# A patient with bilateral neuralgic amyotrophy and hepatitis E virus infection: an educational communication

Bojan Rojc (1,2), Nejc Šarabon (3)

*(1) General Hospital Izola, Neurology Service, Izola, Slovenia; (2) University of Primorska, Faculty of Mathematics, Natural Sciences and Information Technologies, Koper, Slovenia; (3) University of Primorska, Faculty of Health Sciences, Department of Physiotherapy, Izola, Slovenia.*



***Abstract***

Neuralgic Amyotrophy, a peripheral nervous system disorder, is characterized by severe pain and muscle weakness, which can have a significant impact on patients' quality of life. The exact cause of neuralgic amyotrophy is unknown, but it may be linked to immunopathological mechanisms. Recent research has found an association between neuralgic amyotrophy and hepatitis E virus infection. This communication aims to expand knowledge on the clinical phenotype of patients with neuralgic amyotrophy and hepatitis E virus infection, presenting the case of a 55-year-old man diagnosed with bilateral neuralgic amyotrophy and hepatitis E virus infection.

**Key Words**: amyotrophy; peripheral neuropathy; hepatitis E; educational report.

*Eur J Transl Myol 34 (1) 12143, 2024 doi: 10.4081/ejtm.2024.12143*

Neuralgic Amyotrophy (NA), a peripheral nervous system disorder, presents a distinct clinical picture characterized by severe pain and muscle weakness. NA was considered to be a rare disease with an incidence of 1 – 3 per 100,000 per year, but some new studies report a much higher incidence, reaching as high as 1 per 1,000 per year.[1,2] The impact on patients' quality of life could be significant. The classic NA phenotype manifests as new onset pain in one shoulder or upper arm; several hours or days later paresis typically develops involving the long thoracic, suprascapular, and anterior interosseous nerves.[2-4] This clinical presentation may be seen in about two-thirds of patients with NA.[2] The exact pathophysiological mechanism of NA remains unknown, but immune and mechanical mechanisms could be involved.[2] In recent years, an association between NA and Hepatitis E virus (HEV) infection was reported.[5] HEV is a common cause of viral hepatitis; it is estimated that HEV infects about 20 million people each year.[7] HEV has also been linked to various neurological disorders like Guillain-Barré syndrome, encephalitis, myelitis, and NA.[7] About 10% of patients with NA have HEV infection.[6] Patients with NA and HEV present a distinct clinical phenotype that differs from the classic NA phenotype, with predominantly more extensive asymmetric bilateral involvement of the brachial plexus and more often causing damage outside the brachial plexus.[8-10] The aim of this report is to expand the knowledge of the clinical phenotype of patients with NA and HEV.

## Clinical Communication

All procedures performed in this study were in accordance with ethical standards of the 1964 Helsinki Declaration and its later amendments. Written informed consents to participate in this study have been signed by the patient.

A 55-year-old left-handed man, diagnosed with psoriasis and not receiving regular therapy, was referred to our hospital due to weakness in his right shoulder. Half a year ago, he visited the subtropical region of Latin America. The weekend before his admission, he developed fever. On the day of admission, he experienced burning pains in both upper limbs and a sudden weakness on the right side, primarily limiting elevation and abduction in the right shoulder joint. The neurological status revealed reduced muscle strength of abduction in the right shoulder joint and internal and external rotation of the shoulder. Myotatic reflexes were symmetrical. He reported hypesthesia of the lateral part of the right shoulder. Imaging diagnostics, including CT scans of the head and cervical spine, as well as MRIs of the head and cervical spine (Figure 1), showed a normal structure of the brain and cervical spine. The next day, he reported weakness in finger and wrist extension on the left. A suspicion of

