Skeletal Radiology (2019) 48:1451–1456
https://doi.org/10.1007/s00256-019-03174-4

CASE REPORT

Hourglass-like constriction neuropathy of the suprascapular nerve detected by high-resolution magnetic resonance neurography: report of three patients

Du Hwan Kim[1] · Jaelim Kim[2] · Duk Hyun Sung[2]

Received: 7 December 2018 / Revised: 26 January 2019 / Accepted: 28 January 2019 / Published online: 12 February 2019

Abstract
Hourglass-like constriction neuropathy is a neurological condition caused by non-traumatic, non-compressive fascicular constrictions of one or more individual peripheral nerves. Based on clinical manifestations, it is very difficult to differentiate hourglass-like constriction neuropathy from idiopathic neuralgic amyotrophy. Focal hourglass-like constriction neuropathy may be erroneously diagnosed as idiopathic neuralgic amyotrophy. Previous studies demonstrated hourglass-like constrictions of peripheral nerves in the surgical exploration of patients diagnosed with idiopathic neuralgic amyotrophy. Recently, high-resolution ultrasound or magnetic resonance neurography (MRN) have been introduced to detect focal hourglass-like constrictions of peripheral nerves in spontaneous nerve palsy. We present a series of three cases in which the suprascapular nerve was affected by hourglass-like constrictions, which were visualized by high-solution MRN, including a nerve-selective morphological MR pulse sequence with strong fat- and water-signal suppression.

Keywords Brachial plexus · Suprascapular nerve · Hourglass-like constriction · Neuralgic amyotrophy

Introduction

Non-traumatic acute unilateral shoulder weakness is frequently encountered in musculoskeletal clinics. Differential diagnostic approaches to this condition are important for treatment. Possible diagnoses include C5 or C6 cervical radiculopathy caused by a herniated disc or foraminal stenosis, full-thickness rotator cuff tear, space-occupying masses compressing the upper brachial plexus, idiopathic neuralgic amyotrophy, hourglass-like constriction neuropathy, brachial plexitis of various etiologies, and others [1, 2].

The classic phenotype of idiopathic neuralgic amyotrophy is acute severe shoulder girdle pain followed by shoulder weakness [1]. Before the era of high-resolution magnetic resonance neurography (MRN), when diagnostic evaluations such as cervical spine and shoulder magnetic resonance imaging (MRI) failed to explain the cause of shoulder weakness, clinicians tended to diagnose idiopathic neuralgic amyotrophy as the cause of shoulder weakness [2].

Hourglass-like constriction neuropathy is a neurological condition caused by non-traumatic, non-compressive fascicular constrictions of one or more individual peripheral nerves [2, 3]. Recently, various imaging modalities such as high-resolution ultrasound or MRN have been introduced to detect focal hourglass-like constrictions of peripheral nerves in nerve palsy [4–6]. In particular, the bullseye sign of the nerve on MRI, manifested as peripheral hyperintensity and central hypointensity on fat-suppressed imaging, is suggestive of an association with focal hourglass-like constrictions [6]. We present a series of three cases in which the suprascapular nerve was affected by hourglass-like constrictions, which were visualized by high-resolution MRN including a nerve-selective morphological MR pulse sequence with strong fat- and water-signal suppression, and describe the results of the non-surgical treatment of these patients. This study was approved by our institutional review board. Informed consent was obtained from all individuals included in the study.

✉ Duk Hyun Sung
yays.sung@samsung.com

[1] Department of Rehabilitation Medicine, Dongsan Medical Center, School of Medicine, Keimyung University, Daegu, South Korea

[2] Department of Physical and Rehabilitation Medicine, Samsung Medical Center, Sungkyunkwan University School of Medicine, 81 Irwon-Ro, Gangnam-gu, Seoul 06351, South Korea

Case report

Table 1 summarizes the clinical findings of the three cases.

Case 1

A 26-year-old right-handed woman presented with a 5-month history of scapular pain and difficulty in elevating the left arm over the head. Ten days before the development of weakness, she experienced flu-like symptoms for 3 days followed by extremely severe pain (numeric rating scale [NRS] 10) in the shoulder girdle area. As the pain decreased over 10 days, she noticed her shoulder weakness.

