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Dropped Head Syndrome: A Case Series in an Internal Medicine Department

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Abstract

Background: *Dropped head syndrome is a flexion of the head on the trunk secondary to a deficit of the cervical extensor muscles. It has been rarely reported in the literature.*

Cases Presentation: *We presented a case series of six patients with dropped head syndrome admitted during six months. All patients were female, and their median age was 30 years. All patients had a muscle deficit ≤ 2/5. Extra-muscular signs were constant. CPK levels ranged from 2.3 to 358.4 times normal. The diagnoses associated with dropped head syndrome were as follows: Dermatomyositis (two cases), Overlap myositis (one case), Anti-synthetase syndrome (one case), Thyrotoxic hypokalemic periodic paralysis (one case), Unclassified myositis (one case). Corticosteroid therapy was proposed in four patients. Potassium supplementation was done for the patient with hypokalemic paralysis. One patient did not receive any specific treatment. Methotrexate was started in one patient. The evolution was marked by death in three patients, favorable in two patients. One patient was lost to follow-up.*

Conclusion: *Dropped head syndrome, in our experience, was associated with a severe muscle deficit, a poor prognosis and inflammatory myopathies.*

Keywords: *Dropped Head Syndrome, Inflammatory Myopathies, Internal Medicine*

Introduction

Dropped Head syndrome (DHS) is a flexion of the head over the trunk secondary to a deficit in the cervical extensor muscles [1].

It was first described three decades ago [2]. It has been rarely reported in the literature [1]. Most data concern isolated cases [3,4, 5, 6,7]. However, a meta-analysis of 74 studies reported 129 patients with this syndrome [1]. The diagnosis of DHS is clinical but requires a rigorous etiological investigation.

The etiologies associated with this syndrome are variable and may include neurological, neuromuscular, muscular, and sometimes unidentified disorders [1, 6, 7, 8]. Inflammatory myopathies are rarely involved [2]. Treatment and prognosis vary according to the cause.

We presented a cases series describing six observations of DHS in an internal medicine department. The objectives were to determine the clinical features, the etiologies and the prognosis associated with this syndrome.

Cases Presentation

Six female patients with DHS were hospitalized in our institution from June to December 2017. One patient consulted for acute inflammatory myalgias associated with functional impotence of the limbs and an inability to raise the head that settled rapidly. In three other patients, the onset was slower with chronic myalgia. One patient presented a sudden motor deficit in all four limbs and a drooping head without associated myalgias. The last patient was hospitalized with acute chest pain and dyspnea and physical examination revealed myogenic syndrome and DHS.

The characteristics of our study population is noted in table 1.

Table1. *characteristics of our patients*

Parameters	Number
Age median (range)	30 years (16-42)
Gender (% female)	6/6 (100%)
Median length of stay (days)	10 (1-25)
Muscular signs	
DHS*	6/6
Severe deficit of limbs (≤ 2/5)	6/6
Extra muscular signs	6/6
Joints	2/6
Skin	5/6
Otolaryngology	3/6
Cardiac	2/6
Pulmonary	2/6
General	5/6

DHS= dropped head syndrome

All patients with DHS had a muscle deficit of less than or equal to 2/5 on the Medical Research Council (MRC) scale in the proximal muscle groups according to Minimal Muscle Testing (MMT8). Extra-muscular signs were constant. The skin involvement was of the periorbital erythema type in two patients and a "mechanic's hand" appearance in another. It was nonspecific in two other cases. Dysphonia or dysphagia was noted in three patients. Joints, cardiac and pulmonary disorders concerned two patients. One patient had a thyrotoxicosis syndrome associated with bilateral reducible exophthalmos and a goiter. Ocular manifestations or significant amyotrophy were absent. Myasthenic syndrome or extra pyramidal syndrome were not found in any case. Figure 1 illustrate a patient of our series with dopped head syndrome.

Figure1. *patient with Grave's disease presenting a dropped head syndrome*

The creatinine phospho-kinase level varied from 2.3 to 358.4 times normal (normal < 165 UI/L). The blood ionogram revealed an undetectable potassium value in one patient and was normal in the others. An inflammatory syndrome was found in four patients. The muscle biopsy performed in one patient was non-contributory.

Potassium supplementation was initiated in the treatment of hypokalemic myopathy.

Corticosteroid therapy was proposed in the other cases, except in one patient who died before the start of corticosteroid treatment. Methotrexate was started in one patient in combination with motor physiotherapy.

