CASE REPORT / OLGU SUNUMU

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MEDICAL SCIENCES / DAHİLİ TIP BİLİMLERİ

Elsberg Syndrome: One of the Causes of Cauda Equina Syndrome - A Case Series and Review of the Literature

Elsberg Sendromu: Kauda Ekina Sendromunun Nedenlerinden Biri - Olgu Serisi ve Literatürün Gözden Geçirilmesi

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Abstract

Elsberg syndrome (ES) is a rare syndrome characterized by acute or subacute lumbosacral radiculitis and myelitis confined to the lower spinal cord, following an acute or reactivation viral infection. Reported complaints are urinary retention, bowel incontinence, radicular pain and muscle weakness. It is associated with Herpes simplex type 2 infections or reactivation from dorsal root ganglia, there are also cases reported with acute west Nile virus, varicella zoster virus and severe acute respiratory syndrome coronavirus 2. Clinicians do not generally consider this syndrome in the differential diagnosis of acute cauda equina syndrome, and therefore, appropriate diagnostic approaches are missed. Sarcoidosis, lymphoma, vasculitis, other sources of infection and dural arteriovenous fistula are also included in the differential diagnosis.

In order to draw attention to ES, which is a post viral inflammatory condition, two cases presenting acute cauda equina syndrome findings and diagnosed with ES will be presented.

Key Words: Elsberg Syndrome, Herpes Simplex Type 2, Miyelitis

Öz

Elsberg sendromu (ES), akut veya reaktif bir viral enfeksiyonu takiben omurilik alt bölgesinde sınırlı,akut veya subakut lumbosakral radikülit ve miyelit ile karakterize nadir bir sendromdur. Bildirilen semptomlar idrar retansiyonu, bağırsak inkontinansı, radiküler ağrı ve kas güçsüzlüğüdür. Herpes simpleks tip 2 enfeksiyonları veya dorsal kök gangliyonlarından reaktivasyon ile ilişkilidir, ayrıca akut batı Nil virüsü, varisella zoster virüsü ve Şiddetli akut solunum yolu sendromu koronavirüs-2 ile bildirilen olgular da vardır. Klinisyenler genellikle bu sendromu akut kauda ekina sendromunun ayırıcı tanısında değerlendirmez ve bu nedenle uygun tanısal yaklaşımlar gözden kaçar. Sarkoidoz, lenfoma, vaskülit, diğer enfeksiyon kaynakları ve dural arteriyovenöz fistül de ayırıcı tanıda yer almaktadır. Post viral enflamatuvar bir durum olan ES'ye dikkat çekmek amacıyla akut kauda ekuina sendromu bulguları gösteren ve ES tanısı alan iki olgu sunulacaktır.

Anahtar Kelimeler: Elsberg Sendrom, Herpes Simpleks Virüs Tip 2, Miyelit

Introduction

Elsberg syndrome (ES) is a rare syndrome characterized by acute or subacute lumbosacral radiculitis and myelitis limited to the lower spinal cord following an acute or reactivating viral infection. Reported complaints are urinary retention, bowel incontinence, radicular pain and muscle weakness. It is associated with Herpes simplex type 2 (HSV) infections or reactivation from dorsal root ganglia, there are also cases reported with acute west Nile virus (1), varicella zoster virus (2) and severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) (3). Clinicians do not generally consider this syndrome

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in the differential diagnosis of acute cauda equina syndrome, and therefore, appropriate diagnostic approaches are missed. Sarcoidosis, lymphoma, vasculitis, other sources of infection and dural arteriovenous fistula are also included in the differential diagnosis.

In order to draw attention to ES, which is a post viral inflammatory condition, two cases presenting acute cauda equina syndrome findings and diagnosed with ES will be presented.

Case Presentation

Case 1

A 56-year-old woman with no known chronic diseases was admitted to Ankara University Faculty of Medicine Department of Neurosurgery to be operated on with preliminary diagnosis of lumbar disc herniation due to low back and right leg pain, numbness in the inner right leg and urinary and faecal incontinence for 10 days. Since a disc herniation was not detected in lumbar magnetic resonance imaging (MRI) (Figure 1) to explain cauda equina syndrome, the patient was consulted at Neurology and Physical Medicine and Rehabilitation (PMR) Clinics. In an examination by PMR Department, the patient's lumbar movements were found painful but not restricted, and a straight leg raising test was found positive on the right side. On neurological examination, light touch and pain sensations were found mildly impaired through the right L4 dermatome and there was an erythematous patch with a few vesicles on it (Figure 2). Loss of muscle strength in right knee extension [4/5- Medical Research Council Grading (MRC)] was detected and right patellar deep tendon reflex could not be obtained. Anal reflex was decreased at the right side.