Muscle strength tests revealed grade III in the shoulder abductor and external rotator and grade V in the elbow flexor and wrist extensor. There were no abnormalities in sensory tests. Electromyography showed denervation of the supraspinatus and infraspinatus.

At 3 months after the onset, she visited another hospital. Cervical spine MRI (Fig. 1a) revealed no disc herniation or foraminal stenosis, and shoulder MRI (Fig. 1b) demonstrated increased signal intensity of the supraspinatus and infraspinatus. At that time, she was presumptively diagnosed with idiopathic neuralgic amyotrophy. At 5 months after the onset, she visited our clinic and underwent our standard high-solution MRN protocol, which included a nerve-selective morphological MR pulse sequence with strong fat- and water-signal suppression (nerve-SHeath signal increased with INKed rest-tissue RARE Imaging [SHINKEI]) [7]. MRN revealed three focal constrictions of the suprascapular nerve without enlargement or signal change (Fig. 1c, d). Laboratory tests, including cerebrospinal fluid analysis, anti-nuclear autoantibody, and anti-ganglioside antibodies, were normal. Thus, she was diagnosed with hourglass-like constriction neuropathy of the suprascapular nerve rather than idiopathic neuralgic amyotrophy. She was treated with oral steroid (prednisolone 10 mg for 14 days) and ultrasound-guided perineural injection (triamcinolone acetonide 20 mg) once. At 10 months after the onset, muscle strength improved to near normal.

Case 2

A 42-year-old right-handed man presented with a 3-month history of left-shoulder weakness. At the onset, he

Table 1 Clinical findings in three patients with hourglass-like constriction neuropathy

	Case 1	Case 2	Case 3
Sex/age	F/26	M/42	M/52
Side	Left	Left	Left
Involved nerves	Suprascapular	Suprascapular	Suprascapular, radial
Combined cervical spine problems on MRI	None	C3–4, C5–6 disc protrusion disc	C5–6 disc protrusion
Precipitating factors	Flu-like symptoms	C5–6 total disc replacement	Cervical spine interventions
Initial pain (NRS score)	10	9	5
Interval between pain and palsy	10 days	20 days	14 days
Atrophy	Supraspinatus, infraspinatus	Supraspinatus, infraspinatus	Supraspinatus, infraspinatus, brachioradialis, wrist extensors
Muscle strength at MRN	Shoulder external rotator, III; shoulder abductor, III	Shoulder external rotator, grade II; shoulder abductor, grade IV	Shoulder external rotator, grade II; wrist extensor, grade 0
Interval between onset and MRN	5 months	3 months	7 weeks
Number of constrictions	Suprascapular nerve, 3	Suprascapular nerve, 1	Suprascapular nerve, 2; radial nerve, 5
Ancillary tests	CSF, neck CT, chest CT, abdominal CT, autoantibodies—all normal	Autoantibodies—normal	Autoantibodies—normal
Treatment	Oral steroid, local steroid injection	Intravenous steroid	Intravenous steroid
Follow-up	Near normal at 10 months after the onset	Near normal at 15 months after the onset	Shoulder weakness near normal at 5 months after the onset Wrist extensor weakness not improved at 5 months after the onset Wrist extensor weakness not improved at 5 months after the onset

MRI magnetic resonance imaging, *NRS* numeric rating scale, *MRN* magnetic resonance neurography, *CSF* cerebrospinal fluid, *CT* computed tomography

