Coexistence of Transverse Myelitis and Acute Disseminated Encephalomyelitis; a rare case

Transvers Miyelit ve Akut Dissemine Ensefalomiyelit Birlikteliği; Nadir Bir Olgu

Nurten NAS KIRDAR[1], Özlem ASLAN[2]

[1] Department of Physical Medicine and Rehabilitation, Bandırma Onyedi Eylül University, Faculty of Medicine, Balıkesir

[2] Department of Physical Medicine and Rehabilitation, Denizli State Hospital

Aim: Transverse myelitis (TM) is a disease in which motor, sensory and autonomic dysfunction develops as a result of localized inflammation of the spinal cord. Acute disseminated encephalomyelitis (ADEM) is a disease of sudden onset and variable clinical course characterized by demyilination of the central nervous system. Although ADEM and TM are separate entities clinically and radiologically, they have many similarities in terms of treatment and rehabilitation approach.

Case report: A 14-year-old female patient applied to the emergency service with the complaints of lowback pain, weakness in the legs, stumbling and falling. In addition to the paraplegia clinic, there were sensory defect, urinary retention and loss of control of the anal sphincter. ADEM and TM were diagnosed by considering the differential diagnoses due to demyelinating plaques seen in cranial and spinal MRI examinations. High-dose methyl prednisolone and IVIG (intravenous immunoglobulin) were given for treatment. After this treatment, dorsiflexion muscle strength of both ankles improved to 2/5. After starting the rehabilitation program, it was observed that there was a gradual improvement in muscle strength. At the end of the 3rd month, the patient was able to walk independently without support, urinate spontaneously and easily do her daily work.

Conclusion: Despite the poor prognostic factors, we achieved very good results with an effective medical treatment and rehabilitation program in our case with the association of ADEM and TM, which we encountered as a rare condition. We hope that our case will contribute to the literature on the importance of rehabilitation.

Keywords: Acute disseminated encephalomyelitis; neurological rehabilitation; transverse myelitis

Amaç: Transvers miyelit (TM), spinal kordun lokalize inflamasyonu sonucu motor, duyusal ve otonomik disfonksiyonun geliştiği bir hastalıktır. Akut dissemine ensefalomiyelit (ADEM), merkezi sinir sisteminin demiyelinizasyonu ile karakterize, ani başlangıçlı ve değişken klinik seyirli bir hastalıktır. ADEM ve transvers miyelit klinik ve radyolojik olarak ayrı antiteler olsalar da tedavi ve rehabilitasyon yaklaşımı açısından birçok benzerlikleri vardır.

Olgu sunumu: 14 yaşında kadın hasta, bel ağrısı, bacaklarda güçsüzlük, sendeleme ve düşme şikayetleri ile acil servise başvurdu. Parapleji kliniğine ek olarak seviye veren duyu kusuru, idrar retansiyonu ve anal sfinkterin kontrol kaybı mevcuttu. Ayırıcı tanılar da göz önünde bulundurularak kranial ve spinal MRG tetkiklerinde görülen demiyelinizan plaklar nedeniyle ADEM ve TM tanısı konuldu. Tedavi olarak yüksek doz metilprednizolon ve İVİG (intravenöz immünglobulin) verildi. Bu tedaviden sonra her iki ayak bileğinin dorsifleksiyon kas kuvveti 2/5'e yükseldi. Rehabilitasyon programına başladıktan sonra kas kuvvetinde kademeli bir iyileşme olduğu gözlendi. 3. ayın sonunda hasta bağımsız yürüyebiliyor, idrarını spontan yapabiliyor ve günlük işlerini rahatlıkla yapabiliyordu.

Sonuç: Nadir bir durum olarak karşılaştığımız ADEM ve TM birlikteliği olan olgumuzda kötü prognostik faktörlere rağmen etkili bir medikal tedavi ve rehabilitasyon programı ile çok iyi sonuçlar elde ettik. Olgumuzun rehabilitasyonun önemi konusunda literatüre katkı sağlamasını umuyoruz.

