Common Peroneal Nerve and Tarsal Tunnel Release Surgery in an Adolescent Male with Hunter Syndrome: Illustrative Case

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ABSTRACT

BACKGROUND: Children with Hunter syndrome have a high prevalence of nerve compression syndromes given the buildup of glycosaminoglycans in the tendon sheaths and soft tissue structures. These are often comorbid with orthopedic conditions given joint and tendon contractures due to the same pathology. While carpal tunnel syndrome and surgical treatment has been well-reported in this population, the literature on lower extremity nerve compression syndromes and their treatment in Hunter syndrome is sparse.

OBSERVATIONS: We report the case of a 13-year-old male with a history of Hunter syndrome who presented with toe-walking and tenderness over the peroneal and tarsal tunnel areas. He underwent bilateral common peroneal nerve and tarsal tunnel releases, with findings of severe nerve compression and hypertrophied soft tissue structures demonstrating fibromuscular scarring on pathology. Post-operatively, the patient's family reported subjective improvement in lower extremity mobility and plantar flexion.

LESSONS: In this case, peroneal and tarsal nerve compression were diagnosed clinically and treated effectively with surgical release and postoperative ankle casting. Given the wide differential of common comorbid orthopedic conditions in Hunter syndrome and the lack of validated electrodiagnostic normative values in this population, the history and physical examination and consideration of nerve compression syndromes are tantamount for successful workup and treatment of gait abnormalities in the child with Hunter syndrome.

KEYWORDS: Hunter syndrome, MPS II, tarsal tunnel release, peroneal nerve release

INTRODUCTION

Hunter syndrome, or mucopolysaccharidosis type II (MPS II), is a rare, X-linked inherited disorder characterized by a lack of lysosomal iduronate-2-sulfatase enzyme that results in an intracellular and extracellular accumulation of glycosaminoglycans (GAGs).[1] This buildup can damage organs and tissues throughout the body, including peripheral nerves, due to soft tissue becoming "viscous" and fascia becoming thickened.[2] Nerve compression syndromes are highly prevalent among patients with Hunter syndrome, with up to 95% of patients reported to eventually develop carpal tunnel syndrome (CTS).[3] While the literature on CTS and its surgical release in Hunter syndrome is well established,[4–9] nerve compression syndromes of the lower extremity are minimally reported. We present an illustrative case of the diagnosis and surgical treatment peroneal and tibial nerve compression syndromes with release of the common peroneal nerve and tarsal tunnel in the bilateral lower extremities.

ILLUSTRATIVE CASE

The patient is a 13-year-old male with a history of Hunter Syndrome who presented to the plastic surgery office with progressive toe-walking in an externally-rotated gait, and tenderness over the bilateral peroneal and tarsal areas. He had a strong nerve compression history, with bilateral carpal tunnel releases performed at age three, requiring repeat releases at age 11 accompanied at that time by bilateral pronator releases as well as right and middle finger trigger releases. Additional history included reactive airway disease, and intravenous and intrathecal enzyme therapy as part of a clinical trial.

On physical examination, the patient demonstrated a toe-walking gait (**Video 1**). He was only able to descend onto his heels if externally rotating his legs and then gradually internally rotating them until his toes pointed forward.

Video 1. Pre-operative gait [https://vimeo.com/915299525]

Tenderness to palpation was elicited on the outside of the leg just below the knee in the common peroneal nerve area, and on the inner ankle over the tarsal tunnel area. He held his ankles in extension, but they were able to be flexed into a neutral position, which was thought to imply that the Achilles tendons did not need to be lengthened.