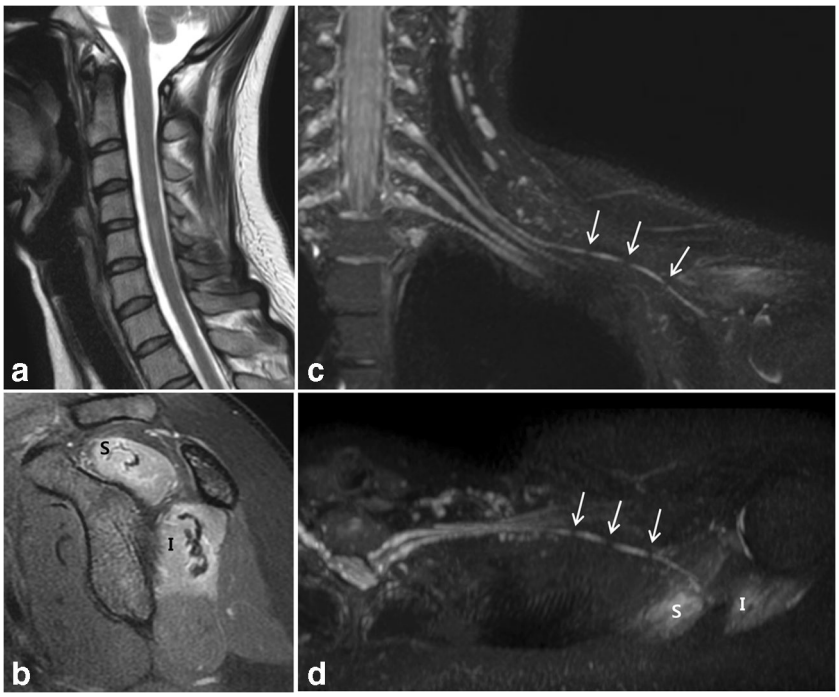

Fig. 1 Case 1. **a** No evidence for cervical disc herniation or foraminal stenosis. **b** Sagittal 2D T2-weighted fat suppression image demonstrates denervation-related edematous changes in the supraspinatus (*S*) and infraspinatus (*I*) muscles. **c** Curved reformatted coronal 3D high resolution magnetic resonance neurography (MRN) including the nerve-SHeath signal increased with INKed rest-tissue RARE Imaging (SHINKEI) sequence reveals three focal constrictions (*arrows*) of the suprascapular nerve. **d** Curved reformatted axial 3D high-resolution MRN including SHINKEI sequence reveals three focal constrictions (*arrows*) of the suprascapular nerve with denervation-related edematous changes in the S and I muscles

experienced sudden-onset severe pain (NRS 9) over the scapular and deltoid muscle areas and was not aware of weakness. He underwent cervical spine MRI, which revealed C3–4 and C5–6 (central to left) disc herniation. Because of severe pain, he received anterior discectomy of the C5–6 intervertebral disc and artificial disc replacement 10 days after the onset. Immediately after surgery, his pain was much relieved. But 10 days later, left shoulder pain in the deltoid muscle area redeveloped and left shoulder weakness developed de novo. At 4 months after the onset of the weakness, he visited our clinic owing to poor recovery of shoulder weakness. Muscle strength tests revealed grade II in the shoulder external rotator and grade IV in the shoulder abductor and flexor. Electromyography showed denervation of the supraspinatus and infraspinatus. High-resolution MRN including the SHINKEI sequence revealed focal constriction of the suprascapular nerve, suggesting hourglass-like constriction neuropathy (Fig. 2). He was treated with intravenous steroid (methylprednisolone 500 mg for 3 days) and tapering of oral prednisolone. His shoulder pain disappeared immediately after administration of intravenous steroid. Shoulder girdle muscle strength improved from 6 months after steroid treatment to near normal at 15 months after the onset.

Case 3

A 52-year-old right-handed man presented with left shoulder weakness and left wrist drop. Nine weeks earlier, he had experienced neck pain (NRS 5) and a tingling sensation in the left thumb. Seven weeks ago, he visited another hospital and underwent cervical MRI, which demonstrated left C5–6 disc protrusion. He received cervical spine procedures including cervical epidural steroid injection, C5–7

