

Review: Magnetic Resonance Imaging of Muscle Denervation Syndromes

Gustav Andreisek, M.D., MBA^{1,2}; Avneesh Chhabra, M.D.³

¹ Radiology, Spital Thurgau, Münsterlingen, Switzerland

² University of Zurich, Zurich, Switzerland

³ Musculoskeletal Radiology, University of Texas Southwestern Medical Center, Dallas, TX, USA

Introduction

Muscles can be affected by a broad variety of pathologies, one of which is muscle denervation syndrome. As the term indicates, muscular denervation syndrome is caused by an underlying neurogenic disorder. There are a broad variety of neurogenic disorders that can cause these muscle abnormalities clinically and on imaging. The list of pathologies includes, but is not exclusively limited to [1]:

1. Systemic nerve diseases

e.g. ischemia, vasculitis, toxic, endocrine and metabolic disorders [2], hereditary motor-sensory neuropathies, amyloidosis, hyperlipidemia, acute and chronic demyelinating inflammatory neuropathies, etc.

2. Local conditions

e.g. plexopathy, nerve injury, perineural or intraneuronal compressive lesions, adhesive neuropathy after surgery, infections and nerve sheath tumors, etc.

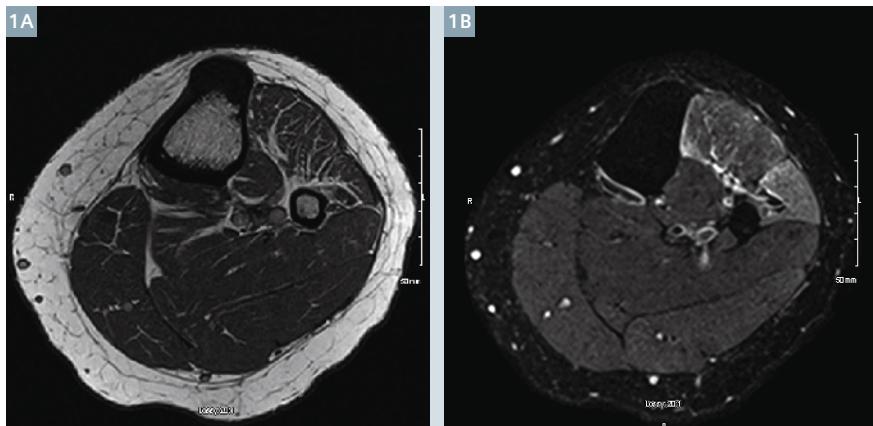
3. Neuropathies related to functional anatomical changes
e.g. habitual leg crossing, repeated typing, obesity and repetitive exercise, which may cause traction mononeuropathy or compressive neuropathy in functional compartment syndromes.

Muscle abnormalities as depicted on magnetic resonance imaging

Neurogenic magnetic resonance (MR) imaging abnormalities of skeletal muscles are related to time course of development. In the early phase, increased muscle signal on fat-suppressed T2-weighted or STIR images is the main imaging finding. It is hypothesized to be related to increased intramuscular fluid, congestion and/or relative shift of intra- and extra-compartmental fluid due to factors, such as altered muscle activity or inactivity. Subsequently, over time, it progresses to muscle atrophy and fatty infiltration with associated increased signal on T1-weighted images (Fig. 1).

In late stages, there might or might not be an increased signal on fat-suppressed T2-weighted or STIR images, and the predominant imaging finding is muscle atrophy characterized by increased signal on T1-weighted images associated with loss of muscle volume. MR imaging has a high sensitivity for the detection of increased muscle signal on fat-suppressed T2-weighted or STIR images and the changes appear earlier than on US or CT imaging.

