparent Diffusion Coefficient image of supratentorial area. D, T2-FLAIR post-contrast image of supratentorial area. E, Axial T2-weighted image of posterior fossa section. F, Axial Diffusion-weighted image of posterior fossa section. G, Apparent Diffusion Coefficient image of posterior fossa section. H, T2-FLAIR post-contrast image of posterior fossa section.

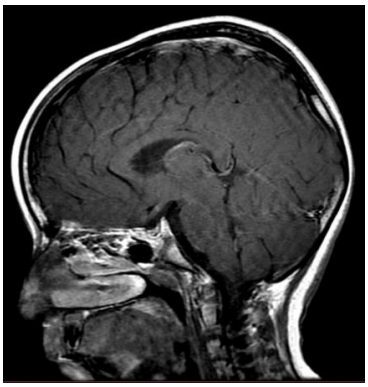

Figure 3. Sagittal T1-weighted image of post-contrast corpus callosum.

She received intravenous high-dose methylprednisolone pulse therapy (30 mg/kg/day for three days and following 20 mg/kg/day for two days) and continued oral prednisolone (was started at 2 mg/kg/day and gradually stopped in a month). She recovered

for visual impairment, ataxic gait, depressed mood, and peripheral facial paralysis findings on follow up.

3. Discussion

ADEM is a rare disorder, which usually occurs following a viral illness. It is an immune-mediated disorder producing multifocal inflammation and subsequent demyelination lesions within the CNS. The incidence rate is about 0.4-0.8 in 100,000. It can occur at any age but more common in children with an average age of 5–7 years. Patients present with non-specific symptoms and neurological symptoms such as confusion, altered level of consciousness, or irritability (10).

ADEM typically presents as a monophasic illness. Characteristic clinical features include sudden onset multifocal neurologic disturbances such as visual field defects, aphasia, motor, and sensory deficits, ataxia, movement disorders, a depressed level of consciousness, focal or generalized seizures, and psychosis (11). Besides, the clinical features of ADEM include multiple cranial nerve involvement. The facial nerve is more common than other cranial nerves involved. In the literature, cases with facial paralysis have rarely been reported. However, the association between facial paralysis and ADEM was not explained clearly (12-16).

In our case, facial palsy was not the only finding like the other ADEM cases. The presence of encephalopathy is an important finding for the diagnosis of ADEM. Our case presented with peripheral facial palsy, depressed mood, nystagmus, and ataxic gait. These findings indicate the presence of mild encephalopathy in the patient. But, in the inspection, the first thing that attracted attention was facial paralysis. Ataxia, hemiparesis, hemiplegia, seizures, and status epilepticus were accompanying findings reported in the literature. Al-Hanshanı et al. (12) reported an 11-year-old girl who had a progressive frontal headache and left eye ptosis, and left facial nerve palsy. Thomas et al. (13) reported a 4-year-old girl who presented with an afebrile partial status epilepticus and right-sided lower motor neuron facial nerve palsy. Tripathy et al. (14) reported a 36-year-old male with behavioral abnormalities, hemispatial neglect of left upper limb, paralysis of the left facial nerve, left arm, and right leg. Patra et al. (15) reported a 10-year-old child who had left upper motor neuron facial palsy with left hemiplegia, paralyzed right lower limb, exaggerated deep tendon reflexes, and bilateral extensor plantar responses. Kaymakamzade et al. (16) reported a 25-year-old male with bilateral central facial palsy and severe quadriparesis.

The cerebrospinal fluid examination's classic features include an increased opening pressure, lymphocytic pleocytosis, normal glucose with raised protein content, and gamma-globulins. Oligoclonal bands are only found in 10% of the ADEM cases and are always a temporary finding (17-19). In our case, CSF analysis was normal, and the oligoclonal band of IgG was negative. The IgG index was found to be elevated, indicating the increase in IgG synthesis in the central nervous system. Intrathecal IgG synthesis can be found to be increased in these patients (20).

Usually, the MRI scans are positive for focal or multifocal lesions, primarily located in the supratentorial or infratentorial white matter and/or in the gray matter of the basal ganglia and/or the thalamus. These lesions are usually 1-2 cm in size and best demonstrated on T2-weighted and FLAIR sequences (17,21,22). Bilateral cerebellar hemispheres, brain stem, middle cerebellar peduncles, basal ganglia, corpus callosum, and cerebral hemispheres in both sizes of confluent T2-weighted hyperintense ring, open ring, and solid enhancement and some non-enhancement lesions are available in our patient’s cerebral MRI.

After a rapid diagnosis, high-dose methylprednisolone pulse therapy was started at the 12th hour of the patient's admission. The patient responded quickly to the treatment and was discharged with complete clinical improvement.

