

A Rare Cause of Proximal Thigh Discomfort and Weakness with Type 2 Diabetes Mellitus

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(Received: 25 May 2017; Accepted: 20 September 2017)

ABSTRACT

Diabetic amyotrophy also known as Bruns-Garland syndrome is diabetic neuropathy subtype affecting the lumbosacral nerve roots and peripheral nerves. There is an ongoing debate on the pathophysiology behind this condition whether is it ischaemic, metabolic (hyperglycaemia) or inflammatory. A 36-year-old man with uncontrolled diabetes mellitus complained of unprovoked weight lost and right proximal thigh discomfort with weakness for one week duration. In neurological examination, his right hip flexion was at medical research council (MRC) grade 3, right hip extension MRC grade 4, his knee and ankle flexion and extension are normal (MRC grade 5). The muscle tones and reflexes were normal. Sensation and proprioception were intact bilaterally. Nerve conduction study (NCS) showed markedly reduced amplitude of the compound muscle action potentials and sensory nerve action potentials, while conduction velocities show only mild slowing. He was started on a course of oral prednisolone 10 mg daily and improved gradually. At three months follow-up, his right lower limb power has recovered fully and he can walk without any assistance. Diabetic amyotrophy was confirmed by suggestive clinical features supported by electrophysiological findings of the affected nerves. This condition is due to metabolic derangement and vasculopathy or immune mediated nerve injury. So, the healthcare providers should be aware about this rare complication of diabetes.

Keywords: diabetic amyotrophy, Bruns-Garland syndrome, diabetic lumbosacral radiculoplexus neuropathy, diabetes mellitus type 2

INTRODUCTION

Diabetic amyotrophy is a rare form of diabetic neuropathies affecting mostly type 2 diabetic mellitus patients.¹ The typical clinical features include sudden, asymmetric, focal onset of pain following by weakness involving the proximal leg, with associated autonomic failure and weight loss.² This condition can progress over months to years and is followed by partial to complete recovery. Some patients with diabetic amyotrophy can develop symmetrical weakness. In most cases, the symptoms and signs progress to affect the opposite limb and in the distal legs.² There are reported cases of foot drop or disturbing neuropathic pain that can persist for years.¹ In the series of 33 patients cited earlier, 48 per cent required wheelchair assistance at a certain point of their illness.² There is no proven effective treatment for diabetic amyotrophy available according to a systemic review published in 2012.³ There are some studies which suggest that targeting treatment employing immune suppression helps in clinical improvement. Such therapies include oral prednisolone, intravenous methylprednisolone, intravenous immune globulin, cyclophosphamide and plasma exchange.³

CASE PRESENTATION

A 36-year-old man with type 2 diabetes for 6 years presented with uncontrolled diabetes due to non-adherence to his medications. His previous oral hypoglycaemic agents were tablet metformin 1 g bd and tablet gliclazide 160 mg bd. Baseline HbA1c was 9 – 10% and he

was not keen for insulin therapy previously. He also reported unprovoked weight lost and right proximal thigh discomfort with weakness for one week duration with no predisposing trauma events. His sugar was controlled with subcutaneous actrapid 22 units tds and insulatard 40 units during his inpatient stay. Neurological examination revealed that his right hip flexion was at medical research council (MRC) grade 3, right hip extension MRC grade 4, his knee and ankle flexion and extension are normal (MRC grade 5). The left lower limb power proximal and distally were MRC grade 5. The tones of both lower limbs were normal, reflexes are 1+ bilaterally at both knee and ankle. Sensation and proprioception were intact bilaterally.

Plantar reflexes normal bilaterally. Neurological examination of his upper limbs and cranial nerves were normal.

Spine X-rays were normal. Nerve conduction study (NCS) showed markedly reduced amplitude of the compound muscle action potentials and sensory nerve action potentials, while conduction velocities show only mild slowing (Figures 1 – 4). On the motor conduction (Figures 1 and 2), the amplitudes are reduced more pronouncedly in the lower limbs than upper limbs. After commencing on a course of steroids and the repeat motor conduction NCS after show improved action potential.

