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Original Research Article

Characterizing dropped head syndrome across neurologic disorders, the hidden epidemic of dropped head syndrome

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ABSTRACT

Background: Dropped head syndrome (DHS) usually characterizes neuromuscular disorders, however, is present in other neurological disorders as Parkinson's disease. Despite being a syndrome seen in the clinic, it's uncommon reported in the literature.
Methods: This retrospective study reviewed electronic medical records of patients hospitalized with neurological disorders from 2018-2022 in a single center. Of 1359 charts screened, 134 patients were confirmed to have co-occurring DHS based on documented neck extensor weakness and an associated neurological condition. Strict inclusion criteria were applied to ensure all included patients had a confirmed neurological disorder and physical exam findings consistent with DHS.
Results: Of 1359 patients hospitalized with neurological disorders, only 134 (0.96%) had co-occurring DHS. The cohort included 69 females (ages 24-79 years) and 65 males (25-87 years). The most prevalent neurological diagnoses linked to DHS were Parkinson's disease, chronic inflammatory demyelinating polyneuropathy, and myasthenia gravis. Less common associations were multiple system atrophy and Alzheimer's disease. The observed DHS prevalence may underestimate the true rate, highlighting the need for enhanced awareness and assessment.
Conclusions: Despite a high prevalence of neurological disorders like Parkinson's disease, chronic inflammatory demyelinating polyneuropathy, and myasthenia gravis at our hospital, dropped head syndrome remained uncommon, affecting only 134 out of 1359 (0.96%) hospitalized neurological patients. DHS showed no sex predilection but predominantly affected older individuals, likely reflective of age-related sarcopenia. This study provides initial data on the prevalence of DHS across different neurologic conditions and underscores the need for early detection to enable timely surgical and nonsurgical interventions. Further research is warranted to elucidate the risk factors and optimal management strategies for this enigmatic condition.

Key words: Fallen head, Neurological diseases, Chronic disease, Dropped head syndrome

INTRODUCTION

In the year of 1980 Katz reported the floppy head syndrome in a case with dropped head accompanied by muscle weakness of the neck.[1] In 1992 it was described by Suarez and Kelly Jr the isolated type of dropped syndrome, they described that this type of dropped head syndrome (DHS) affects commonly to elderly population and is caused by noninflammatory myopathy restricted to the paraspinal muscles of the neck.[2] Despite the DHS being prevalent, this disorder is uncommonly reported, especially in our country, raising the need to approximate this prevalence in our community; Specially because this disorder is characterized by weakness of neck extensor muscles (paraspinal muscles), the flexibility of neck movements is preserved, unlike ankylosing spondylitis in

which the neck is rigid.[3] A review on DHS made by Sharan et al suggests that the semispinalis cervicis and capitis muscles may be primarily responsible for neck extension. In the literature, some authors explain that DHS it is a result of selective injury to the lower motor neurons in the spinal cord, but on the other hand some other authors suggest that the spinal roots or peripheral nerves are the primary sites of injury. Curiously both myopathic and neurogenic electromyographic changes have been reported. The muscle weakness leads to cervical kyphotic posture, which causes the forward drop of the head, resulting in a fixed flexion deformity of the cervicothoracic spine until a chin-on-chest condition, however, this condition differs from camptocormia, because, this last is an entity characterized by a kyphosis lumbar reductible in supine decubitus.[4] DHS can be observed both in extrapyramidal and neuromuscular disorders, as well as in dystrophic, metabolic, non-inflammatory or inflammatory myopathies.[5]

The most common disorders related to DHS are polymyositis, chronic inflammatory demyelinating polyneuropathy (CIDP), myasthenia gravis, amyotrophic lateral sclerosis (ALS), inclusion body myositis (IBM) etc.[6] Another condition that could mimic DHS is the *"antecollis"*, which is a rare condition which is presented in metabolic diseases, Parkinson, ALS, myasthenia gravis, Lambert-Eaton's syndrome, facioscapulohumeral dystrophy, CIDP, hypokalemic, hyperparathyroidism related myopathy; and related with dopamine and neuroleptic drugs.[7] It is thought that radiation damage the mature muscle fibers, and this led to DHS, the radiation dose is around 40 Gy.