Increased signal on fat-suppressed T2-weighted or STIR images of muscles is also seen with a number of other muscle disorders. The list of possible pathologies causing increased T2 signal is broad and includes trauma, early myositis ossificans, inflammatory myopathies such as dermatomyositis, polymyositis, eosinophilic myositis, proliferative myositis, or myositis associated with connective tissue diseases, infectious myositis, infiltrating neoplasm, rhabdomyolysis, muscle infarction, sickle cell disease, or overuse syndromes. Whereas some differential diagnosis can be excluded based on clinical examination or labo-



1 Axial images of a 36-year-old man with traumatic common peroneal nerve injury. MR images show a typical muscle denervation pattern in the subacute phase with increased signal and mild loss of bulk on T1-weighted (1A) and fat saturated T2-weighted (1B) images affecting the tibialis anterior, extensor digitorum and peroneal muscles.

ratory data, many of them may remain on the list of possible differential diagnosis. MR imaging is also highly sensitive but specificity may vary due to factors such as reader skill, technique and unavailable clinical history and findings at the time of interpretation. Therefore, it is important to understand the role of MR imaging in these conditions, as MRI cannot be the 100% problem-solving tool in all cases [3]. However, its real value is in excluding many other differential diagnoses, therefore supporting establishment of the main diagnosis [4]. This role is important as it can help in lowering healthcare costs by avoiding unnecessary exams or assessments as well as reducing the risk of a delayed diagnosis. In fact, this might become even more important in healthcare systems where ideology and planning exists to reimburse procedures based on their value (value-based healthcare model).

The role of MR imaging in neurogenic muscle disorders includes, but is not exclusively limited to [1]:

1. Systemic nerve diseases

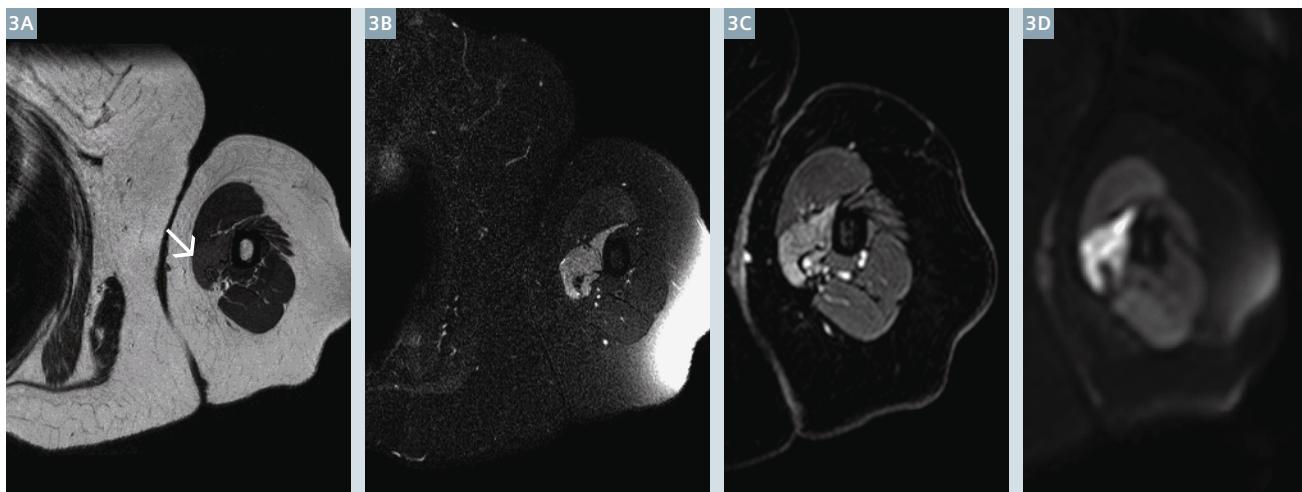
MR imaging is not expected to make these diagnoses alone. However, MRI may be used to confirm the clinical suspicion (based on symptoms, clinical exam, electrodiagnostic tests and laboratory findings) by demonstrating abnormality of the innervating and regional muscle denervation changes or, exclude any structural cause for neuropathies in these cases. The muscle denervation changes are limited to the territory of neuropathic nerves, are diffuse without intramuscular hemorrhage and there is no associated perimuscular or perifascial edema as could be conspicuous in myopathy cases.

2. Local conditions

MR imaging has a major role in these cases as it supplements information gained from the clinical exam and electrodiagnostic tests or provides information, which may not be possible to attain from other (imaging) modalities. Not only the muscle denervation change is visible but also the offending lesion, such as an intra- or perineural lesion (Fig. 2) or other tumors (Fig. 3), can be identified.