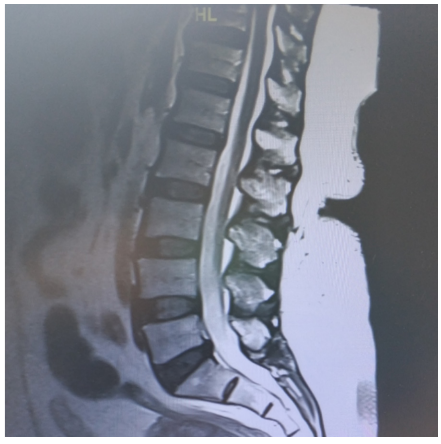

Figure 1: Coronal section of lumbar MRI showing multilevel disc bulging which could not explain the clinic findings

MRI: Magnetic resonance imaging

Consultation from the Dermatology and Neurology Clinic were requested. Dermatology Department evaluated vesicular lesions as HSV infection. Serum HSV IgM and IgG levels were requested. IgG test resulted positive and IgM resulted negative. Neurology Department recommended performing ENMG with a preliminary diagnosis of radiculopathy and lumbar MRI with contrast to exclude neoplasm. Contrast-enhanced lumbar MRI revealed multiple bulging and thickening of the right L4 root, and contrast enhancement in the right L4 root in post-contrast series (Figure 3, 4). The patient's first ENMG revealed normal findings. ENMG was repeated 2 weeks later and it was found

Figure 2: Case 1) Grouped vesicles on erythematous base

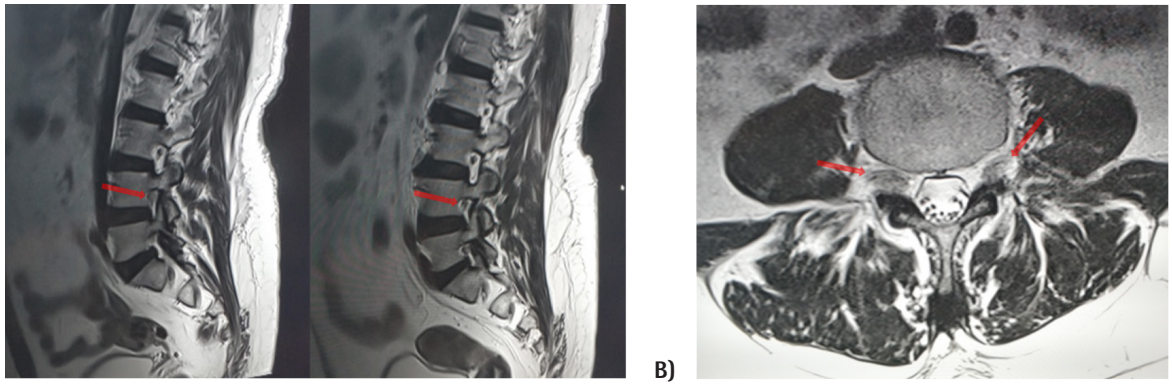

Figure 3: Case 1) **A)** Coronal section of lumbar MRI. Right L4 root was thickened **B)** Axial section of lumbar MRI. The right L4 root was thicker than the left one

MRI: Magnetic resonance imaging

to be compatible with L4 root involvement. A lumbar puncture procedure was planned for the patient for Cerebrospinal fluid analysis. However, the patient refused. With these findings, the patient was diagnosed with ES. The treatment was initiated with an oral valacyclovir 1000 mg every 8 hour for 7 days. The full sensation of the need to defecate had not returned and she still had occasional episodes of incontinence of faeces. The patient did not want to come to the hospital for control due to the coronavirus disease-2019 (COVID-19) pandemic. A telephone interview at 2 months revealed that even though there was further improvement in urinary continence control, faecal incontinence was still going on (Figures 4, 5).