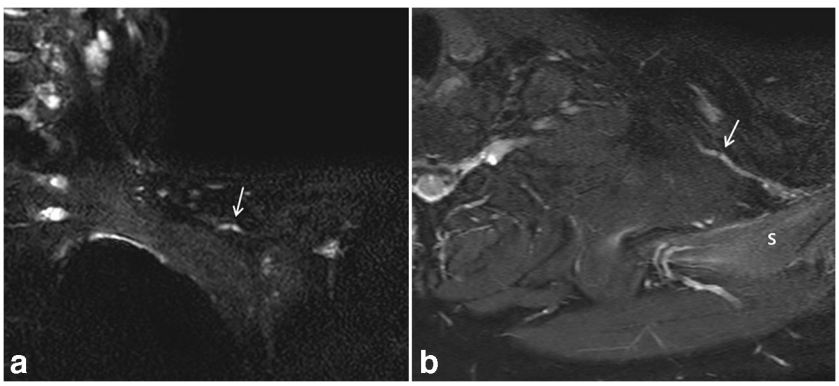

Fig. 2 Case 2. **a**, **b** High-resolution magnetic resonance neurography including the SHINKEI sequence reveals one focal constriction (*arrow*) of the suprascapular nerve, suggesting hourglass-like constriction neuropathy

medial branch block, and suprascapular and axillary nerve block. About 3 h after the procedures, he noticed left wrist drop. Four weeks earlier, he had recognized left shoulder girdle muscle atrophy. On physical examination, left upper extremity muscle strength tests revealed grade II in the shoulder external rotator; grade IV in the shoulder flexor; and grade 0 in the wrist extensor and finger extensor. Pinprick and touch senses over the left first dorsal web space were decreased. Electromyography showed denervation potentials of the suprascapular and radial nerve–innervated muscles except the triceps brachii. High-resolution MRN of the brachial plexus including the SHINKEI sequence revealed diffuse swelling of the C6 nerve with increased signal intensity and two focal constrictions of the suprascapular nerve (Fig. 3a, b) and T2-weighted fat-suppressed images of arm MRI showed two twisting constrictions with diffuse swelling of the radial nerve in the radial groove and three other constrictions with diffuse swelling of the radial nerve in the proximal forearm (Fig. 3c–f), suggesting hourglass-like constriction neuropathy of the suprascapular and radial nerves. He was treated with intravenous steroid (methylprednisolone 500 mg for 3 days). Three months after steroid treatment, the muscle strength of the shoulder girdle had returned to normal. However, no improvement in strength was noted in radial nerve–innervated muscles.

Fig. 3 Case 3. **a**, **b** High-resolution magnetic resonance neurography of the brachial plexus including the SHINKEI sequence reveals diffuse swelling of the C6 nerve (*arrowheads*) with increased signal intensity and two focal constrictions (*arrows*) of the suprascapular nerve. **c**, **d** Upper-arm MRI showed two twisting constrictions (*arrows*) with diffuse swelling of the radial nerve in the radial groove. **e**, **f** Forearm MRI showed three other constrictions (*arrows*) with diffuse swelling of the radial nerve in the proximal forearm

Discussion

We report three cases of hourglass-like constriction neuropathy, including two cases of isolated suprascapular nerve lesions and one case of suprascapular and radial nerve lesions. Hourglass-like constriction neuropathy around the shoulder region, which is usually confirmed by exploratory surgery, is relatively less frequently reported than that of the elbow region [2, 3, 8, 9]. The classic manifestation of idiopathic neuralgic amyotrophy is severe pain followed by weakness in single or multiple nerves [1, 10]. Idiopathic neuralgic amyotrophy is a clinical diagnosis based on clinical symptoms, imaging findings suggestive of the absence of compressive lesions, and electrodiagnostic testing. Differential diagnosis between hourglass-like constriction neuropathy and idiopathic neuralgic amyotrophy based on clinical findings seems to be nearly impossible because both diseases share common clinical manifestations [2]. In 2000, Yamamoto et al. reported that a patient diagnosed with idiopathic neuralgic amyotrophy had multiple hourglass-like constrictions in the anterior interosseous nerve in surgical exploration [11]. If hourglass-like constriction neuropathy were not taken into consideration, our three cases would be erroneously diagnosed as idiopathic neuralgic amyotrophy. However, high-resolution MRN revealed hourglass-like constrictions of individual nerves. In a clinical setting with severe pain followed by weakness in an upper extremity, high-resolution MRN of the brachial plexus and extremity may be helpful in differentiating hourglass-like constriction neuropathy from idiopathic neuralgic amyotrophy or other mimicking causes.

