CASE REPORT / Olgu Sunumu

Syringomyelia in The Patient Following The Diagnosis of Amyotrophic Lateral Sclerosis

Amyotrofik Lateral Skleroz Tanısı ile Takip Edilen Hastada Siringomiyeli

Emel Oğuz Akarsu, Bekir Enes Demiryürek

Sakarya Üniversitesi Eğitim Araştırma Hastanesi Nöroloji kliniği /Sakarya

Yazışma Adresi / Correspondence:
Bekir Enes Demiryürek

Sakarya Üniversitesi Eğitim Araştırma Hastanesi Adapazarı Sakarya
T: **+90 536 933 79 04** E-mail: **bekirenes10@gmail.com**

Geliş Tarihi / Received : **29.07.2018** Kabul Tarihi / Accepted : **27.09.2018**

Abstract

The syringomyelia refers to the development and progression of the cerebrospinal fluid (CSF) filling space within the spinal cord. We wanted to share with you the 56 years old male case with syringomyelia that misdiagnosed as amyotrophic lateral sclerosis (ALS) and using riluzole therapy. We wanted to share this phenomenon, which was diagnosed with ALS in terms of emphasizing the importance of neuroimaging. **(Sakarya Med J, 2018, 8(4):859-862).**

Keywords: syringomyelia; amyotrophic lateral sclerosis; trauma

Öz

Siringomiyeli, omurilikteki beyin omurilik sıvısı (BOS) dolgu boşluğunun gelişimini ve ilerlemesini ifade eder. 56 yaşında torakal siringomiyelisi olan ancak yanlış olarak ALS tanısı ve riluzol tedavisi alan erkek olguyu paylaşmak istedik. Klinisyenlerin nörogörüntülemenin önemini dikkate almalarını vurgulamak istedik. **(Sakarya Tıp Dergisi, 2018, 8(4):859-862)**

Anahtar kelimeler: siringomiyeli; amyotrofik lateral skleroz; travma

Background

The syringomyelia refers to the development and progression of the CSF filling space within the spinal cord. As its name implies, this particular type occurs after a syringomyelia, known or suspected trauma. It differs from syringomyelia due to congenital malformations, such as Arnold-Chiari, where blockage of normal CSF flow through the cerebral ventricles may result in a syrinxs that is a secondary phenomenon.[1] We wanted to share with you the case with syringomyelia that misdiagnosed as ALS.

Case

A 56-year-old male patient presented with weakness and unstable walking complaints.

In his complaints of about six months, he said that there has not been an increase in the amount of everything since the beginning.

The patient noted pain and numbness in his legs. There was no urinary incontinence.

In the background; DM, cigarette smoking, a two-year-old lumber disc operation and a car traffic accident story a year ago. There was no property in her family history.

Neurological examination revealed no upper motor and sensory disturbance on the upper extremity, lower extremity 4/5 motor force, and DTRs lower extremity +++ / +++. Bottom skin reflex was bilateral extensor. There was no sensory malfunction.

In ENMG performed on the absence of abnormal findings in the cranial, cervical and lumbar MR patients who were admitted to the another hospital four months ago, moderate motor nerve BCAP responses were decreased in all extremities, decreased in motor speeds, and sensorial DSAP responses were low. Sensory disorders were diagnosed as diabetic polyneuropathy and the patient was diagnosed as Amyotrophic Lateral Sclerosis (ALS) and discharged with riluzole 2 * 50 mg/day treatment.

Thoracic spinal MRI, suspected of being diagnosed due to the absence of abnormal examination findings in the upper extremity of the patient, was found to be compatible with syrings of five vertebra sizes at the level of T5-T10 in the T1 and T2 sequence (Figure). The syringomyelia was diagnosed. Riluzole theraphy was stopped and the patient was transferred to the brain surgery department.

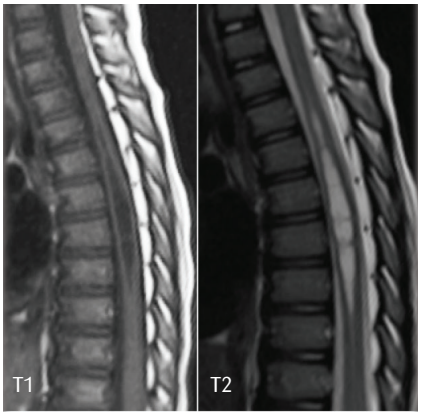

Figure: Thoracic spinal MRI with T1 and T2 sequences compatible with syrings at T5-T10

Discussion:

Syringomyelia is more common in the mid-lower cervical region, and more rarely in the thoracic and lumbosacral region and medulla oblongata (syringobulbi).[2] Symptoms such as syringomyelia spasticity, weakness, vitality in reflexes, impaired balance, numbness, paresthesia, sensory loss, muscle atrophy can be seen. Symptoms may be either a single limb for a long time or bilateral. [3,4] ALS was first suspected on the basis of a clinical concurrence of both upper and lower motoneurons signs, involving the four limbs and the bulbar region. The complete biochemical, electrophysiologic and imaging work-up then allowed the diagnosis of clinically definite ALS, according to the El Escorial-WFN revised criteria.[6] Syringomyelia may produce a clinical picture similar to ALS. However, the slow progression of the disease and the presence of sensory signs, along with the MRI evidence of the syringomyelic cyst generally allow the differential diagnosis.Spinal MRI is important in terms of differential diagnosis in cases of ALS considered because of similar clinical picture.[7]

Conclusion:

We wanted to share this phenomenon, which was diagnosed with ALS in terms of emphasizing the importance of neuroimaging, but which identified syrinxs in Thoracic MRI.

1. Yeo J, Cheng S, Hemley S, Lee BB, Stoodley M, Bilston L. Characteristics of CSF Velocity-Time Profile in Posttraumatic Syringomyelia. AJNR Am J Neuroradiol. 2017 Sep;38(9):1839-1844.
2. Smoker W: MR imaging of cranivertebral junction. Magn Res Imaging Clin North Am 2000; 8:635-651,
3. Curnes J, OakesW, Boyko O: MR imaging of hindbrain deformity in Chiari II patients with and without symptoms of brainstem compression. AJNR Am J Neuroradiol 1989;10:293-302,
4. Dones J, De Jesus O, Colen CB, Toledo MM, et al: Clinical outcomes in patients with Chiari I malformation: a review of 27 cases. Surg Neurol 2003;60(2):142-147
5. Brooks BR, Miller RG, Swash M, Munsat TL. El Escorial revisited: Revised criteria for the diagnosis of amyotrophic lateral sclerosis. Amyotroph Lateral Scler. 2000; 1:293-299.
6. Lo Coco D, Spataro R, Taiello AC, La Bella V. Thoracic Syringomyelia in a Patient with Amyotrophic Lateral Sclerosis. INDJ, 2015;3(4):136-140
7. Braddom R. Physical Medicine and Rehabilitation. 4th Ed. Philadelphia: W.B.Saunders, 1996. p. 729-755.