Anahtar Kelimeler: Akut dissemine ensefalomiyelit; nörolojik rehabilitasyon; transvers miyelit

Corresponding Author: Nurten Nas Kırdar **e-mail:** nrtnns@gmail.com
Received: 9 May 2024 **Accepted:** 8 August 2024 **DOI:** 10.33716/bmedj.1480959

INTRODUCTION

Transverse myelitis is a disease in which motor, sensory and autonomic dysfunction develops as a result of localized inflammation of the spinal cord. It progresses with demyelination and neuronal damage in the spinal cord (Defresne, 2001).

Acute disseminated encephalomyelitis (ADEM) is a disease of sudden onset and variable clinical course characterized by demyelination of the central nervous system (Stonehouse, 2003).

In this article, we will present a rare case of ADEM and transverse myelitis association.

CASE REPORT

A 14-year-old female patient, who stated that her first complaint was low back pain that started 2 days ago, applied to the emergency service with the complaints of weakness in the legs, stumbling and falling. In the physical examination, bilateral lower extremity muscle strengths (hip flexion, extension, abduction, knee extension, ankle dorsiflexion, big toe extension, ankle plantar flexion) were evaluated as 0/5. There was loss of sensation below the umbilical level. Lower extremity deep tendon reflexes could not be obtained, and there were no pathological reflexes. The patient had difficulty urinating and had lost control of the anal sphincter. Bilateral upper extremity muscle strengths were 5/5, deep tendon reflexes were normoactive, and there were no pathological reflexes.

The patient's vital signs (fever, blood pressure measurement, pulse, respiration) were stable. Complete blood count, kidney function tests, liver function tests, electrolytes, CRP, blood sugar results and complete urinalysis were normal.

In the radiological imaging, lumbar MRI performed due to low back pain did not reveal any finding other than "L5-S1 central focal bulging". Contrast-enhanced cranial MRI: Gliotic signals reaching a size of approximately 19x9 mm, extending to callosa septal surfaces in the periventricular and supraventricular deep white matter, suggested demyelinating disease. There is no pathological contrast enhancement in the cranial neural parenchyma in the post-contrast series (Figure 1).

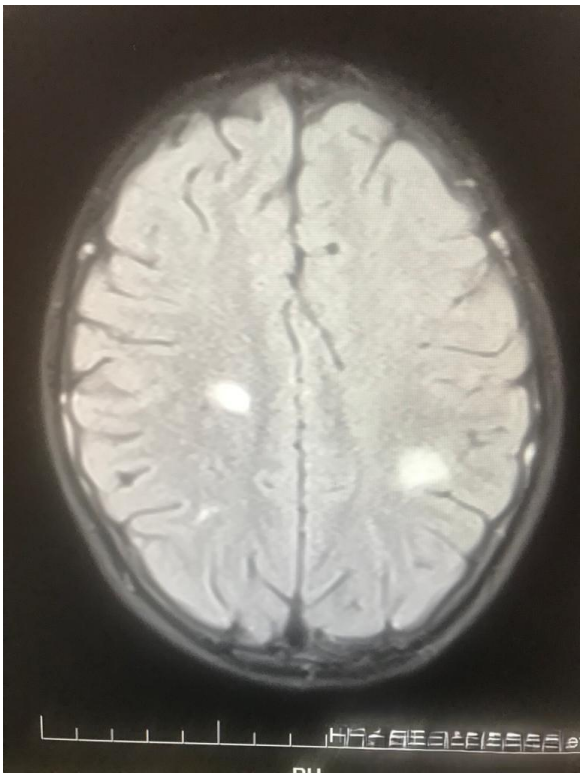

Figure 1: Contrast-enhanced cranial MRI: Gliotic signals reaching a size of approximately 19x9mm, extending to callosa septal surfaces in the periventricular and supraventricular deep white matter, suggested demyelinating disease. There was no pathological contrast enhancement in the cranial neural parenchyma in the post-contrast series.

Cervical MRI: T2 hyperintense signal increases in the cervical spinal cord at the C2-C3 level suggested a demyelinating plaque (Figure 2).