We expect that high-resolution imaging modalities may alter the concept or classification of idiopathic neuralgic amyotrophy. Conventional MRI of the cervical spine, shoulder, and brachial plexus is frequently performed in patients with non-traumatic acute unilateral shoulder weakness to find compressive lesions such as a herniated disc, foraminal stenosis, or paralabral ganglion cyst, and to visualize the patterns of denervation-related muscle edema or signal changes in the brachial plexus [12]. Before the introduction of high-resolution MRN, if compressive lesions were not found and conventional brachial plexus MRN demonstrated a hyperintense signal in the upper trunk, the diagnosis of classic-type idiopathic neuralgic amyotrophy tended to be made [12]. In 2006, van Alfen et al. reported various clinical spectra of idiopathic neuralgic amyotrophy in 199 cases [10]. Fifty patients underwent brachial plexus MRI, yielding focal T2 hyperintensities of the brachial plexus in three patients and no cases of hourglass-like constriction neuropathy [10]. We speculate that if high-resolution MRN had been applied in this population, several cases might have been proven to be hourglass-like constriction neuropathy. In non-traumatic acute unilateral shoulder weakness, high-resolution MRN should be performed to determine the presence of hourglass-like constriction neuropathy if there is no evidence for compressive C5 or C6 radiculopathy or space-occupying masses compressing individual nerves.

In these cases, we adopted high-resolution MRN including the SHINKEI sequence. This sequence includes two parts: a fat suppression pre-pulse and the improved motion sensitized driven equilibrium (iMSDE) pre-pulse to suppress signal from vessels, followed by a readout section with a 3D tissue-specific variable refocusing a flip-angle rapid acquisition with relaxation enhancement (RARE) sequence to acquire contrast-efficient T2-weighting [13]. This sequence can yield high-resolution and volumetric neurographic images by suppressing both fat and vessel signals to detect torsion sites along the longitudinal axis of an individual branch from the brachial plexus [7, 13]. However, this technique is not popular because its acquisition time is longer than that of fat suppression using spectral adiabatic inversion recovery alone.

The diagnostic yields of high-resolution MRN in hourglass-like constriction neuropathy around the brachial plexus have not been determined. In 2009, Vigasio and Marcoccio reported that preoperative differential diagnosis between hourglass-like constriction neuropathy and idiopathic neuralgic amyotrophy is unreliable [9]. Until recently, hourglass-like constriction neuropathy had to be surgically confirmed [2, 3]. In the future, we expect high-resolution MRN to be helpful in the diagnosis of hourglass-like constriction neuropathy and in deciding the methods of treatment and the surgical approach if needed.

The clinical implications for the use of high-resolution MRN to ascertain the presence of hourglass-like constrictions are not yet clear. The bullseye sign of the nerve on MRI refers to the finding of peripheral hyperintensity and central hypointensity on fat-suppressed imaging, perpendicular to the direction of the nerve [6]. This sign may reflect focal edematous swelling of the nerve proximal to the constriction sites. It is unclear whether the bullseye sign is pathognomonic for hourglass-like constrictions. Sneag et al. reported that a bullseye sign of the nerve was identified immediately proximal to 21 out of 23 constriction sites in 6 patients [6]. However, 2 of 3 patients with suprascapular nerve involvement did not reveal the bullseye sign [6]. Recent research by Sneag et al. also reported that although brachial plexus MRI was revealed to be normal in 24 out of 27 patients with idiopathic neuralgic amyotrophy, focal severe caliber decreases, suggestive of focal intrinsic constriction, were detected in 32 out of 38 nerves [14]. Our three cases with hourglass-like constriction neuropathy of suprascapular nerve did not demonstrate any obvious bullseye sign. We suggest that the sensitivity of the bullseye sign might be related to the caliber or obliquity of individual nerves.

The natural history of hourglass-like constriction neuropathy is unclear. It is known that the paralysis in hourglass-like constriction neuropathy is complete and permanent [3, 15]. However, our three patients with hourglass-like constriction neuropathy of suprascapular nerve nearly recovered after non-surgical treatments. Unfortunately, we did not perform follow-up MRN to elucidate whether hourglass-like constrictions resolved spontaneously. Also, it is unclear whether the patterns (numbers, extent, and severity) of constrictions are related to clinical severity at onset and recovery, and the exact mechanism of hourglass-like constriction neuropathy also remains unclear. Future research is necessary to elucidate the diagnostic value and clinical implications of high-resolution MRN in hourglass-like constriction neuropathy.