We thought multiple sclerosis (MS), Lyme disease, and tumoral lesions in the differential diagnosis. We considered ADEM diagnosis with the first occurrence of the findings, preadolescent age, history of infection a week ago, accompanying ataxia, multiple findings, encephalopathy, and the oligoclonal band's negativity of IgG. However, due to the patient's high IgG index, we will continue to follow up on possible recurrent attacks, considering a case with MS or other demyelinating disorders of central nervous system who had his first attack as ADEM.

4. Conclusion

Acute disseminated encephalomyelitis may be associated with facial paralysis, and the response of our patient to high-dose methylprednisolone pulse therapy is excellent. In cases with facial paralysis, and encephalopathy ADEM should be considered in the differential diagnosis.

REFERENCES

1. Jayne MN. Demyelinating disorders of the central nervous system. In: Kliegman RM, Stanton BF, St Geme JW, Schor NF, Behrman RE editors. *Nelson Textbook of Pediatrics.* 20th ed. Philadelphia: Elsevier; 2016;2919–2925.
2. Kaya A, Acikgoz M, Ustyol L, et al. A case of acute disseminated encephalomyelitis mimicking leukodystrophy. *Kurume Med J.* 2010;57:85-9.
3. Madan S, Aneja S, Tripathi RP, Batra A, Seth A, Taluja V. Acute disseminated encephalomyelitis--a case series. *Indian Pediatr.* 2005;42:367-71.
4. Garg RK. Acute disseminated encephalomyelitis. *Postgrad Med J.* 2003;79:11-7.
5. Kinomoto K, Okamoto Y, Yuchi Y, Kuriyama M. Acute encephalomyelitis associated with acute viral hepatitis type B. *Intern Med.* 2009;48:241-43.
6. Sidhu J, Maheshwari A, Gupta R, Devgan V. Acute disseminated encephalomyelitis after Plasmodium vivax infection: case report and review of literature. *Pediatr Rep*. 2015;7:5859.
7. Jayakrishnan MP, Krishnakumar P. Clinical profile of acute disseminated encephalomyelitis in children. *J Pediatr Neurosci.* 2010;5:111–14.
8. Ashrafi MR, Amirkashani D, Hirbod-Mobarakeh A, et al. Acute disseminated encephalomyelitis mimicking acute meningoencephalitis. *Acta Clin Croat*. 2013;52:523-28.
9. Murthy SN, Faden HS, Cohen ME, Bakshi R. Acute disseminated encephalomyelitis in children. *Pediatrics.* 2002;110(2Pt1):e21.
10. Zeb Q, Alegria A. Acute disseminated encephalomyelitis (ADEM) following a H3N3 parainfluenza virus infection in a pregnant asthmatic woman with respiratory failure. *BMJ Case Reports.* 2014;2014:bcr2013201072.
11. Kumar P, Kumar P, Sabharwal RK. Acute disseminated encephalomyelitis: case report and brief review. *J Family Med Prim Care*. 2014;3:443-45.
12. Al-Hanshani AA, Otaif MY, Shati AA, Alsuheel AM, Huneif MA. Acute disseminated encephalomyelitis presenting with ptosis and facial palsy. *Med J Cairo Univ*. 2014;82:269-72.
13. Thomas GS, Hussain IH. Acute disseminated encephalomyelitis: a report of six cases. *Med J Malaysia.* 2004;59:342-51.
14. Tripathy S, Routray PK, Mohapatra AK, Mohapatra M, Dash SC. Acute demyelinating encephalomyelitis after anti-venom therapy in Russell's viper bite. *J Med Toxicol*. 2010;6:318-21.
15. 15. Patra KC, Shirolkar MS, Ghane VR. Acute disseminated encephalomyelitis: extremely rare presentation of pediatric human immunodeficiency virus infection. *J Peadiatr Neurosci*. 2014;9:150-53.
16. Kaymakamzade B, Karabudak R, Kurne AT, Nurlu G. Acute disseminated encephalomyelitis after oral therapy with herbal extracts: a case report. *Balkan Med J*. 2016;33:366-69.
17. Sheikh AAE, Sheikh AB, Sagheer S, et al. Acute intermittent porphyria: a rare cause of acute disseminated encephalomyelitis. *Cureus.* 2018;10:e2989.
18. Garg RK. Acute disseminated encephalomyelitis. Postgrad Med J. 2003;79:11-17.
19. Marchioni E, Tavazzi E, Minoli L, et al. Acute disseminated encephalomyelitis. *Neurol Sci.* 2008;29:286-88.
20. Yang HQ, Zhao WC, Yang WM, et al. Clinical profiles and short-term outcomes of acute disseminated encephalomyelitis in adult Chinese patients. *J Clin Neurol*. 2016;12:282–88.
21. Lee YJ. Acute disseminated encephalomyelitis in children: differential diagnosis from multiple sclerosis on the basis of clinical course. *Korean J Pediatr*. 2011;54:234-40.
22. Mader I, Stock KW, Ettlin T, Probst A. Acute disseminated encephalomyelitis: MR and CT features. *AJNR Am J Neuroradiol.* 1996;17:104-9.