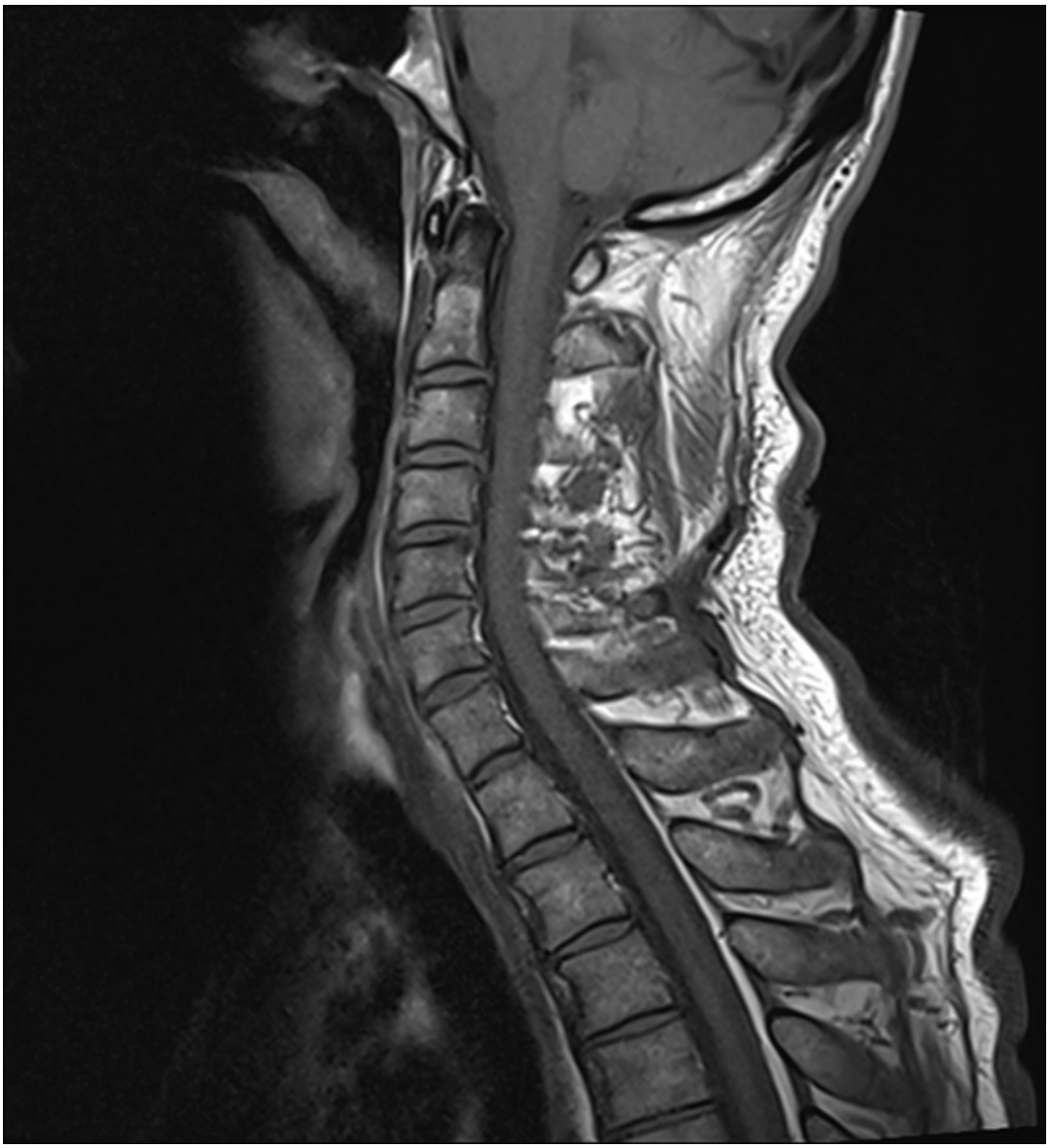

***Fig 1.*** *Magnetic resonance imaging of the cervical spine of the patient.*

mononeuritis multiplex with damage to the right axillary nerve and posterior interosseous branch of the left radial nerve was established. The electrodiagnostic exemination (EDX) examination confirmed acute denervation in both nerves. In the cerebrospinal fluid, leukocytes were borderline elevated. There were no oligoclonal bands in the cerebrospinal fluid, nor was there a disruption of the blood-brain barrier. The results for infections with Borrelia Burgdorferi, syphilis, cytomegalovirus, Epstein Barr virus, Varicella Zoster virus, HIV, hepatitis B, hepatitis C, and West Nile virus were all negative. Antiganglioside and paraneoplastic antibodies, determined in the blood and cerebrospinal fluid, were all negative. Rheumatological blood tests were also negative. Hemogram, liver tests, thyroid hormones, vitamin B12, folic acid, and proteinogram were all within the normal range. The patient had both IgG and IgM positive antibodies against HEV. The control EDX, conducted four days after the first examination, showed additional denervation activity in the right supraspinatus muscle. Based on the distribution of the lesion, a diagnosis of bilateral NA with HEV was established. The follow up exam after 6 months demonstrated an improvement of muscle strength. After 18 months the patient's muscle strength was in a normal range.

## Discussion

Patients with HEV associated NA are more likely to be middle-aged males (55 years) with bilateral NA (80% vs 8.6% in non-HEV NA).[8,9] The findings in our patient are consistent with previous reports; at the time of diagnosis,

he was 55 years old. The presentation was bilateral from the beginning of the manifestation of muscle weakness, with involvement of the right axillary and left posterior interosseous nerve at first, followed in the next days by subacute involvement of the right subscapular nerve. Our patient had mild pleocytosis in CSF, as was reported before in 25% of patients.[8] The level of liver enzymes at the time of diagnosis was in the normal range and he was not icteric; he had a predominantly neurological clinical picture, as was also reported in other cases.[8] Unlike in other cases, our patient did not have any extra brachial involvement; also, the phrenic nerve was not compromised.[8,9] He recovered after 18 months, as was expected from previous reports.[3,8]

In conclusion, our communication confirms the findings of previous reports and studies about the clinical phenotype of HEV associated NE.

## List of acronyms

EDX - electrodiagnostic exemination.
HEV - Hepatitis E virus
NA - neuralgic amyotrophy

## Contributions of Authors

Both authors conceptualized the paper, final proof read and approved it. Introduction and discussion was written by both co-authors, while the core case report was drafted by the first author. Both authors read and approved the final edited typescript.