Case 2

A 52-year-old woman was admitted to the PTR department with radicular pain in the back and left leg that started 2 days ago and progressed gradually. She also suffered from abdominal distension due to difficulty with defecation and forced urination. She had no comorbidity in his past medical history and her vital signs were stable. In musculoskeletal physical examination; the patient's lumbar movements were found painful but not restricted and a straight leg raising test was found positive on the left side. On neurological examination, pain and light touch sensations were found impaired mildly through the left S1 dermatomal area. Achille tendon jerk was slightly decreased compared to the right. There was weakness in left ankle plantar flexion and it was graded as 4/5 according to MRC. Anal reflex was decreased at the left side. Other neurological examination was unremarkable. On examination, vesicular lesions were found above the gluteal fold in the S1 dermatome area on the left side (Figure 5). Laboratory results (complete blood count, biochemical tests, inflammatory and autoimmune markers for rheumatic diseases, coagulation profile) revealed within normal range. Nasopharyngeal SARS-CoV-2 reverse-transcription polymerase chain reaction was negative twice. The screening for Epstein-Barr virus, Cytomegalovirus, HSV, and Herpes Zoster Virus, HIV was negative for IgM on serum. The varicella-zoster and HSV type 1 was positive for IgG on serum. While cervical and thoracal MRI was normal, an increase in thickness is observed in the left S1 nerve root compared to the right was shown in lumbar MRI (Figure 6).

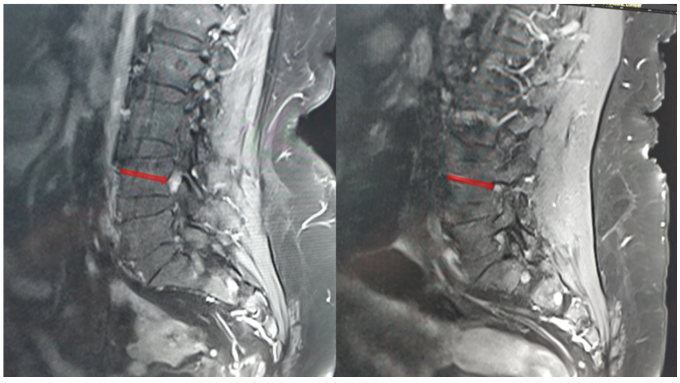

Figure 4: Case 1) Contrast-enhanced coronal lumbar MRI revealed thickening of the right L4 root with contrast enhancement in the right L4 root in postcontrast series

MRI: Magnetic resonance imaging

Figure 5: Case 2) Grouped vesicles above the gluteal fold in S1 dermatome

Lumbar puncture procedure was planned for the patient for cerebrospinal fluid (CSF) analysis. The patient refused it. The treatment was started with oral acyclovir 5x800/day for 5 days. The patient received 1 g per day of methylprednisolone for 3 days, concurrently. She gradually improved after 2 weeks and did not have any problem with urination and defecation. ENMG findings were consistent with S1 root involvement. The patient was able to empty her bladder, with a residual volume of less than 20 mL measured by ultrasound.

Discussion

We herein present the 2 cases with ES who had radicular pain, saddle hypoesthesia, and urinary retention associated with vesicular lesions on corresponding

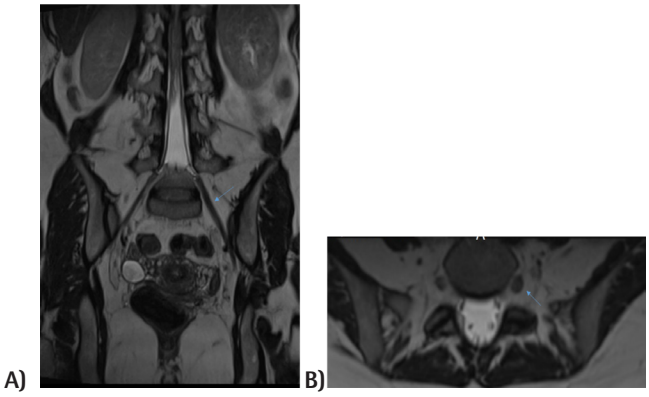

Figure 6: Case 2) **A:** T2-weighted coronal plane image. An increase in thickness is observed in the left S1 nerve root compared to the right. **B:** T2-weighted axial plane image. An increase in thickness is observed in the left S1 nerve root compared to the right

