

Images in Hospital Medicine

Claw Hand in Parsonage Turner syndrome

Michael E Lazarus¹, Tyler B Larsen^{1,2}

¹ Internal Medicine, Department of Medicine, University of California, Los Angeles, CA, USA,

Journal of Brown Hospital Medicine

Vol. 3, Issue 1, 2024

Article Information

Keywords: Sudden upper extremity pain, Parsonage Turner Syndrome, Neuralgic amyotrophy, SARS-CoV-2, acute multi-focal motor neuropathy, ganglioside GD1a antibody

https://doi.org/10.56305/001c.90068

Submitted: October 29, 2023 EST

Accepted: November 11, 2023 EST

Abstract

An elderly patient with an extensive smoking history presented with acute severe right-hand pain and evolving weakness with patchy sensory deficit. He had minimal relief from multiple analgesics, gabapentin, and acupuncture. Motor deficits in his right-hand progressed. A nerve conduction study confirmed motor-predominant polyneuropathy and magnetic resonance imaging (MRI) of his C-spine and brachial plexus ruled out apical lung mass or other structural lesions. A positive GalNAc-GD1a ganglioside antibody (IgG) was noted. A diagnosis of immune mediated neuralgic amyotrophy or Parsonage-Turner Syndrome was made. Intravenous immune globulin (IVIG) was started with partial improvement in motor function, six months later. Although this patient did not have SARS-CoV-2 infection nor was he recently vaccinated, this case is of interest to hospital-based internists as the Covid pandemic has seen an increase in the in reported cases of neuralgic amyotrophy worldwide due to the virus itself as well as the associated COVID-19 vaccines.

A man in his 70s presented with one month of sudden right-hand weakness, numbness, and pain. Gabapentin, over-the-counter analgesics, and acupuncture provided no relief. He denied any other symptoms or any preceding trauma or injury. His medical history included hypertension and chronic obstructive pulmonary disease (COPD) and a one hundred fifty-pack year smoking history. He worked as a lawyer and denied recent viral infections, including any prior SARS-CoV-2 infection or recent vaccinations. Examination of his upper extremities revealed inability to close or open the fingers of his right hand (Fig 1, and Fig. 2). Power in right wrist dorsiflexion was 4/5, deep finger flexors 3/5, finger extension 2/5 with the weakest finger abduction being 1/5. Sensory deficit was noted in a patchy distribution over the right hand and forearm. Initial lab findings were unremarkable. His chest radiograph revealed no lung masses. A right upper extremity nerve conduction study revealed extensive motor-predominant polyneuropathy. Magnetic resonance imaging (MRI) of his cervical spine and brachial plexus ruled out apical mass or other structural lesions. Serologic testing for immune markers and paraneoplastic disease were obtained and revealed a positive anti-N-acetylgalactosaminyl GD1a (anti-GalNAc-GD1a) antibodies, a marker associated with immune mediated acute motor axonal neuropathy, and which predicts a good response to intravenous immune globulin (IVIG) therapy. A final diagnosis of immune-mediated right upper extremity focal neuropathy associated with GD1a antibody consistent with Parsonage-Turner Syndrome (PTS) was made.



Figure 1. Right (total) claw hand deformity with wasting of all intrinsic muscles of the hand: lumbricals, palmar interossei, and most prominently, the thenar eminence.

Treatment with IVIG started immediately and then at three-week intervals. At five-month follow-up, wrist extensors and triceps had improved power, but flexors and intrinsic hand muscles were still weak. IVIG and occupational therapy are currently ongoing. Given his extensive smoking history, regular health screening for an occult malignancy is also a priority. High on the initial differential diagnosis was a right sided lung malignancy in the apical right lobe invading the lower brachial plexus (Pancoast tumor) but this was quickly ruled out.