## Acknowledgments

The authors are grateful to the patient for his kind cooperation during the study

## Funding

NŠ declares financial support for his scientific research activities by the Slovenian Research Agency through the research program KINSPO - Kinesiology for the effectiveness and prevention of musculoskeletal injuries in sports (P5-0443).

## Conflict of Interest

The authors declare they have no financial, personal, or other conflicts of interest.

## Ethical Publication Statement

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

## Corresponding Author

Nejc Šarabon, Faculty of health Sciences, University of Primorska, 6310 Izola, Slovenia. Phone +386 40 429 505
ORCID iD: 0000-0003-0747-3735.
Email: nejc.sarabon@fvz.upr.si

*E-mail and ORCID iD of co-author*

*Bojan Rojc: bojan.rojc@upr.si*
*ORCID iD: 0009-0000-3813-1127*

## References

1. van Alfen N, van Eijk JJ, Ennik T, Flynn SO, Nobacht IE, Groothuis JT, Pillen S, van de Laar FA. Incidence of neuralgic amyotrophy (Parsonage Turner syndrome) in a primary care setting--a prospective cohort study. PLoS One. 2015 May 27;10(5):e0128361. doi: 10.1371/journal.pone.0128361. PMID: 26016482; PMCID: PMC4445915.
2. Van Eijk JJ, Groothuis JT, Van Alfen N. Neuralgic amyotrophy: An update on diagnosis, pathophysiology, and treatment. Muscle Nerve. 2016 Mar;53(3):337-50. doi: 10.1002/mus.25008. Epub 2016 Jan 20. PMID: 26662794.
3. van Alfen N, van Engelen BG. The clinical spectrum of neuralgic amyotrophy in 246 cases. Brain. 2006 Feb;129(Pt 2):438-50. doi: 10.1093/brain/awh722. Epub 2005 Dec 21. PMID: 16371410.
4. Ferrante MA, Wilbourn AJ. Lesion distribution among 281 patients with sporadic neuralgic amyotrophy. Muscle Nerve. 2017 Jun;55(6):858-861. doi: 10.1002/mus.25422. Epub 2017 Feb 3. PMID: 27680713.
5. Fong F, Illahi M. Neuralgic amyotrophy associated with hepatitis E virus. Clin Neurol Neurosurg. 2009 Feb;111(2):193-5. doi: 10.1016/j.clineuro.2008.09.005. Epub 2008 Nov 4. PMID: 18986757.
6. van Eijk JJ, Madden RG, van der Eijk AA, Hunter JG, Reimerink JH, Bendall RP, Pas SD, Ellis V, van Alfen N, Beynon L, Southwell L, McLean B, Jacobs BC, van Engelen BG, Dalton HR. Neuralgic amyotrophy and hepatitis E virus infection. Neurology. 2014 Feb 11;82(6):498-503. doi: 10.1212/WNL.0000000000000112. Epub 2014 Jan 8. PMID: 24401685; PMCID: PMC3937863.
7. Webb GW, Dalton HR. Hepatitis E: an expanding epidemic with a range of complications. Clin Microbiol Infect. 2020 Jul;26(7):828-832. doi: 10.1016/j.cmi.2020.03.039. Epub 2020 Apr 3. PMID: 32251845.
8. van Eijk JJJ, Dalton HR, Ripellino P, Madden RG, Jones C, Fritz M, Gobbi C, Melli G, Pasi E, Herrod J, Lissmann RF, Ashraf HH, Abdelrahim M, Masri OABAL, Fraga M, Benninger D, Kuntzer T, Aubert V, Sahli R, Moradpour D, Blasco-Perrin H, Attarian S, Gérolami R, Colson P, Giordani MT, Hartl J, Pischke S, Lin NX, Mclean BN, Bendall RP, Panning M, Peron JM, Kamar N, Izopet J, Jacobs BC, van Alfen N, van Engelen BGM. Clinical phenotype and outcome of hepatitis E virus-associated neuralgic amyotrophy. Neurology. 2017 Aug 29;89(9):909-917. doi: 10.1212/WNL.0000000000004297. Epub 2017 Aug 2. PMID: 28768846.

9. Scanvion Q, Perez T, Cassim F, Outteryck O, Lanteri A, Hatron PY, Lambert M, Morell-Dubois S. Neuralgic amyotrophy triggered by hepatitis E virus: a particular phenotype. J Neurol. 2017 Apr;264(4):770-780. doi: 10.1007/s00415-017-8433-z. Epub 2017 Feb 28. PMID: 28247042.
10. Garofoli R, Zauderer J, Seror P, Roren A, Guerini H, Rannou F, Drapé JL, Nguyen C, Lefèvre-Colau MM. Neuralgic amyotrophy and hepatitis E infection: 6 prospective case reports. RMD Open. 2020 Nov;6(3):e001401. doi: 10.1136/rmdopen-2020-001401. PMID: 33219125; PMCID: PMC8011528.

## Disclaimer



Submission: November 28, 2023
Accepted for publication: December 28, 2023