2 35-year-old man with physical signs of a winged scapula after motor vehicle accident. Coronal T1-weighted MR image shows atrophy of the right serratus anterior muscle compared to the normal contralateral left side (2A, arrows). The muscle is innervated by the long thoracic nerve which appears both, thickened on T1-weighted MR images (2B, arrow), and with increased signal intensity on fat suppressed T2-weighted MR images (2C). For comparison, corresponding MR images of the left normal long thoracic nerve are shown (2D, E). Please note that the long thoracic nerve directly arises from C5-7 without entering the brachial plexus. It does not innervate other muscles.



3 Axial MR images of a 46-year-old woman with clinical weakness of the coracobrachialis muscle (arrow). MR images show increased signal on T1-weighted (3A) and fat saturated T2-weighted (3B) MR images. However, there is also enhancement on postcontrast fat suppressed T1-weighted MR images as well as diffusion restriction on b=800 diffusion-weighted images raising the suspicion of a non-neurogenic disorder. In this case, MR-targeted muscle biopsy revealed muscle lymphoma.

3. Neuropathies related to functional anatomical changes

The role of MR imaging is limited in these circumstances. These disorders are mostly diagnosed on the basis of clinical and pressure catheter studies. Sometimes, ultrasound examinations are used to unveil dynamic nerve compression or dislocation. However, rarely, MRI may be used in clinically confounding cases with imaging performed before and after the exercise/effort in question. Indirect signs such as significant prolongation of T2 signal intensity within the affected muscle compartment may aid in the diagnosis of compartment syndrome.

Nerve disorders

The underlying cause for muscle denervation syndrome includes a broad variety of neurogenic diseases that can be divided according to the anatomic site of affliction (neuromuscular junction or nerve cell body, axon and myelin sheath) or functional involvement (sensory, motor or autonomic) [5].

Neuromuscular junction (NMJ) disorders are primarily related to acetylcholine receptor abnormalities. They can be familial/congenital, such as myasthenia gravis, or acquired, such

as drug induced, botulism or paraneoplastic (Eaton-Lambert syndrome). Whereas many neuromuscular disorders show predominant involvement of lower extremities, NMJ disorders present with facial and extraocular muscle weakness, however, with normal tendon reflexes and normal sensation. The latter is usually confirmed by electrodiagnostic tests showing normal electromyograms and normal nerve conduction velocities. By contrast, nerve diseases distal to the NMJ may present with motor weakness, sensory symptoms and autonomic symptoms depending upon the affected nerve and its functional role. In these cases, electrodiagnostic studies usually show pathologic electromyograms as well as delayed nerve conduction [1].

With predominant axonal involvement, distribution of involvement and clinical presentation are usually distal > proximal, and legs > arms. Nerve biopsy may be useful in axonal and myelin disorders. The pathologist usually classifies the histologic alterations as axonal neuropathy, demyelinating neuropathy, inflammatory neuropathy, or as a process of the supporting and/or vascular tissues, such as vasculitis, storage disease, and infectious or neoplastic infiltration. However, MR imaging findings of muscle denervation can be very