PTS is an often-overlooked acute multifocal motor neuropathy described by Parsonage and Turner in 1948.²

² Department of Medicine, VA Greater Los Angeles Healthcare System



Figure 2. Dorsal view of hands: Right total claw hand (Lower trunk of Brachial Plexus C8/T1) associated with Parsonage Turner Syndrome. Right Metacarpophalangeal (MCP) joints are extended, proximal interphalangeal (PIP) and distal interphalangeal (DIP) joints are flexed. Weakness in thumb abduction and opposition.

The preferred term is neuralgic amyotrophy (NA)³ but brachial neuritis, and acute brachial neuropathy⁴ have also been used. It most commonly affects middle aged men and usually presents with sudden and severe unilateral upper limb pain followed by motor weakness and patchy sensory deficit.⁵ It may also affect the diaphragm, recurrent laryngeal nerve, lower limbs, or the shoulder.⁶, ⁷ The etiology is generally multifactorial and is associated with autoimmune causes (systemic lupus erythematosus, giant cell arteritis and polyarteritis nodosa) or viral infections-most recently SARS-CoV-2.8 Vaccinations (including Covid), Hepatitis E, surgery, or trauma may play a role. The diagnosis of NA is fundamentally clinical and based on a typical history and a focal neurologic exam. MRI and nerve conduction studies are helpful in confirming the diagnosis. If the condition is immune mediated, steroids and/or IVIG are efficacious especially when added to analgesics in patients in whom pain is the predominant feature. Most cases improve or resolve with time but there is a twenty-five percent chance of recurrence.

Author Contributions

All authors have reviewed the final manuscript prior to submission. All the authors have contributed significantly to the manuscript, per the International Committee of Medical Journal Editors criteria of authorship.

- Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; AND
- Drafting the work or revising it critically for important intellectual content; AND
- Final approval of the version to be published; AND
- Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Disclosures and conflicts of Interest

The authors have no conflicts of interest to disclose.

Corresponding Author

Dr. Michael E. Lazarus Professor of Clinical Medicine 757 Westwood Plaza, Suite 7501C, Los Angeles, CA 90095

Tel: 310-267-9645

Email: mlazarus@mednet.ucla.edu



This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CCBY-NC-4.0). View this license's legal deed at https://creativecommons.org/licenses/by-nc/4.0/legalcode for more information.

REFERENCES

- 1. Moriguchi K, Miyamoto K, Takada K, Kusunoki S. Four cases of anti-ganglioside antibody-positive neuralgic amyotrophy with good response to intravenous immunoglobulin infusion therapy. *J Neuroimmunol*. 2011;238(1-2):107-109. doi:10.1016/j.jneuroim.2011.08.005
- 2. Parsonage MJ, Turner JW. Neuralgic amyotrophy; the shoulder-girdle syndrome. *Lancet*. 1948;251(6513):973-978. doi:10.1016/s0140-6736(48)90611-4
- 3. IJspeert J, Janssen RMJ, van Alfen N. Neuralgic amyotrophy. *Curr Opin Neurol*. 2021;34(5):605-612. doi:10.1097/wco.00000000000000068
- 4. Tsairis P, Dyck PJ, Mulder DW. Natural history of brachial plexus neuropathy: report on 99 patients. *Arch Neurol*. 1972;27(2):109. doi:10.1001/archneur.1972.00490140013004

- 5. Feinberg JH, Radecki J. Parsonage-Turner Syndrome. *HSS Jrnl*. 2010;6(2):199-205. doi:10.1007/s11420-010-9176-x
- 6. van Alfen N, van Engelen BGM. The clinical spectrum of neuralgic amyotrophy in 246 cases. *Brain*. 2005;129(2):438-450. doi:10.1093/brain/awh722
- 7. England JD, Sumner AJ. Neuralgic amyotrophy: an increasingly diverse entity. *Muscle Nerve*. 1987;10(1):60-68. doi:10.1002/mus.880100112
- 8. Ameer MZ, Haiy AU, Bajwa MH, et al. Association of Parsonage–Turner syndrome with COVID-19 infection and vaccination: a systematic review. *J Int Med Res*. 2023;51(7):3000605231187939. doi:10.1177/03000605231187939