Consideration of clinical factors led to the hypothesis that peroneal nerve compression was affecting ankle extension, and that tarsal tunnel compression was causing plantar nerve irritation and pain, culminating in it being easier and less painful to walk on his toes instead of the soles of his feet. Given his robust positive response to prior nerve decompression in the upper extremities and the natural history of GAG buildup in the soft tissues, peroneal nerve and tarsal tunnel release surgeries were offered. Electrodiagnostic studies for further workup were discussed but forgone, given the strong history and physical exam findings, as well as the lack of known normative electrodiagnostic values for the lower extremity in children, let alone children with Hunter syndrome. Second opinions also offered the options of Achilles tendon lengthening and plantar fasciotomy. Ultimately, the family chose to pursue nerve decompression with post-operative casting of ankles in a neutral position.

The patient was taken to the operating room where general anesthesia was induced using a laryngeal mask airway and fiber-optic bronchoscopy confirmation given a history of difficult intubation due to macroglossia and cervical spine immobility, as well as reactive airway disease. A local direct subcutaneous block over the four incision sites was administered. Esmarch exsanguination followed by tourniquet inflation to 225 mmHg was performed.

For the common peroneal nerve releases, a curvilinear incision was made using a #15 scalpel blade on the lateral portion of the left lower extremity. The fat was dissected from the muscle using tenotomy scissors. The nerve was not readily palpable. A linear incision was made in the fascia covering the extensor digitorum longus and peroneus longus, which was noted to be extremely thickened. The nerve was identified and noted to be extremely flattened. It was dissected proximally and distally. The anterior and posterior crural intermuscular septae, the deep tendinous fascia, and the innominate intermuscular septum were incised and removed until the nerve appeared free. By the end of this decompression, the intrinsic blood vessels in the nerve became more visible as blood was able to fill them more freely as compression was released (**Figure 1**).

Figure 1. Peroneal nerve beneath the peroneus fascia

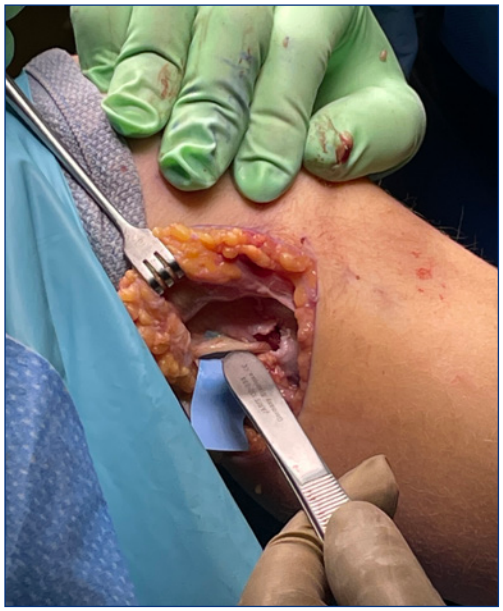
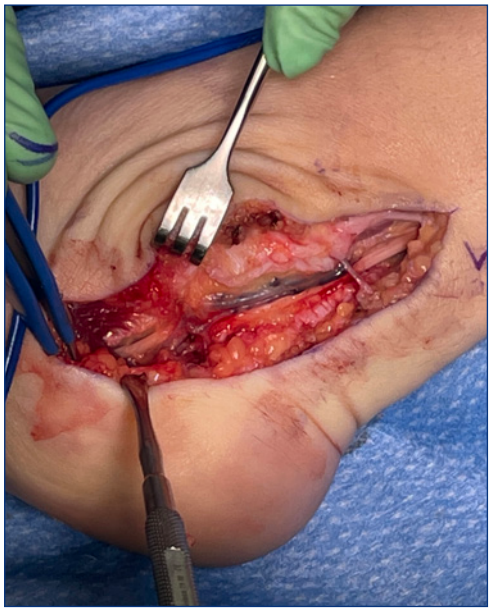