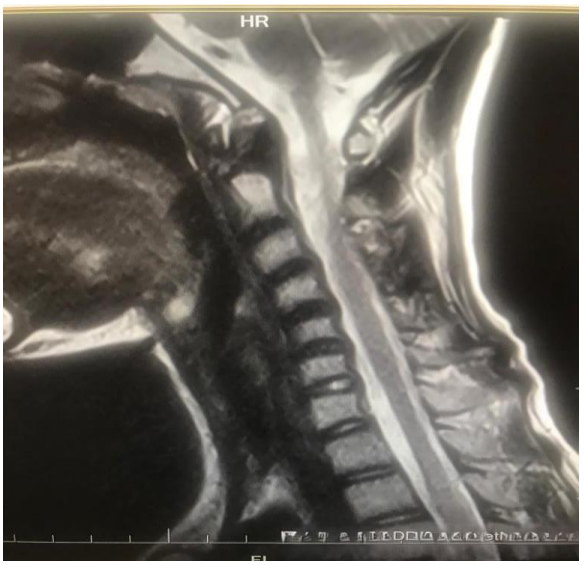

Figure 2: Cervical MRI: T2 hyperintense signal increases in the cervical spinal cord at the C2-C3 level suggested a demyelinating plaque.

Thoracic MRI: Signal increases were observed between T1 and T3 in the spinal medullary cord at the upper thoracic levels. This appearance is suspicious for demyelinating plaques (Figure 3).

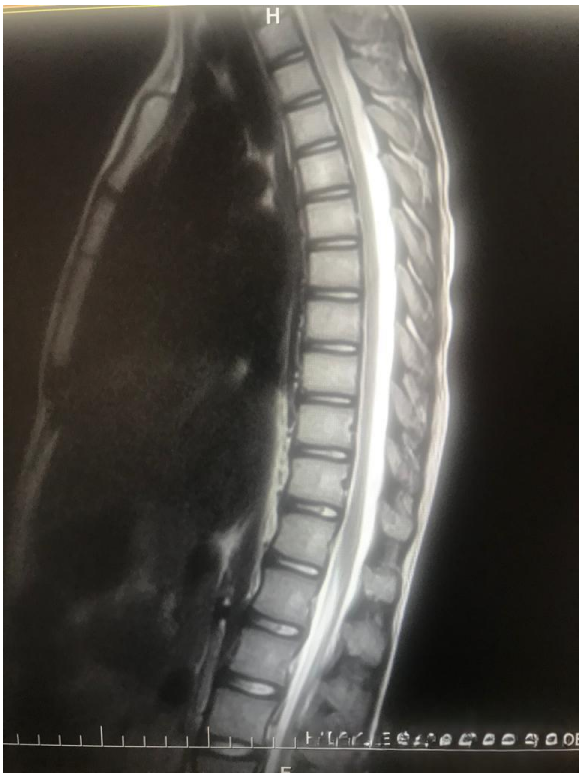

Figure 3: Thoracic MRI: Signal increases were observed between T1 and T3 in the spinal medullary cord at the upper thoracic levels. This appearance is suspicious for demyelinating plaques.

Based on these results, the patient was admitted to the pediatric neurology service.

Etiological examinations were performed in the pediatric neurology service. The test results for viral and bacterial agents were negative. Tests such as ANA, anti ds DNA, which are examined in terms of autoimmune diseases, were negative. In the cerebrospinal fluid examination; glucose: 94 mg/dl (normal CSF glucose value: 40-70 mg/dl), protein: 46.9 mg/dl (normal CSF protein: 15-45 mg/dl), CSF pressure: 15 cmH2O. No cells were seen in direct examination and bacteria did not grow in the CSF culture. Urinary system USG: moderate dilatation was observed in both kidney collecting systems. Pelvis AP diameter was measured 8 mm on the left and 9 mm on the right. Bladder was observed as a prominent globe. A urinary catheter was then inserted into the patient. The patient, who had acute onset walking difficulties, loss of muscle strength in the legs, and inability to urinate, was diagnosed with transverse myelitis after excluding the differential diagnoses, considering clinical, laboratory and imaging findings.

