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Case Report

Dropped Head Syndrome-A Rare Clinical Presentation of Myasthenia Gravis

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***ABSTRACT*:**
'Dropped head syndrome' (DHS) can be associated with several neurological diseases. It is particularly of great challenge to manage such cases where there is absence of neurological clues to the underlying cause of DHS. The restricted weakness of extensor muscles of the neck is relatively a rare condition which has been designated as DHS or 'dropped head syndrome' and it may be associated with a significant number of neurological diseases, such as motor-neuron, neuromuscular and neurodegenerative disorders. When DHS is accompanied by neurological features of such disorders, potential causes, like dystonia and myositis can be confirmed, thereby facilitating the selection of the treatment. We report a case of myasthenia gravis (MG) who was treated with Pyridostigmine and Methyl Prednisolone which demonstrated significant improvement.
Key words: Dropped Head Syndrome (DHS), Myasthenia gravis, neurological disease.

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INTRODUCTION:
The Dropped head syndrome (DHS) is characterized by severe weakness of the paravertebral extensor muscle, resulting in the chin-on-chest deformity.[1] There are various conditions associated with the DHS that have been reported, including the neuromuscular diseases, e.g. motor neuron disease, myasthenia gravis, congenital myopathy, mitochondriopathy and chronic inflammatory demyelinating polyneuropathy.[2] Among these conditions, some presents with DHS are systemic and progressive, and also have a grave prognosis, like amyotrophic lateral sclerosis.[3] While some others have benign clinical course without any generalized neuromuscular disorder and are diagnosed as an isolated neck extensor myopathy (INEM). We report here a case of DHS due to myasthenia gravis that was successfully treated successfully by Pyridostigmine and Methyl Prednisolone.

CASE REPORT: A 63 year old man came with a history of sudden onset dropping of head of sub-acute duration. On clinical examination, the patient was found to have neck extensor weakness. Bilateral ptosis and bilateral external opthalmoplegia was present. Patient was having reduced palatal movements and on examination, the cough reflex was reduced. Diurnal variation of symptoms was present. Motor examination of the patient showed proximal muscle weakness (bilateral lower extremities), patient was investigated for the differentials of dropped head syndrome and considering the sensory examination being normal, pure motor syndrome was the probable diagnosis. So, the possibilities kept in mind were: Myasthenia Gravis**,** Hypothyroidism, Polymyositis, Lambert-Eaton Syndrome and ALS
So, keeping the various differentials in mind, CT-Chest was done to rule out thymoma and oat-cell carcinoma of lung which was normal. Then, Repeated Nerve Stimulation (RNS) was done which showed significant decremental response in the bilateral orbicularis oculi muscle. Among the serological tests, Serum Ach-R Antibody was done which was found to be positive. So, our diagnosis of Myasthenia Gravis was made based on above investigations. His motor examination showed proximal muscle weakness (bilateral lower extremities) due to which he was

investigated for the differentials of dropped head syndrome and considering the sensory examination being normal, pure motor syndrome was the probable diagnosis. Dropped Head Syndrome is usually a challenge to neurologists since it has a broad differential diagnosis. The causes vary from muscular, neurological to neuro-muscular. In the present case report, the patient suffered from the complication of DHS due to Myasthenia Gravis which is a neuro-muscular disorder in which anti-bodies are formed against the Ach-receptors at the post-synaptic membrane. Due to this reason, detection of Ach-R antibodies in our patient's serum confirmed our diagnosis. As the serological testing confirmed Myasthenia Gravis, thereby the final diagnosis of Dropped Head Syndrome due to Myasthenia Gravis was made and the patient was started on Pyridostigmine-30 mg-6 hour which is a cholinomimetic alkaloid and helps improve the concentration of Acetyl Choline available post-junctionally. Along with this, he was started on Methyl Prednisolone-60 mg in divided doses which is a corticosteroid. Patient significantly improved over a period of 60 days on follow up.

DISCUSSION: DHS is characterized by the severe weakness of the cervical paraspinal muscles.[4]Various literatures on the surgical management include the optimal timing of intervention and the levels which will involve decompression and instrumented fusion. While some cases of DHS are systemic as well as progressive with a grave prognosis, while others present with the absence of generalized neuromuscular disorder and have a benign clinical course.[5] The various differential diagnoses are: Neurological- Amylotrophic Lateral Sclerosis, Parkinson's disease; Muscular- Polymyositis, Scleromyositis, Hypothyroidism; Neuro-muscular-Myasthenia Gravis, Lambert-Eaton Syndrome; Others- Malignancy, Post-surgical. [6]Although, it has been reported that paraspinal cervical muscles develop pathological abnormalities with increasing age and both the raggedred fibers as well as the accumulation of mitochondria are frequent findings in aging cervical muscles.[7] Myasthenia gravis (MG) should be considered as a possible cause of INEM. The clinical history of various patients affected by MG is usually characterized by the weakness as well as the fatigability of the muscles on repeated activity that improve after rest. The patients usually present not only difficulties of holding their heads upright, but also with the weakness of legs, diplopia, ptosis and slurring of speech. Recently, D'Amelio et al. described an unusual case of MG, presenting the isolated weakness of neck extensor muscles.[8] A Tensilon test or edrophonium test is essential for the diagnosis of MG. Due to the small number of reported cases; it is yet controversial whether the surgical treatment is indicated for the drooped head syndrome. There has been as reported case of failure after the surgical treatment with posterior correction and the fusion without anterior reconstruction.[9] In the present article, DH responded to steroids and anticholinesterase agents. MG can present with DH as well as neck weakness is initial manifestation in 3% of the patients with MG. The myasthenia gravis is known to affect the neck muscles, predominantly the neck flexors and DH has developed along with the bulbar, ocular or limb involvement late in the course of disease. Although, about 20% of the patients with MG have no anti-AchR antibody, which often creates the difficulty in diagnosing MG when only DH is the manifestation. This responsiveness to pyridostigmine and steroid suggests that a neuromuscular junction disorder probably caused DH. Interval from the onset of DH to the presentation of typical MG features differed between patients with and without anti-AchR antibody. [10]

CONCLUSION: DHS is relatively a rare condition that may occur as a result of several muscular, neurological, neuromuscular and other causes. It is of great important to differentiate between neuromuscular (NM) and non-neuromuscular (non-NM) causes in order to offer the best treatment options to the patients. A detailed history taking and appropriate physical examinations is the key to make an accurate diagnosis. As reported in the present case, the patient was managed medically for DHS in myasthenia gravis patient with significant improvement over a period of 60 days on follow up.

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