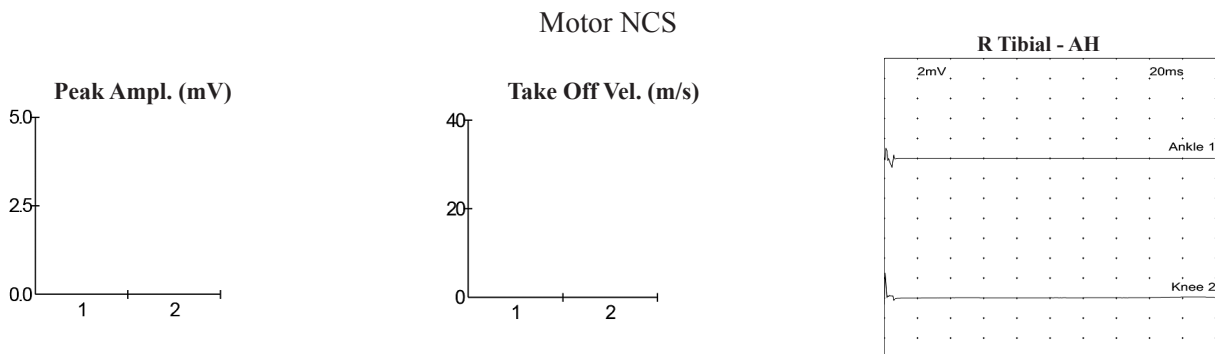


Figure 1 Right Tibial – AH (initially, before starting treatment)

After commencing on a course of steroids and the repeat NCS after show improved action potential in the NCS recording:

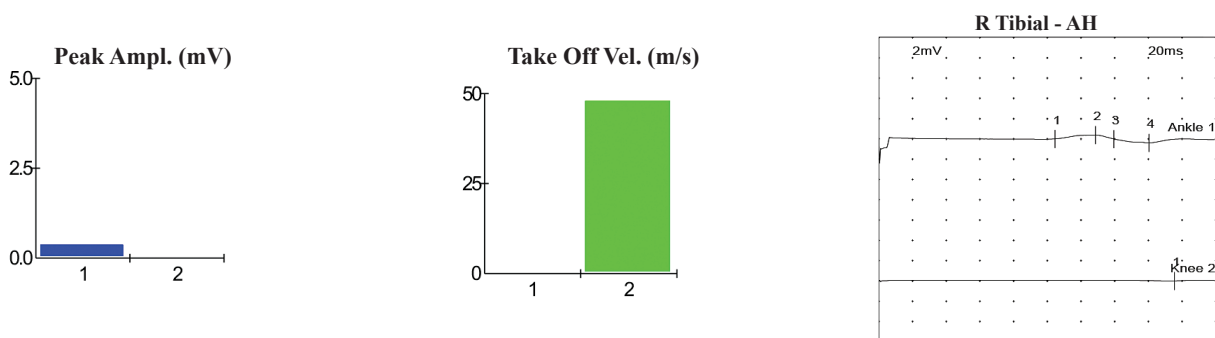


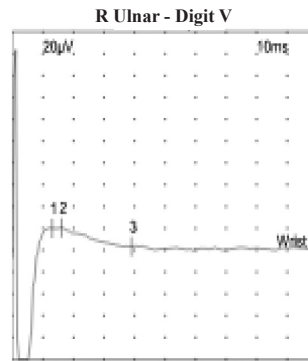
Figure 2 Right Tibial – AH (repeated NCS after treatment)

On sensory conduction (Tables 1 – 2, Figures 3 and 4), minimal amplitudes are seen before and after treatment in the upper limbs. There are no signals detected on the lower limbs sensory NCS study before and after treatment.

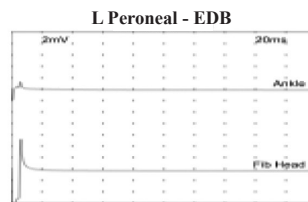
Table 1 Sensory NCS for right ulnar at wrist level

Nerve / Sites	Rec. Site	Latency (ms)	Peak Ampl. (μ V)	Distance (cm)	Velocity (m/s)
R Ulnar – Digit V					
Wrist	V	1.30	0.61	12	92.2

R Ulnar - Digit V



L Peroneal - EDB



L Tibial - AH

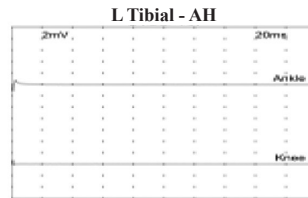
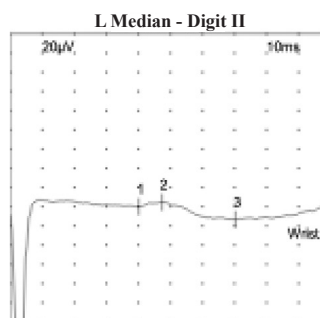


Figure 3 Right ulnar at wrist level before commencing on treatment. The lower limbs sensory NCS conducted failed to pick up any signals.