High dose methyl prednisolone 30 mg/kg/day (1000 mg) was started and given for 7 days. Since there was no improvement in muscle strength, IVIG (intravenous immunoglobulin) (400 mg/kg/day) was started 3 days later. IVIG was given for 5 days. After this treatment, dorsiflexion muscle strength of both ankles improved to 2/5, more prominently on the left. The patient whose general condition was stable and started to benefit from the treatment was transferred to our Physical Medicine and Rehabilitation service for the rehabilitation process.

The rehabilitation program was started with bilateral lower extremity joints ROM (range of motion) exercise, weak muscles strengthening exercise, neuromuscular electrical stimulation, bilateral upper extremity strengthening and back muscle strengthening exercises, sitting balance and balance coordination exercises, and progressive ambulation training. The intensity of the program was gradually increased in the follow-ups according to the patient's condition. Necessary precautions were taken against possible accompanying conditions such as DVT (deep vein thrombosis) and pressure ulcers. The patient was given bladder exercises for neurogenic bladder and bowel training for neurogenic bowel. In addition, 1 mg/kg/day methyl prednisolone, which was started as an oral maintenance dose, was given for 4 weeks, then the dose was tapered and stopped.

After about 10 days, the patient was able to sit unsupportedly and tolerated standing on the parapodium for half an hour. There was no new muscle strength gain other than minimal ankle dorsiflexion for 3 weeks. The absence of motor recovery for such a long period of time started to suggest that the prognosis would be poor, but hip flexion and abduction muscle strengths began to return at the beginning of the fourth week. Then she started to do knee extension. After a few days of standing on the side of the bed, locking the knees and stepping, she started to mobilize for short distances with a walker. She walked with a walker for 1 month. Meanwhile, in the follow-ups for the bladder,

postvoiding residual urine amount, which was 1450 cc at the beginning, was measured as 400 cc and 150 cc, 2 weeks apart, respectively. Urinary catheter and bladder exercises were continued. The patient, who had some improvement in muscle strength, went out of the walker and started walking short distances with the support of 1 person. Then she started going up and down stairs with support. At the end of 3 months, in the patient's examination; bilateral upper extremity muscle strengths 5/5, right lower extremity ankle dorsiflexion 4+/5, others 5/5, left lower extremity whole muscle strength was 5/5. Bilateral lower extremity deep tendon reflexes were hyperactive, Babinski reflex was positive, clonus was positive. Hoffmann sign was negative. There was no sensory defect, no spasticity. Anal sphincter control was partially present but inadequate. The urinary catheter was removed after the last amount of residual urine checked was 80 cc, and the patient started to urinate spontaneously. Control MRI examinations of the patient were performed 3 months later. It was observed that the lesions in the cranial and thoracic regions were healed, and the lesions in the cervical were completely disappeared. The patient was now able to sit, stand, walk and do daily work independently. Informed consent was obtained from the family to publish this report.

DISCUSSION

With this case, we aimed to draw attention to the association of ADEM and transverse myelitis and the importance of an effective rehabilitation process in addition to medical treatments.

Although ADEM and transverse myelitis are separate entities clinically and radiologically, they have many similarities in terms of treatment and rehabilitation approach (Yiu et al., 2009). When we review the literature, we see a few cases with ADEM and transverse myelitis coexistence (Poyrazoğlu et al., 2022; Sarioglu et al., 2014). In these cases, we see that COVID-19 and HSV infection are etiological factors. In our case, however, we could not find any cause in the investigations we performed for the etiology.

High-dose methyl prednisolone and IVIG intravenous immunoglobulin) are often sufficient in the treatment of myelitis. Rarely, cases that do not respond to these treatments require plasmapheresis (Aslan & Güngör, 2021; Nishiyama et al., 2019). In our case, improvement began to be seen after pulse steroid and IVIG treatment. Plasmapheresis was not required.

