ORIGINAL RESEARCH PAPER

Neurology

KEYWORD: Guillain barre syndrome, finger drop , tongue wasting, clawing of fingers

GUILLAIN BARRE SYNDROME WITH TONGUE ATROPHY AND FINGER DROP SIGN- A RARE CASE REPORT

B Rama Abhishek*	Resident In Neurology, Department of Neurology, Stanley medical college, Chennai,Tamilnadu *Corresponding Author
K Mugundhan	Associate Professor of Neurology, Department of Neurology, Stanley medical college, Chennai,Tamilnadu
M Sathish Kumar	Assistant Professor of Neurology, Department of Neurology, Stanley medical college, Chennai,Tamilnadu
K Malcolm Jeyaraj	Assistant Professor of Neurology, Department of Neurology, Stanley medical college, Chennai,Tamilnadu
S Sakthi Velayutham	Assistant Professor of Neurology, Department of Neurology, Stanley medical college, Chennai,Tamilnadu
P R Sowmini	Assistant Professor of Neurology, Department of Neurology, Stanley medical college, Chennai,Tamilnadu
S. Vivekasaravanan	Associate Professor of Neurology, Department of Neurology, Stanley medical college, Chennai,Tamilnadu

ABSTRACT

Guillain barre syndrome(GBS) is an acquired autoimmune polyradiculoneuropathy with several variants. We report here a case of 50 year old male who presented with areflexic quadriparesis, tongue atrophy,tongue fasciculations and claw hand with finger drop was diagnosed as GBS supported by electrophysiological study. Patient's neurological deficits improved after a course of Intravenous immunoglobulin therapy.Limb Weakness, tongue atrophy with fasciculations and finger drop with clawing of fingers improved after treatment. Knowledge about these atypical clinical features in GBS will avoid unnecessary delay in investigations and treatment.

INTRODUCTION

Guillain barre syndrome is classically described as an acute inflammatory polyradiculoneuropathy characterised by rapidly progressive,symmetrical, areflexicflaccid weakness associated with albumino-cytological dissociation.[1] Over the past 20 decades, the clinical and pathological spectrum of GBS is expanding which includes variable and atypical presentations apart from miller fisher syndrome.[2] Although, tongue wasting and finger drop in GBS have been described previously, we would like to highlight those rare findings in our case.

CASE REPORT

A 50 year old male, smoker ,non alcoholic , who is carpenter by occupation presented to our hospital with history of weakness of all four limbs with sensory disturbances of 10 days duration. He initially had dull aching pain in both lower limbs for 2 days following which he had difficulty in getting up from squatting position along with slippage of footwear while walking. Two days later,the weakness progressed to upper limbs in the form of difficulty in lifting weights , gripping objects and mixing food. The weakness was acute in onset, symmetrical in nature and progressed within 7 days. He also had sensory disturbances in the form of paresthesias on both feet .He denied difficulty in swallowing ,nasal regurgitation and breathlessness.He had no neck pain, bowel or bladder dysfunction. On examination he was hemodynamically stable.His single breath count was 30. Cranial nerve examination revealed significant tongue atrophy (figure -01) with prominent fasciculations and weakness of tongue movements.He had wasting of intrinsic hand muscles and clawing of both hands without any fasciculations. Hypotonia was noted in all four limbs .His power was 3/5 (MRC grade) with hand grip of 30% of normal. Distal muscles were more affected than proximal muscles with wrist drop and finger drop (figure 02). There was clawing of fingers in both hands(figure 03).All Deep tendon reflexes were sluggish. Elicitation of Plantar reflex resulted in flexor response bilaterally. Sensory examination showed graded sensory loss(vibration and position sense) in all four limbs.

Blood Investigations were within normal limits. Nerve conduction study showed prolonged distal latency in both median and ulnar CMAPs(compound motor action potentials) with reduction in amplitude and conduction velocity. Peroneal and tibial nerve CMAPs were not obtained. SNAPs(sensory nerve action potentials) were also not obtained. F waves were absent. The electrophysiological findings were suggestive of demyelinating sensorimotor polyradiculoneuropathy. CSF analysis revealed albuminocytological dissociation. Electromyography(EMG) of tongue muscles demonstrated neurogenic pattern with fasciculations. Vasculitic workup and Antinuclear antibody with extractable nuclear antigen panel were negative. Patient was started on Intravenous immunoglobulin (0.4gm/kg/day) for 5 days along with physiotherapy. Significant improvement was noted in weakness with power improving to 4+/5(MRC),hand grip improved to 90 percent after treatment. Tongue fasciculations and finger drop with clawing of hands disappeared after treatment (figure 04-06).Repeat NCS was done after 6 months of treatment, which showed sensory motor demyelinating polyradiculoneuropathy with mild improvement in conduction velocities and CMAP amplitude compared to baseline values.