Other case reports were related with the use of chemotherapy, mainly with MOPP (Mecloretamin, Oncovin®, Procarbazine and Prednison) or ABVD (Adriamicine, Bleomicine, Vinblastin, Dacarbazine), even these therapies were combined with radiation therapy for the treatment of Hodgkin disease. The agents combined with mantle irradiation might enhance radiation-induced chronic adverse effects.[8,9] Djaldetti et al have described that camptocormia is a rare condition in Parkinson's disease, these suggest that dropped head syndrome and camptocormia in Parkinson's disease may occur because of similar pathophysiological process affecting paraspinal muscles, however, both syndromes have a different clinical picture.[10]

Primary objective of the study was to determine the prevalence of Dropped Head Syndrome in patients with neurologic disorders hospitalized from 2018 to 2022.

METHODS

This was a retrospective, observational study. We reviewed electronic medical records for adult patients hospitalized with neurological disorders at Hospital de Especialidades Antonio Fraga Mouret, CMN La Raza between 2018-2022.

We included patients aged ≥18 years who had documented neck extensor muscle weakness or other conditions that could explain DHS.

All patients had a neurological disorder that either preceded hospitalization or developed during hospitalization as a complication of prolonged immobilization. We excluded patients with incomplete medical records, or those with non-neurological disorders that could explain DHS (e.g. ankylosing spondylitis, previous trauma, radiotherapy, chemotherapy, kyphotic deformities). We also excluded those with generalized weakness from conditions like severe hypokalemia or mechanical ventilation, as these were considered possible non-neurological causes of DHS. The main criteria for DHS were neck extensor weakness with preserved flexibility of neck movements attributed to a neurological disorder. All included patients had a prior diagnosis of DHS made by a neurologist based on clinical evaluation, electrophysiological testing, or neuroimaging. Of the initial 1359 patients screened, 1225 were excluded based on the criteria above (Figure 1). The final analytic sample included 134 patients. Information extracted from the medical records included demographic data, medical history, neurological diagnosis, presence of neck extensor weakness, and other potential causes of DHS.

We reported descriptive statistics as frequencies and percentages for categorical variables, and means with standard deviations or medians with interquartile ranges for continuous variables. Neurological diagnoses were summarized by sex (Table 1) and graphed into a sankey plot (Figure 2). All analyses were conducted using SPSS version 22.0 (IBM Corp, Armonk, NY) and Rstudio.

RESULTS

Of the 1359 patients screened, 1225 were excluded based on predefined criteria (Figure 1), leaving 134 patients in the final analytic sample. This included 69 females (age range 24-79 years) and 65 males (25-87 years) (Table 1). The leading neurological diagnoses associated with DHS were Parkinson's disease in 31 patients (19 males, 12 females, age range 58-87 years), chronic inflammatory demyelinating polyneuropathy in 25 patients (11 males, 14 females, 30-79 years), and myasthenia gravis in 15 patients (3 males, 12 females, 24-69 years).

Other diagnoses linked to DHS were cervical myelopathy in 10 patients, neoplastic disorders such as astrocytoma and meningioma in 8 patients, inclusion body myositis in 7 patients, amyotrophic lateral sclerosis in 5 patients, rapidly progressive dementia in 4 patients, autoimmune encephalitis in 3 patients, polymyositis in 3 patients, spinal cord atrophy in 2 patients, anoxic-ischemic encephalopathy in 2 patients, muscular dystrophy in 2 patients, infectious encephalopathy in 2 patients, and 1 case each of multiple system atrophy, Alzheimer's disease, multiple sclerosis, and anterior horn cell disorders (Table 1).

Table 1: Neurological diagnoses among patients with dropped head syndrome.