In a study evaluating the prognostic parameters in terms of acute transverse myelitis in children, pleocytosis in CSF examination, absence of tetraparesis and a long time to maximal motor deficit were found to be associated with good prognosis (Ganelin-Cohen et al., 2020). In another study, back pain, rapid progression of symptoms, severe motor deficit and spinal shock were found to be poor prognostic factors (Chen et al., 2013). Considering our case, we thought that the prognosis would be poor because of back pain, rapid progression of symptoms, and the patient's becoming paraplegic in a short time. In addition, delayed response to medical treatment and accompanying urinary retention and loss of anal sphincter control suggested that the process would be difficult. In a study investigating neurogenic bladder in children with transverse myelitis, it was reported that 55% of the patients had urinary retention (Hannallah et al., 2021). In our case, a permanent urinary catheter was used for approximately 2.5 months, but then the catheter was withdrawn due to the improvement in bladder functions. As a result, despite the poor prognostic factors, we achieved very good results with an effective medical treatment and rehabilitation program in our case with the association of ADEM and transverse myelitis, which we encountered as a rare condition. We also hope that our case will contribute to the literature on the importance of rehabilitation.

Authorship Contributions: Study conception and design: NNK, ÖA; Data collection: NNK, ÖA; analysis and interpretation of results: NNK, ÖA; draft manuscript preparation: NNK, ÖA. All authors reviewed the results and approved the final version of the manuscript.

Conflict of Interest: The authors declare that there is no conflict of interest.

Ethical approval: Informed consent was obtained from the family to publish this report.

Funding: No funding sources.

REFERENCES

Aslan, M., & Güngör, S. (2021). Çocukluk çağında miyelit ile başvuran olguların değerlendirilmesi. *İstanbul Tıp Fakültesi Dergisi*, 84(2). https://doi.org/10.26650/IUITFD.2019.0048

Chen, L., Li, J., Guo, Z., Liao, S., & Jiang, L. (2013). Prognostic indicators of acute transverse myelitis in 39 children. *Pediatric Neurology*, 49(6), 397–400. https://doi.org/10.1016/j.pediatrneurol.2013.08.022

Defresne, P. (2001). Efficacy of high dose steroid therapy in children with severe acute transverse myelitis. *Journal of Neurology, Neurosurgery & Psychiatry*, 71(2), 272–274. https://doi.org/10.1136/jnnp.71.2.272

Ganelin-Cohen, E., Konen, O., Nevo, Y., et al. (2020). Prognostic parameters of acute transverse myelitis in children. *Journal of Child Neurology*, 35(14), 999–1003. https://doi.org/10.1177/0883073820947512

Hannallah, A., Passoni, N., Peters, C. A., Shakir, N., Greenberg, B., & Jacobs, M. (2021). Bladder management in children with transverse myelitis. *Journal of Pediatric Urology*, 17(4), 522.e1–522.e6. https://doi.org/10.1016/j.jpurol.2021.04.003

Nishiyama, M., Nagase, H., Tomioka, K., et al. (2019). Clinical time course of pediatric acute disseminated encephalomyelitis. *Brain and Development*, 41(6), 531–537. https://doi.org/10.1016/j.braindev.2019.02.011

Poyrazoğlu, H. G., Kırık, S., Sarı, M. Y., Esen, İ., Toraman, Z. A., & Eroğlu, Y. (2022). Acute demyelinating encephalomyelitis and transverse myelitis in a child with COVID-19. *The Turkish Journal of Pediatrics*, 64(1), 133. https://doi.org/10.24953/turkjped.2020.3385

Sarioglu, B., Kose, S. S., Saritas, S., Kose, E., Kanik, A., & Helvaci, M. (2014). Severe acute disseminated encephalomyelitis with clinical findings of transverse myelitis after herpes simplex virus infection. *Journal of Child Neurology*, 29(11), 1519–1523. https://doi.org/10.1177/0883073813513334

Stonehouse, M. (2003). Acute disseminated encephalomyelitis: Recognition in the hands of general paediatricians. *Archives of Disease in Childhood*, 88(2), 122–124. https://doi.org/10.1136/adc.88.2.122

Yiu, E. M., Kornberg, A. J., Ryan, M. M., Coleman, L. T., & Mackay, M. T. (2009). Acute transverse myelitis and acute disseminated encephalomyelitis in childhood: Spectrum or separate entities? *Journal of Child Neurology*, 24(3), 287–296. https://doi.org/10.1177/0883073808323522