In conclusion, although hourglass-like constriction neuropathy is rare, it should be considered a potential cause of spontaneous nerve palsy, especially unilateral shoulder weakness. Because hourglass-like constriction neuropathy clinically mimics idiopathic neuralgic amyotrophy, high-resolution MRN should be performed to differentiate hourglass-like constriction neuropathy from idiopathic neuralgic amyotrophy.

Compliance with ethical standards

Ethics approval All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional review board.

Disclosure We did not receive any financial support or other benefits from commercial sources for the work reported in this manuscript, nor did any of the authors have any financial interests with regard to the work that may create a potential conflict of interest or the appearance of one. This work has not been previously presented, submitted, or published in any form or any language.

Conflicts of interest The authors declare that they have no conflicts of interest.

References

1. Van Alfen N, van Eijk JJ, Ennik T, et al. Incidence of neuralgic amyotrophy (parsonage turner syndrome) in a primary care setting—a prospective cohort study. PLoS One. 2015;10:e0128361.
2. Vigasio A, Marcoccio I. Hourglass-like constriction of the suprascapular nerve: a contraindication for minimally invasive surgery. J Shoulder Elbow Surg. 2018;27:e29–37.
3. Pan Y, Wang S, Zheng D, et al. Hourglass-like constrictions of peripheral nerve in the upper extremity: a clinical review and pathological study. Neurosurgery. 2014;75:10–22.
4. Aranyi Z, Csillik A, Devay K, et al. Ultrasonographic identification of nerve pathology in neuralgic amyotrophy: enlargement, constriction, fascicular entwinement, and torsion. Muscle Nerve. 2015;52:503–11.
5. ArAnyi Z, Csillik A, DeVay K, et al. Ultrasonography in neuralgic amyotrophy: sensitivity, spectrum of findings, and clinical correlations. Muscle Nerve. 2017;56:1054–62.
6. Sneag DB, Saltzman EB, Meister DW, Feinberg JH, Lee SK, Wolfe SW. MRI Bullseye sign: an indicator of peripheral nerve constriction in parsonage-turner syndrome. Muscle Nerve. 2017;56:99–106.
7. Kasper JM, Wadhwa V, Scott KM, Rozen S, Xi Y, Chhabra A. SHINKEI—a novel 3D isotropic MR neurography technique: technical advantages over 3DIRTSE-based imaging. Eur Radiol. 2015;25:1672–7.
8. Lieba-Samal D, Jengojan S, Kasprian G, Wober C, Bodner G. Neuroimaging of classic neuralgic amyotrophy. Muscle Nerve. 2016;54:1079–85.
9. Vigasio A, Marcoccio I. Homolateral hourglass-like constrictions of the axillary and suprascapular nerves: case report. J Hand Surg Am. 2009;34:1815–20.
10. Van Alfen N, van Engelen BG. The clinical spectrum of neuralgic amyotrophy in 246 cases. Brain. 2006;129:438–50.
11. Yamamoto S, Nagano A, Mikami Y, Tajiri Y. Multiple constrictions of the radial nerve without external compression. J Hand Surg Am. 2000;25:134–7.
12. Duman I, Guvenc I, Kalyon TA. Neuralgic amyotrophy, diagnosed with magnetic resonance neurography in acute stage: a case report and review of the literature. Neurologist. 2007;13:219–21.
13. Yoneyama M, Takahara T, Kwee TC, Nakamura M, Tabuchi T. Rapid high resolution MR neurography with a diffusion-weighted pre-pulse. Magn Reson Med Sci. 2013;12:111–9.
14. Sneag DB, Rancy SK, Wolfe SW, et al. Brachial plexitis or neuritis? MRI features of lesion distribution in Parsonage–Turner syndrome. Muscle Nerve. 2018;58:359–66.
15. Guerra WK, Schroeder HW. Peripheral nerve palsy by torsional nerve injury. Neurosurgery. 2011;68:1018–24; discussion 1024.