Neurologic disorder	Age (years)	N	%	Females, N (%)	Males, N (%)
Parkinson disease	68 (58-87)	31	23.1	12 (38.7)	19 (61.2)
Chronic inflammatory demyelinating polyneuropathy	48 (30-79)	25	18.7	14 (56)	11 (44)
Myasthenia gravis	44 (24-69)	15	11.2	12 (80)	3 (20)
Narrow medullary canal	50 (25-70)	11	8.2	6 (54.5)	5 (45.4)
Cervical myelopathy	44 (25-80)	10	7.5	5 (50)	5 (50)
Medullary neoplasia	53 (32-79)	8	6	5 (62.5)	3 (37.5)
Body inclusion myositis	59 (51-74)	7	5.2	4 (57.1)	3 (42.8)
Amyotrophic lateral sclerosis	59 (55-65)	5	3.7	3 (60)	2 (40)
Rapidly progressive dementia	56 (44-71)	4	3	1 (25)	3 (75)
Autoimmune encephalitis	27 (23-30)	3	2.2	0	3(100)
Polymyositis	50 (34-61)	3	2.2	1 (33.3)	2 (66.6)
Spinal cord atrophy	32 (24-39)	2	1.5	2 (100)	0
Anoxoischemic encephalopathy	44 (42-46)	2	1.5	1 (50)	1 (50)
Muscular dystrophy	30 (24-35)	2	1.5	0	2 (100)
Infectious encephalopathy	37(32-41)	2	1.5	1 (50)	1 (50)
Multiple systems atrophy	79	1	0.7	0	1 (100)
Alzheimer disease	63	1	0.7	1 (100)	0
Multiple sclerosis	36	1	0.7	1 (100)	0
Medullary anterior syndrome	65	1	0.7	0	1 (100)
Total		134	100	69	65

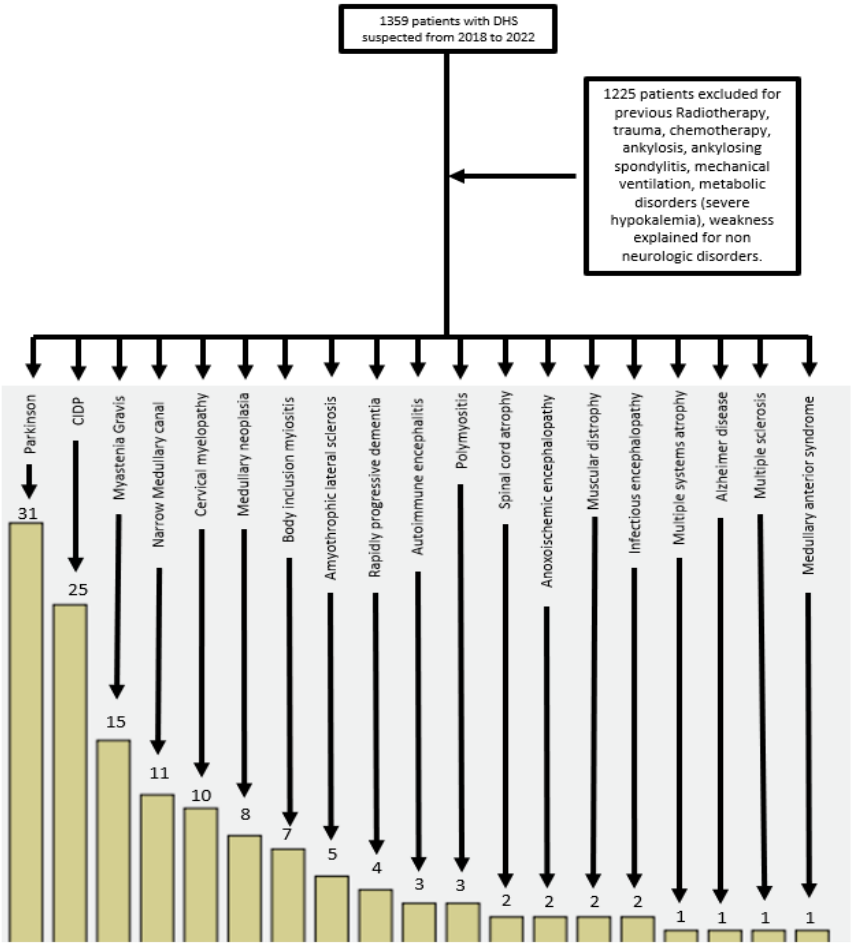

Figure 1: Study patient's flowchart, with patients included in the final analysis, and the neurologic disorders related to DHS.