Figure 2. Tibial nerve in the tarsal tunnel



For the tarsal tunnel releases, an incision was made with a #15 blade scalpel posterior to and along the path of the posterior tibial nerve. Blunt dissection was used to preserve longitudinal crossing nerve branches. Vessels were cauterized with bipolar cautery or clipped. The nerve was identified proximal to the flexor retinaculum, and the retinaculum was divided over the nerve. The nerve was significantly flattened and pearly white/avascular appearing (**Figure 2**). In the left tarsal tunnel, inflammatory fluid was noted within the tunnel and the flexor retinaculum was severely tight. The abductor hallucis was identified. There was notable synovitis in the tarsal tunnel as well as significant compression at the foot along the undersurface of the abductor hallicus. Its fascia was divided; the muscle was retracted; and deep fascia was divided. The scissor was inserted into the medial and lateral plantar canals and the roofs were opened. The septum between the nerves was divided to create a large tunnel.

The wounds were closed with deep dermal 3-0 monocryl interrupted stitches, and a running subcuticular 4-0 monocryl for dermal and dermal-epidermal layers. Steristrips were then placed on the skin. The ankles were placed in a neutral position and casted using synthetic casting material. Post-operatively, the patient was admitted to a floor level of care for one night of observation. At his two-week follow up appointment, he was able to ambulate with improved plantarflexion compared to baseline (**Video 2**). Histopathology of the hypertrophied fascia demonstrated fibromuscular tissue scarring. The patient has since received routine physical therapy services.

Video 2. Post-operative gait [https://vimeo.com/915297064]

DISCUSSION

Observations

We report the case of a 13-year-old male with Hunter syndrome who developed progressive toe-walking and tenderness over the peroneal and tarsal areas, successfully treated with bilateral common peroneal nerve and tarsal tunnel releases, during which severe nerve compression and hypertrophied soft tissue structures were encountered. Several orthopedic manifestations have been identified in patients with Hunter syndrome.[10,11] However, conditions such as tarsal tunnel syndrome are infrequently reported in this patient population.[12]

Lessons

Musculoskeletal manifestations of Hunter syndrome primarily consist of spinal conditions (e.g., cervical stenosis), hip dysplasia, and, most commonly, carpal tunnel syndrome (CTS).[12,13] While CTS is highly prevalent in other MPS subtypes (e.g., Hurler, Maroteaux-Lamy syndrome), other nerve entrapment syndromes, such as tarsal tunnel syndrome (TTS), occur less commonly in among these patients.[12]

In their 2019 case series of 19 MPS patients, Williams et al report mixed results in the diagnosis and post-operative outcomes of four patients (two with MPS I, two with MPS VI) with a suspected diagnosis of TTS.[12] One patient with MPS VI was clinically diagnosed with TTS and experienced symptom relief following bilateral tarsal tunnel decompressions. Two patients had inconclusive nerve conduction studies but had resolution of lower extremity pain following tarsal tunnel decompression for one patient (MPS VI) and spinal fusion for the other (MPS I). The final patient with MPS I had inconclusive lower extremity nerve conduction studies but underwent tarsal tunnel release with subsequent pain resolution post-operatively. These findings indicate mixed results in the clinical and electromyographical diagnosis of patients with MPS, and variable yet promising results following nerve decompression surgery.

In this case, peroneal and tarsal nerve compression were diagnosed clinically and treated effectively with surgical release and postoperative ankle casting. Given the wide differential of common comorbid orthopedic conditions in Hunter syndrome and the lack of validated electrodiagnostic normative values in this population, the history and physical examination and consideration of nerve compression syndromes are valuable for successful workup and treatment of gait abnormalities in a child with Hunter syndrome. Furthermore, communication barriers in patients with Hunter syndrome may limit symptom reporting, and subtle signs of nerve compression may be overlooked by higher acuity medical conditions, thereby putting this population at increased risk of more advanced disease at the time of diagnosis.[3]

Limitations

This is a single case report of a rare disease case and therefore does not intend to provide a high level of evidence to inform treatment decisions.

