Debilitating triad: Myelopathy, lymphoma, and severe polyneuropathy resulting from human T-lymphotropic virus

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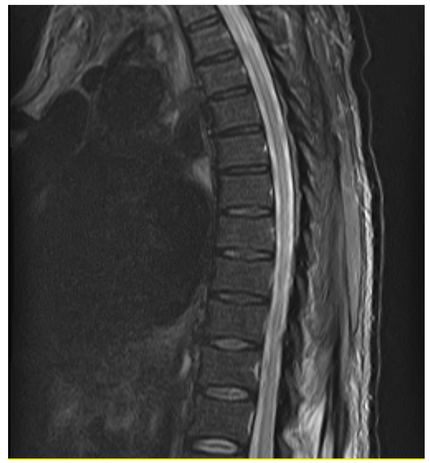
Case Description

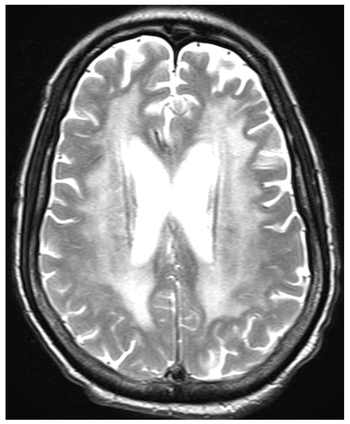
A 70-year-old male with a history of adult t-cell lymphoma leukemia (ATLL) and rapidly progressive myelopathy, both secondary to human T-lymphotropic virus type 1, presents to the emergency department from subacute rehab with complaints of worsening global weakness and hypophonic speech.

Physical exam on admission is consistent with C5 AIS B tetraplegia as well as hypophonic speech, lower extremity flaccidity, hyporeflexia, and absent Hoffman's sign. MRI spine shows central cord enhancement from C7 to conus. Serum studies are positive for HTLV 1/2. CSF studies demonstrate mildly elevated protein; however, HTLV1 CSF PCR was not available. Mixed upper and lower motor neuron findings as well as lack of expected response to plasmapheresis and corticosteroid treatment created suspicion for concurrent pathological process. Neurophysiologic studies showed evidence of severe axonal sensorimotor polyneuropathy. Treatment with pulsed corticosteroids provides improvement in symptoms to L2 AIS B, which is deemed to be his baseline functional status in the setting of likely malignancy-associated polyneuropathy.

Disclaimer:
This case presentation did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors. There are no conflicts of interest for the authors. His material is the result of work supported with resources and the use of facilities at the James A. Haley Veterans' Hospital. The contents do not represent the views of the Department of Veterans Affairs or the United States Government



Increased T2/STIR signal predominantly in the central cord extending from C7 down through the conus



Extensive confluent T2/FLAIR hyperintense signal involving the periventricular, deep, and subcortical white matter, the internal capsule, thalamus, midbrain, pons and medulla, both cerebellar hemispheres and middle cerebellar peduncles.

Discussion

HTLV-1 virus rarely leads to HTLV-1-associated myelopathy (also known as tropical spastic paraparesis) and ATLL, though the impact from these secondary diagnoses is severe and no treatments exist to reduce long-term disability. Pulsed-dose corticosteroids show promise for the possibility of breakthrough improvement in myelopathy; however, if clinical findings are not consistent with myelopathic pathology alone, concurrent polyneuropathy should be considered as this diagnosis can be associated with both virus and malignancy. In this case, the discovery of concomitant polyneuropathy allowed for reasonable conclusion of functional baseline and rehabilitation goals.

References

- Giam CZ. HTLV-1 Replication and Adult T Cell Leukemia Development. Recent Results Cancer Res. 2021;217:209-243. doi: 10.1007/978-3-030-57362-1_10. PMID: 33200368.
- Zuo X, Zhou R, Yang S, Ma G. HTLV-1 persistent infection and ATLL oncogenesis. J Med Virol. 2023 Jan;95(1):e28424. doi: 10.1002/jmv.28424. PMID: 36546414.
- Araujo A, Bangham CRM, Casseb J, Gotuzzo E, Jacobson S, Martin F, Penalva de Oliveira A, Puccioni-Sohler M, Taylor GP, Yamano Y. Management of HAM/TSP: Systematic Review and Consensus-based Recommendations 2019. Neurol Clin Pract. 2021 Feb;11(1):49-56. doi: 10.1212/CPJ.0000000000000832. PMID: 33968472; PMCID: PMC8101298.
- Iordan I, Onisâi M, Vlădăreanu AM, Mambet C, Marinescu EC, Nistor R, Bumbea H. Particularities of Neurological Manifestations in Adult T-Cell Leukemia/Lymphoma: Need for a Multidisciplinary Approach-A Narrative Review. Medicina (Kaunas). 2022 Oct 28;58(11):1553. doi: 10.3390/medicina58111553. PMID: 36363509; PMCID: PMC9693620.
- Bastos Ferreira AP, do Nascimento ADFS, Sampaio Rocha-Filho PA. Cerebral and spinal cord changes observed through magnetic resonance imaging in patients with HTLV-1-associated myelopathy/tropical spastic paraparesis: a systematic review. J Neurovirol. 2022 Feb;28(1):1-16. doi: 10.1007/s13365-021-01043-2. Epub 2022 Jan 3. PMID: 34981435.
- Yamauchi J, Tanabe K, Sato T, Nakagawa M, Matsuura E, Tsuboi Y, Tamaki K, Sakima H, Ishihara S, Ohta Y, Matsumoto N, Kono K, Yagishita N, Araya N, Takahashi K, Kunitomo Y, Nagasaka M, Coler-Reilly A, Hasegawa Y, Araujo A, Jacobson S, Grassi MFR, Galvão-Castro B, Bland M, Taylor GP, Martin F, Yamano Y. Efficacy of Corticosteroid Therapy for HTLV-1-Associated Myelopathy: A Randomized Controlled Trial (HAMLET-P). Viruses. 2022 Jan 12;14(1):136. doi: 10.3390/v14010136. PMID: 35062340; PMCID: PMC8780460.
- Tsutsumi S, Sato T, Yagishita N, Yamauchi J, Araya N, Hasegawa D, Nagasaka M, Coler-Reilly ALG, Inoue E, Takata A, Yamano Y. Real-world clinical course of HTLV-1-associated myelopathy/tropical spastic paraparesis (HAM/TSP) in Japan. Orphanet J Rare Dis. 2019 Oct 21;14(1):227. doi: 10.1186/s13023-019-1212-4. PMID: 31639014; PMCID: PMC6802124.