Table 2 Sensory NCS for left median and right ulnar at wrist level

Nerve / Sites	Rec. Site	Latency (ms)	Peak Ampl. (μ V)	Distance (cm)	Velocity (m/s)
L Median – Digit II					
Wrist	II	4.01	4.7	16	39.9
R Ulnar – Digit V					
Wrist	V	6.98	6.0	14	20.1

L Median - Digit II



R Ulnar - Digit V

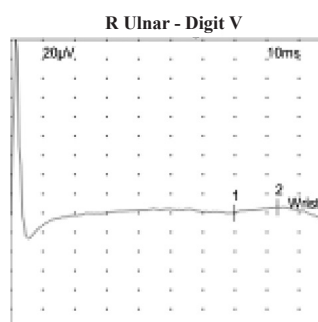


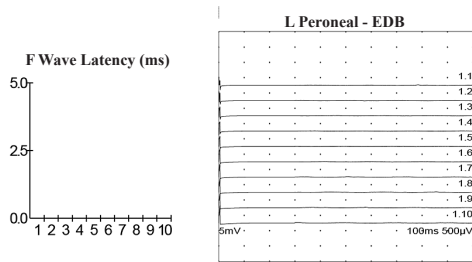
Figure 4 Left median and right ulnar at wrist level after commencing on treatment. Unable to detect any signals from the lower limbs nerves sensory NCS.

On F waves (Table 3, Figures 5 and 6), the F waves showed improvement after treatment in both upper and lower limbs. The repeated NCS after treatment showed improvement in both the motor conduction action potential and F waves of the lower limbs. This NCS shows predominantly axonal peripheral neuropathy which improved with treatment.

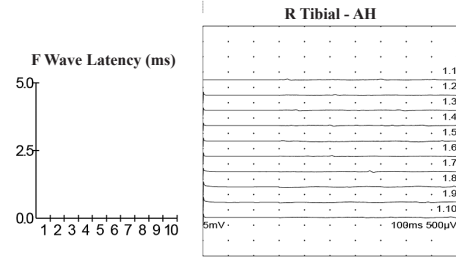
Table 3 F waves

Nerve	Min F Lat (ms)	Max F Lat (ms)	Mean F Lat (ms)
R Tibial – AH	63.28	70.89	65.64

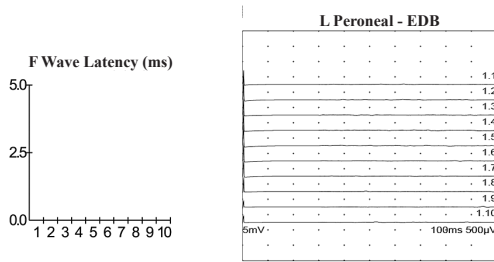
L Peroneal – EDB



L Tibial – AH



R Peroneal – EDB



R Tibial – AH

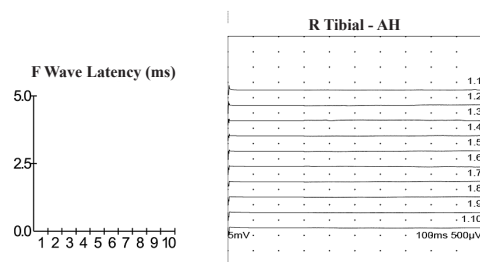
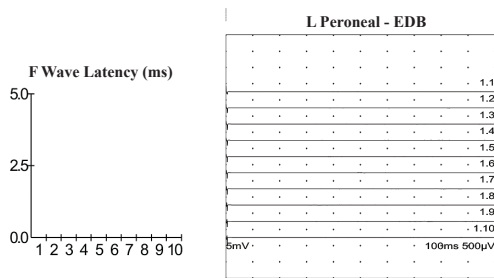
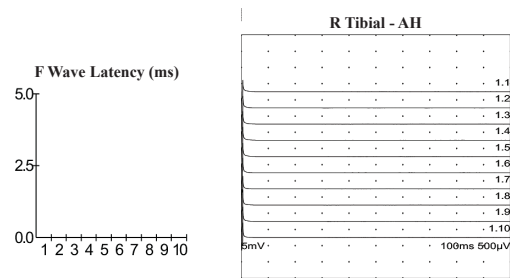