Images Prior To Treatment

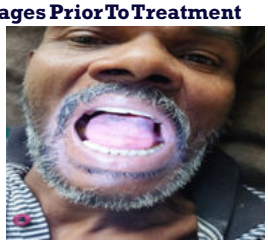

Figure 01-showing Tongue Wasting

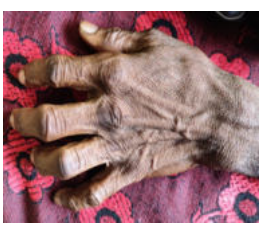

Figure 02-showing finger drop sign

Figure 03-showing clawing of fingers

Images Showing Improved Findings After Treatment

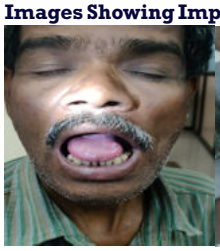

fig 04- improvement of tongue fasciculations

fig 05- improvement of finger drop

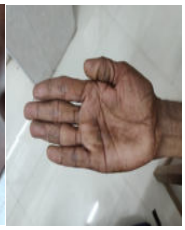

Fig 06- improvement of clawing of fingers

DISCUSSION

Guillain Barre Syndrome is a heterogenous entity and most frequent cause of acute or subacute onset neuromuscular weakness worldwide. Often it is a monophasic illness with 60% of cases have preceeding infection .Almost 30% of GBS patients require ventilator due to respiratory failure which explains the severity of the disease.[3] Although weakness in GBS is usually more proximal than distal,many can have predominant distal weakness .Of several variants in GBS, patients with AMAN variant present with severe distal weakness compared to proximal weakness.Our patient had more distal weakness than proximal weakness. Finger Drop sign is another rare entity described in GBS, where severe finger extensor weakness in the presence of relatively normal power of finger flexors or wrist flexors causes clawing of fingers which was seen in our patient. Finger drop sign or predominant finger extensor weakness is relatively specific for AMAN variant of GBS.[4]In patients with AIDP variant, finger extensors are equally involved as finger flexors which was not so in our patient.Few patients can have hand-onset GBS,with weak hand grip and clumsy fingers.[5] Cranial nerve involvement in GBS is seen in around 45-75% of cases, of which facial nerve involvement is most common followed by extraocular muscle and lower cranial nerve involvement although,few studies have showed that bulbar palsy is more common than facial nerve involvement. Hypoglossal nerve involvement in GBS was also described as rare.[6] Till date, only four GBS cases have been reported with hypoglossal palsy either isolated or associated multiple cranial nerve palsy.[7] Our patient had atrophy of tongue with fasciculations. In our case, patient had no other cranial nerve palsy apart from twelfth nerve involvement. Tongue wasting and fasciculations had improved after 5 months of treatment. Finger and wrist drop with clawing of fingers improved after 2 weeks of immunotherapy.

CONCLUSION

This case is reported to highlight the atypical findings of Guillain barre syndrome.Distal predominant weakness in the form of finger drop sign with clawing of fingers and isolated 12th nerve palsy were observed in our patient who had AIDP variant of GBS. Early recognition of these unusual findings expands the clinical awareness among physicians which inturn paveway for timely appropriate management. Knowledge about these atypical clinical features in GBS will avoid unnecessary delay in investigations and treatment.

REFERENCES

1. Hiraga A, Mori M, Ogawara K, Hattori T, Kuwabara S. Differences in patterns of progression in demyelinating and axonal Guillain-Barré syndromes. Neurology. 2003;61(4):471-474. doi:10.1212/01.wnl.0000081231.08914.a1
2. Tsivgoulis G, Tsakaldimi S, Vadikolias K, et al. Bilateral claw hand: an uncommon presentation of regional Guillain-Barré syndrome. J Neurol Sci. 2013;334(1-2):24-25.doi:10.1016/j.jns.2013.07.009.
3. Mazen M. Dimachkie, MD, FAAN, FANA; David S. Saperstein, MD Peripheral Nervous System Disorders p. 1241-1260 October 2014, Vol.20,No.5.doi: 10.1212/01.CON.0000455883.91426.12
4. George A, Aburehiman P, James J. "Finger drop sign" in Guillain-Barré syndrome. Neurol India. 2009;57(3):282-286. doi:10.4103/0028-3886.53276
5. Mori I, Koga M, Hirata K, Yuki N. Hand weakness onset Guillain-Barré syndrome. J Neurol Neurosurg Psychiatry. 2004;75(1):169-170.
6. Tan AK, Chee MW. Fulminant Guillain-Barré syndrome with quadriplegia and total paresis of motor cranial nerves as a result of segmental demyelination. J Neurol Sci. 1995;134(1-2):203-206. doi:10.1016/0022-510x(95)00235-8.
7. Chakrabarti S, Pan K. Hypoglossal nerve palsy in a case of Guillain-Barré syndrome. Ann Trop Med Public Health 2014;7:270: doi: 10.4103/1755-6783.155026.
8. Eswaradass PV, Ramasamy B, Kalidoss R, Gnanashanmugham G. A rare cause of twelfth-nerve palsy: Guillain-Barré Syndrome. Arch Med Health Sci 2017;5:140-1. doi: 10.4103/2321-4848.208186
9. Nanda SK, Jayalakshmi S, Ruikar D, Surath M. Twelfth cranial nerve involvement in Guillian Barre syndrome. J Neurosci Rural Pract. 2013;4(3):338-340. doi:10.4103/0976-3147.118804.
10. Polo JM, Alañá-García M, Cacabelos-Pérez P, Ortín-Castaño A, Ciudad-Bautista J, López-Alburquerque JT. Síndrome de Guillain-Barré atípico: neuropatía craneal múltiple [Atypical Guillain-Barré syndrome: multiple cranial neuropathy]. Rev Neurol. 2002;34(9):835-837.