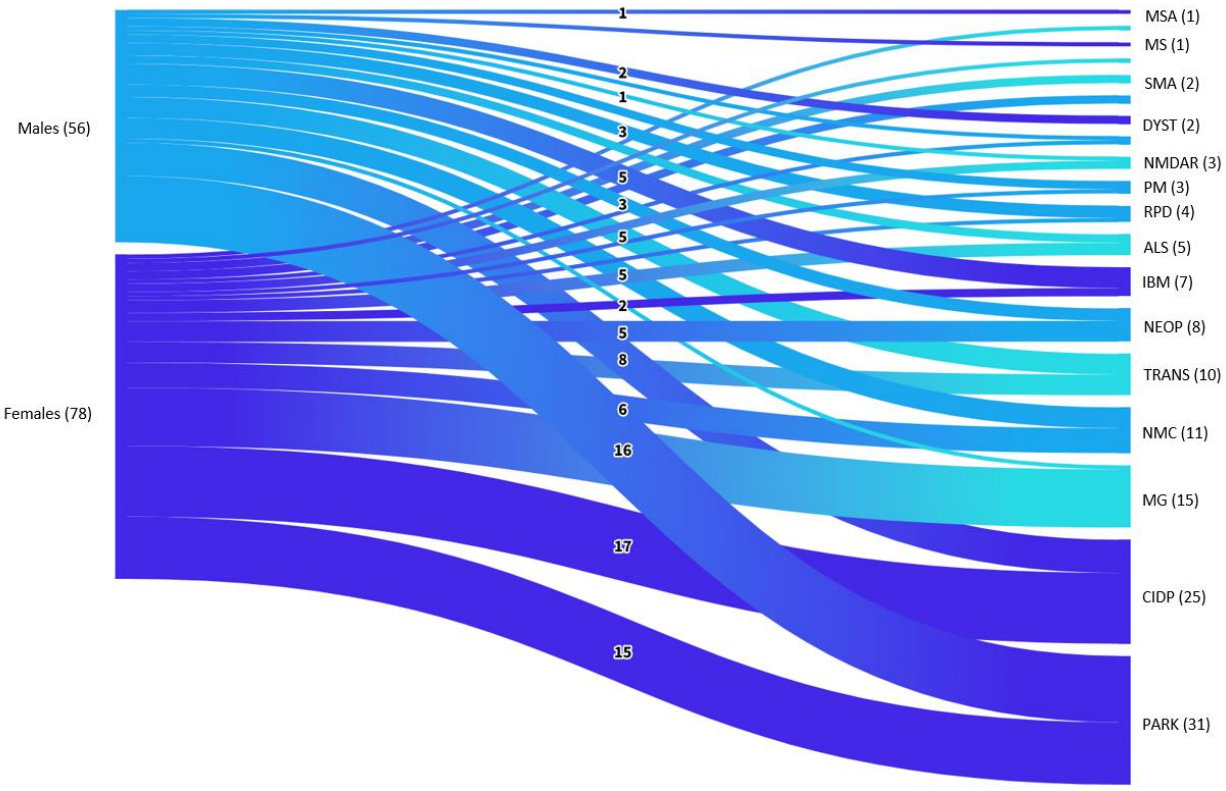

Figure 2: Sankey Plot; sex-specific patterns of leading neurological conditions associated with dropped head síndrome; SMA (Spinal Muscular Atrophy); MSA (Multiple System Atrophy); ANOX (Anoxic-Ischemic Encephalopathy); TRANS (Transverse Myelitis); CIDP (Chronic Inflammatory Demyelinating Polyneuropathy); NMC (Narrow Medullary Canal); DEM (Dementias); DYST (Dystrophies); RPD (Rapidly Progressive Dementia); ALS (Amyotrophic Lateral Sclerosis); MS (Multiple Sclerosis); INF (Infectious); MG (Myasthenia Gravis); IBM (Inclusion Body Myositis); NEOP (Neoplasms); anti-NMDAR (Anti-NMDA Receptor Encephalitis); PARK (Parkinson's); PM (Polymyositis).

While no clear sex predominance was observed across neurological disorders leading to DHS, some less common causes were only present in female patients. In general, DHS showed a tendency to occur in older individuals regardless of sex. Further research is warranted to better characterize the associations between specific neurological conditions and risk of DHS.

DISCUSSION

The purpose of our study was to assess the prevalence of DHS in neurologic diseases, our data showed that despite that the DHS is a rare condition, most of literature reported as a case report, mainly reported in rheumatological, trauma, oncologic, mechanical, mineral and neurological disorders. Considering that during this period, around 13916 patients with neurological disorders were hospitalized, we obtain a prevalence of 0.96% in our unit, however, this prevalence might be overestimated, because the main criteria used to determine DHS was clinical diagnosis, lacking imaging, electromyography, or laboratory test to confirm this condition. Based on our results, the prevalence in neurological disorders is high, and hardly applicable for the general population.