Figure 5 F waves recorded before starting treatment

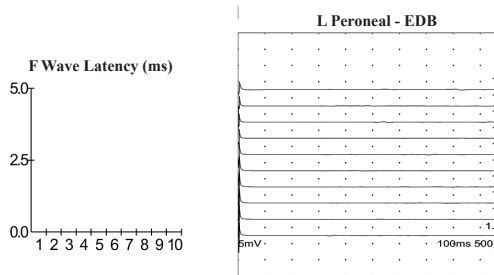
R Peroneal – EDB



R Tibial – AH



R Peroneal – EDB



R Tibial – AH

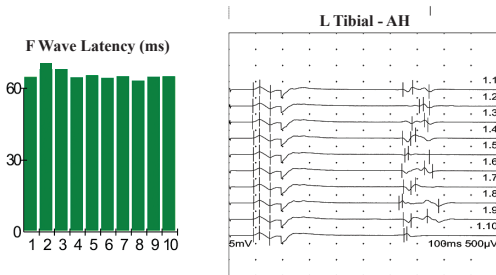


Figure 6 Repeated F waves after treatment showed remarkable improvement in action potential captured

MRI whole spine done was normal. He was started on a course of oral prednisolone 10 mg daily. After one week of treatment, he was able to ambulate with walking frame in the ward. He was under the care of dedicated rehabilitation team. He was subsequently discharged with oral prednisolone 10 mg daily and was given a neurology clinic appointment. At three months follow-up, his right lower limb power has recovered fully and he can walk without any assistance. Oral prednisolone was stopped after 3 months of treatment. His glycaemic control has been optimized and his latest HbA1c has reduced to 7%.

DISCUSSION

Diabetic amyotrophy also known as Bruns-Garland Syndrome or diabetic lumbosacral radiculoplexus neuropathy is a rare subtype of diabetic neuropathies affecting the lumbosacral plexus and its nerve roots.¹ Patient with this condition presented initially with acute to subacute onset of lower limb proximal muscle pain followed by weakness which is often asymmetry.⁴ Occasionally, the thoracic nerve roots and brachial plexus may be involved giving rise to thoracic wall pain and proximal upper limbs pain as well as weakness respectively.⁴ The aetiology for diabetic amyotrophy is not known despite being in existence for over a century. The proposed mechanism for this condition is due to metabolic derangement and vasculopathy.⁵ However this proposal has fallen out of favour as most affected patients have relatively controlled diabetes mellitus. Recent studies have indicated that immune mediated nerve injury has a stronger basis for the mechanism of diabetic amyotrophy yet all the studies were of small scaled without a randomised control method.⁵ This has led to the usage of immunosuppressants in the treatment of diabetic amyotrophy.⁴

In the case illustrated, the diagnosis of diabetic amyotrophy was confirmed by suggestive clinical features supported by

electrophysiological findings of the affected nerves. The patient initially presented with right proximal thigh discomfort and weakness which progresses for a week. This presentation is typical of diabetic amyotrophy. His HbA1c was documented at 9% suggesting that his diabetic was uncontrolled but not to the severe extent. There was no explainable cause, for instance preceding trauma, to explain his symptom. Electrophysiological study in the form of nerve conduction study showed reduced compound muscle action potential of the femoral nerves consistent with reduced velocity of the sensory potential consistent with axonal neuropathy, thus supporting the clinical diagnosis.⁶ There was also subclinical abnormality of the apparently normal left lower limbs to suggest asymmetrical nerve involvement further pointing towards diabetic amyotrophy.

The patient showed dramatic improvement after a course of oral steroid and aggressive rehabilitation for three months. The observed improvement is expected for diabetic amyotrophy. Some investigators reported that the expected mean time recovery was three months and complete recovery was achieved by 18 months in a case series of 27 diabetic amyotrophy patients.⁷

CONCLUSION

Diabetic amyotrophy is a rare cause of proximal thigh weakness and discomfort in type 2 diabetic patient. This case report will help to create awareness among healthcare providers regarding diabetic amyotrophy.

ACKNOWLEDGEMENTS

The authors would like to thank the Director General of Health Malaysia for the permission to publish this paper.

CONFLICT OF INTEREST

The authors declare that they have no competing interests.

CONSENTS

Written informed consent was obtained from the patient to publish the case. A copy of written consent is available for review by the Chief Editor.

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