CONCLUSION

In this case of a 13-year-old male with Hunter syndrome, peroneal and tarsal nerve compression were diagnosed clinically and treated effectively with surgical release and postoperative ankle casting. Given the wide differential of common comorbid orthopedic conditions in Hunter syndrome and the lack of validated electrodiagnostic normative values in this population, the history and physical examination and consideration of nerve compression syndromes are tantamount for successful workup and treatment of gait abnormalities in the child with Hunter syndrome.

References

1. Hashmi MS, Gupta V. Mucopolysaccharidosis Type II. In: *StatPearls*. StatPearls Publishing; 2023. Accessed July 18, 2023. http://www.ncbi.nlm.nih.gov/books/NBK560829/
2. Stapleton M, Kubaski F, Mason RW, et al. Presentation and Treatments for Mucopolysaccharidosis Type II (MPS II; Hunter Syndrome). *Expert Opin Orphan Drugs*. 2017;5(4):295-307. doi:10.1080/21678707.2017.1296761
3. Argenta AE, Davit A. Carpal Tunnel Syndrome in the Setting of Mucopolysaccharidosis II (Hunter Syndrome). *Plast Reconstr Surg Glob Open*. 2017;5(8):e1477. doi:10.1097/GOX.0000000000001477
4. Wraith JE, Alani SM. Carpal tunnel syndrome in the mucopolysaccharidoses and related disorders. *Arch Dis Child*. 1990;65(9):962-963. doi:10.1136/adc.65.9.962
5. Norman-Taylor F, Fixsen JA, Sharrard WJ. Hunter's syndrome as a cause of childhood carpal tunnel syndrome: a report of three cases. *J Pediatr Orthop B*. 1995;4(1):106-109. doi:10.1097/01202412-199504010-00018
6. Haddad FS, Jones DH, Vellodi A, Kane N, Pitt MC. Carpal tunnel syndrome in the mucopolysaccharidoses and mucolipidoses. *J Bone Joint Surg Br*. 1997;79(4):576-582. doi:10.1302/0301-620x.79b4.7547
7. Van Heest AE, House J, Krivit W, Walker K. Surgical treatment of carpal tunnel syndrome and trigger digits in children with mucopolysaccharide storage disorders. *J Hand Surg Am*. 1998;23(2):236-243. doi:10.1016/S0363-5023(98)80120-2

8. Yuen A, Dowling G, Johnstone B, Kornberg A, Coombs C. Carpal tunnel syndrome in children with mucopolysaccaridoses. *J Child Neurol*. 2007;22(3):260-263. doi:10.1177/0883073807300528
9. Jadhav T, Kornberg AJ, Peters H, Lee J, Ryan MM. Carpal tunnel syndrome in pediatric mucopolysaccharidoses. *Clinical Neurophysiology*. 2014;125(4):e4-e5. doi:10.1016/j.clinph.2013.10.033
10. Link B, de Camargo Pinto LL, Giugliani R, et al. Orthopedic manifestations in patients with mucopolysaccharidosis type II (Hunter syndrome) enrolled in the Hunter Outcome Survey. *Orthop Rev (Pavia)*. 2010;2(2):e16. doi:10.4081/or.2010.e16
11. Link B, Botha J, Giugliani R. Characterization of orthopedic manifestations in patients with mucopolysaccharidosis II using data from 15 years of the Hunter Outcome Survey. *JIMD Rep*. 2023;65(1):17-24. doi:10.1002/jmd2.12401
12. Williams N, Willet J, Clark D, Ketteridge D. Tarsal tunnel syndrome in the mucopolysaccharidoses: A case series and literature review. *JIMD Rep*. 2019;46(1):16-22. doi:10.1002/jmd2.12021
13. Stepien KM, Bentley A, Chen C, et al. Non-cardiac Manifestations in Adult Patients with Mucopolysaccharidosis. *Frontiers in Cardiovascular Medicine*. 2022;9. Accessed July 24, 2023. https://www.frontiersin.org/articles/10.3389/fcvm.2022.839391

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