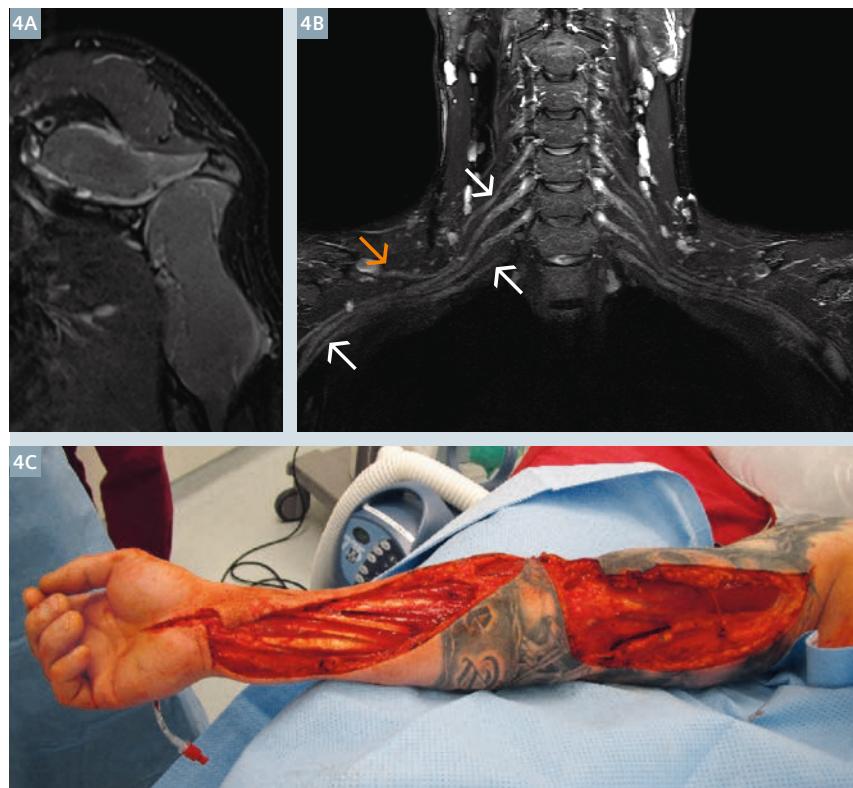
similar and should be correlated with clinical, available electrodiagnostic, and other clinical and/or laboratory information to establish the final diagnosis [6]. MR imaging, especially MR neurography, is particularly helpful in demonstrating the nerve lesions and localization of the pathology. The affected nerve shows most signal, fascicular and/or caliber alteration, at or immediately proximal to the site of compression or injury. The affected muscles often show enhancement on post gadolinium imaging but there is no significant restriction on diffusion-weighted imaging (DWI). MR neurography is also helpful in grading the nerve injury into stretch injury, neurroma in continuity and complete nerve discontinuity (Fig. 4).

The differential diagnosis of myopathies (muscular dystrophy/myositis) may result from infectious, endocrine, metabolic, autoimmune, myoglobinuria or familial causes, such as limb-girdle syndrome. Myopathies present with isolated motor symptoms and the distribution is usually proximal and symmetric with absent tendon reflexes in the involved muscles. Electrodiagnostic testing is usually pathologic with respect to the motor unit potential analysis (e.g. abnormal amplitude, duration, number of phases, and firing rate). In addition, serologic exams might show high creatine kinase levels

in myopathies. Muscle biopsy is often used to make the final diagnosis and the role of MR imaging is to show one or a combination of edema like signal intensity changes on fat suppressed T2-weighted MR images, fatty infiltration or atrophy depending upon the stage of the disease, similar to denervation muscle changes. However, the involved muscles may not correspond to single nerve territory and regional nerves will show normal signal intensity, thereby excluding neuropathy as the cause of muscle findings.

Patchy involvement, multicompartiment affliction, perimuscular fascial edema and patchy intramuscular and/or fascial enhancement may be seen with myositis, which is absent with muscle denervation changes. On DWI, the presence of a mass lesion can show diffusion restriction (Fig. 3). In myopathy cases, the nerves show normal signal and MR imaging appearance.

In conclusion, MR imaging in muscle denervation syndromes is very valuable in either establishing the diagnosis, ruling out related pathologies, or a combination of both.



4 MR image of a 32-year-old man after a high-voltage accident and with palsy of the right arm. MR neurography shows increased T2-signal of supraspinatus and infraspinatus muscles of the right shoulder (**4A**) as well as mild thickening and hyperintensity of the right brachial plexus (white arrows) and suprascapular nerve (orange arrow) on coronal STIR SPACE image (**4B**) in keeping with traction injury with possible ischemia. Subsequently, muscle compartment arm and forearm decompression surgery was performed (**4C**).

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Avneesh
Chhabra



Gustav
Andreisek

Contact

PD Dr. Gustav Andreisek, M.D., SCMR
Executive MBA HSG
Head of Radiology
Kantonsspital Münsterlingen
Spitalcampus 1
8596 Münsterlingen
Switzerland
Phone: +41 71 686 23 30
Fax +41 71 686 26 74
gustav@andreisek.de