The weakness of neck extensor muscles of paraspinal muscles that characterizes the DHS, as well as, preservation of flexibility of neck movements preserved play an important role in the clinical diagnosis of these patients, however, the etiology of this condition differs from any disorder, in ALS the head drop can be attributed to the involvement of anterior horn cells innervating the paraspinal muscles, patients are unable to lift their chin off the chest wall and find difficulty in looking forward while walking, talking, or eating, using their hands to support their chin.[11]

On the other hand, it is reported that DHS is rare in parkinson disease, in advanced and early stages, however, it is known to progress subacutely over a period of several days, according to some studies, DHS is relatively common in parkinson disease, the main mechanism is either dystonia of flexor neck muscles or weakness of extensor neck muscles. Moreover, in parkinson, the camptocormia is an entity characterized by a kyphosis lumbar reductible in supine decubitus, associated to dysfunctions in computed tomography scans and histologics in the paravertebrals muscles of the lumbar segment, a feature that is not presented in the DHS.[12]

The DHS in the myopathic illness may initially present as an isolated head drop, it is described that myasthenia gravis with MuSK-positive receptor can be presented as DHS, the dropped head may be non-fluctuating and nonfatigable, however, the repetitive nerve stimulation is useful in distinguishing between myasthenia gravis and isolated neck extensor myopathy. In the myopathies the EMG helps in its diagnosis, showing a neurogenic pattern with increased spontaneous activity, fibrillations and fasciculation potentials. In muscular biopsy may be similar to those in isolated neck extensor myopathy or may show inflammatory changes. Finally other less common causes of head drop include CIDP, which generally involves other muscle groups along with cervical muscles.[13]

Despite being a less common cause of DHS, our study showed 25 cases during this period, however, electro-phisiological studies and imaging are needed to confirm this diagnosis, which were not done, or registered in the electronic file; this might overestimate the DHS, contrasting to other studies which mention a lower prevalence.

The diagnosis is mainly clinical, but we suggest complementary studies such as electromyography, as well as imaging studies to confirm this diagnosis, because, must be distinguished from the others conditions that affect this muscle groups. Previous studies have reported lower prevalence of DHS compared to our findings. For example, a study by Suarez and Kelly found a DHS prevalence of only 2.3% among patients with connective tissue disorders.[14] Similarly, a retrospective case series by Munhoz et al. documented a DHS frequency of just 0.025% in patients undergoing botulinum toxin injections.[15]

The higher prevalence in our sample may be partly explained by lack of confirmatory electrophysiological or imaging studies, resulting in possible overestimation.

As we suggested, additional diagnostic testing is warranted to confirm DHS and distinguish it from other mimicking conditions affecting the same muscle groups. Standardized diagnostic criteria may improve accuracy and allow better comparison to other studies.

Limitations

Current study have several limitations, the most important limitation is that this disorder was mainly diagnosed by a neurologist, with a few cases diagnosed by specialist in neurological disorders, moreover, this is a retrospective study, and the clinical picture of the patients could be overestimated in the clinical files, however, this is an approximation to the prevalence of DHS in neurological disorders in our community, and future, prospective studies are needed to determine more exactly prevalence of this condition in our community.

CONCLUSION

In this retrospective study, we aimed to determine the prevalence of DHS across various neurological disorders. Despite differing underlying etiologies and pathophysiologies, neck extensor weakness represents a shared clinical sign among conditions as diverse as neuropathies, neurodegenerative diseases, myopathies, mechanical spinal cord compression, and other miscellaneous entities. While DHS can be diagnosed clinically, our findings reinforce the need for electrophysiological testing and neuroimaging to confirm the diagnosis and distinguish from mimicking conditions affecting the same muscle groups. During evaluation of suspected DHS, it is particularly important to differentiate from camptocormia given the close link between these two disabling movement disorders. However, a key differentiating feature is that camptocormia manifests as lumbar kyphosis reducible with recumbency, which is not seen in DHS. Moving forward, large prospective multicenter studies are warranted to elucidate the natural history of DHS, with longitudinal follow-up, electromyography, imaging, muscle biopsy, and creatine kinase testing. This foundational work can pave the way for future interventional trials aimed at prevention and treatment of DHS across the broad range of associated neurological conditions. Our study provides an important first step, but further research is needed to deepen understanding of this enigmatic syndrome.

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