dermatomes. Immunocompromised states [such as HIV (4-7), immunosuppressing treatment (6-8), diabetes mellitus (9), myeloma (10), surgery (11)] make patient more susceptible to ES. It is usually not considered in the differential diagnosis of acute cauda equina syndrome. The diagnosis of ES is challenging for clinicians. There are small series and case reports in the literature (1-9). However, it is considered very rare (2). Although ES is thought to be mostly associated with HSV infections, cases associated with varicella-zoster, HIV, and currently COVID-19 infections have also been described (2,3). Savoldi et al. (2) recently proposed a set of diagnostic criteria based on clinical, radiological, and microbiologic features for ES (Table 1). Five levels of categories are defined for the diagnosis of ES: Laboratory-supported definite, clinically definite, clinically probable, clinically possible, and excluded. According to these diagnostic criteria, our cases were evaluated as "clinically definite"s ES with acute onset cauda equina syndrome and detection of radiculitis in MRI, root involvement in ENMG, and HSV-related skin lesions. Although the determined radiological findings are not specific, they are important in excluding other diagnoses in the differential diagnosis, which is also stated by Savoldi et al. (2) Evidence for causative virus infection and exclusion of alternative causes are important in the definitive diagnosis of ES. Viral detection in CSF is the diagnostic gold standard. But the absence of infection in the CSF does not rule out the diagnosis. Considering the sensitivity of serologic and virologic studies, the difficulty in obtaining samples from the CSF, and delayed sampling, clinical and radiological evidence may suggest ES regardless of microbiologic findings. Clinicians should also consider potential other viral infections in patients who do not have a rash but may have ES. Diagnostic criteria for ES provided flexibility for clinicians not to miss the diagnosis, especially in the absence of serological findings. Both of our cases were diagnosed "clinically definite" ES with acute onset cauda equina syndrome, detection of radiculitis in MRI, root involvement in ENMG and HSV-associated skin lesions according to these diagnostic criteria (2).

The ideal treatment of ES is debatable. Varicella-zoster and HSV are considered as causes; early acyclovir treatment is thought to be effective. In addition, corticosteroid treatment and plasma exchange have been used alone or in combination with acyclovir (2). Acyclovir treatment was applied in both of our cases. In the second case, additional corticosteroid treatment was given. In our second case, the recovery was complete. In the first case, the treatment response was incomplete due to the occasional episodes of faecal incontinence. There is a need to

Table 1: Elsberg syndrome according to diagnostic certainty

Categories	Criteria
1. Laboratory-supported definite	(A1 OR A2) AND B5
2. Clinically definite	A1 OR A2; B1 AND two of B2-B4; B1 and B2 (if concomitant)
3. Clinically probable	A1 OR A2; B1 AND one of B2-B4
4. Clinically possible	A1 OR A2; one of B1-B4
5. Excluded	Neither of A1 nor of A2; any of D1-D3
A. Required	
A1. Clinical symptoms and signs of cauda equina involvement: urinary hesitancy or retention; bowel incontinence, or severe constipation (erectile dysfunction insufficient on its own)	
A2. MRI or electrophysiologic evidence of cauda equina involvement: enhancement of cauda equina; EMG evidence of radiculopathy	
B. Supportive but not required	
B1. Time course: acute/subacute onset; no relapse; progression over less than 3 mo	
B2. Coexisting or recently preceding symptoms of genital herpes infection OR other clinical symptoms of herpes virus infection	
B3. Clinical (e.g., exaggerated reflexes and Babinski signs) or MRI evidence of myelitis in conus	
B4. CSF pleocytosis	
B5. Documented herpes virus infection from CSF by PCR, culture, or detection of IgM (immunoglobulin M); serology	
C. Red flags	
C1. Relapses beyond 1 y from onset	
D. Exclusionary	
D1. Myelitis extending rostral to T9	
D2. Other neurologic symptoms suggestive of alternative etiology: optic neuritis, brain/brainstem syndrome	
D3. Other etiology proven/more likely for syndrome: NMOSD (neuromyelitis optica spectrum disorder), dural arteriovenous fistula, viral transverse myelitis, other causes of myelopathy	

This table is taken from an article written by Savoldi et al. (2)
MRI: Magnetic resonance imaging, EMG: Electromyography, CSF: Cerebrospinal fluid, PCR: Polymerase chain reaction

document the treatment agent, its duration and dose, and the appropriate combination in the treatment of ES.

Conclusion

ES is a rare diagnosis that is not generally considered in the differential diagnosis and therefore can be missed. Low back pain and accompanying cauda equina syndrome are among the red flags for low back pain. It is important to make the differential diagnosis appropriately in order not to make unnecessary interventions to the patients. The fact that CSF analysis could not be performed in the presented cases is a limitation, but their clinical and radiological features are sufficient for diagnosis.

Ethics

Informed Consent: Informed consent taken from patients.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: H.O.A., Ş.K., S.G., A.G., Concept: H.O.A., Ş.K., S.G., A.G., Design: H.O.A., Ş.K., S.G., A.G., Data Collection and Processing: H.O.A., Ş.K., S.G., A.G., S.Ü., E.P., Z.Ö.A., Analysis or Interpretation: H.O.A., Ş.K., S.G., A.G. Literature Search: H.O.A., Ş.K., S.G., A.G. Writing: H.O.A., Ş.K.

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