The evolution was marked by the death of three patients from cardiac or respiratory complications. It was favorable in two others. One patient was lost to follow-up. Table 2 shows the clinical, etiological, therapeutic, and evolutionary data associated with DHS in our series.

Table2. *clinical, etiological, therapeutic, and evolutionary data of our cases series*

Cases	Associated Manifestations	Etiologies	Treatment	Evolution
1	Myogenic syndrome Periorbital erythema Skin ulcers Myocarditis Fever	Dermatomyositis	Corticosteroids	Death
2	Myogenic syndrome Arthritis Péricarditis Interstitial pneumopathy Dysphagia Palmar hyperkératosis Fever	Anti-synthétase syndrome	Corticosteroids	Death
3	Myogenic syndrome Periorbital erythema Photo exposed erythema Dysphagia Dysphonia Fever	Dermatomyositis	Corticosteroids	Lost to follow up
4	Myogenic syndrome Thyrotoxic syndrome Exophtalmos Goitre Mélanoderma	Thyrotoxic hypokaliemic paralysis (Basedow)	Potassium	Favorable
5	Myogenic syndrome Arthritis Interstitial pneumopathy Sclérodactylia Hypochromic macules Fever	Scléromyositis	Corticosteroids Methotrexate	Favorable
6	Myogenic syndrome Dysphagia Fever	Unclassed myositis	No pécific treatment	Death

Discussion

We reported six observations of DHS. This is a rare clinical situation and its multiple etiologies can be neurological, neuromuscular, muscular and sometimes unidentified [1, 6, 7, 8]. The term isolated neck extensor myopathy (INEM) is used for isolated forms with no other neuromuscular abnormality found [8]. DHS reflects a deficit of the neck extensor muscles. This syndrome is more frequently encountered in elderly subjects with a female predominance [1]. However, in our series, a young age characterized our study population, all of whom were women.

The etiologies of DHS are dominated by neurological pathologies (Parkinson's disease, amyotrophic lateral sclerosis), neuromuscular pathologies (myasthenia gravis) and the isolated form, the other causes being rarer [1, 5, 6, 7]. This contrasts with the data from our series where inflammatory myopathies are the main cause found (five cases out of six). No cases of neurological or neuromuscular pathology were found in our series. One case of hypokalemic paralysis secondary to hyperthyroidism was noted. In a recent review, the diagnoses associated with DHS were dominated by amyotrophic lateral sclerosis and neuromuscular junction disorders, notably myasthenia gravis, while inflammatory myopathies were less frequently involved [2]. Their association with this syndrome was found in the literature in the form of case reports [9, 10]. In a recent review of 107 cases of DHS, only 16 were associated with an inflammatory myopathy [11]. This difference could be related to the exclusive recruitment of our patients in an internal medicine department with the almost constant presence of extra-muscular manifestations pointing to a systemic pathology. A recent study of DHS in inflammatory myopathies showed that this syndrome was associated with late age, the presence of extra-muscular signs, a limb deficit in most cases, scleromyositis and inclusion myositis [12]. Other publications have reported an association of DHS with scleromyositis [3, 4]. African series on inflammatory myopathies have not reported an association with DHS [13, 14, 15, 16]. A clinical case of association of DHS with scleromyositis has been reported by an African team [17].

Hypokalemic thyrotoxic paralysis found in one of our patients is also a rare complication of hyperthyroidism, especially in black subjects, like our population [18].

In our series, corticosteroid therapy was initiated in most cases and an immunosuppressant was added in one patient. In the literature, medical treatment based on corticosteroids with or without an immunosuppressant is the standard treatment in association with rehabilitation [1]. Surgery, used as a second-line treatment, remains a therapeutic option that has not benefited from enough experience, with results varying according to the series, positive in some and poor in others [1, 6, 8].

The evolution was unfavorable in half of the cases in our series. Deaths were mainly related to cardiac or respiratory complications. In the study of DHS in inflammatory myopathies, except for those with inclusion myositis, all patients received treatment with corticosteroids and immunosuppressant. Half of them also received intravenous immunoglobulin. In this study, the course was favorable in the vast majority of cases but was worse in cases of inclusion myositis. One patient had cancer before the diagnosis of myopathy and one patient died of ischemic cardiomyopathy [12].

In a publication reporting five cases of this syndrome associated with inflammatory myopathies, the presence of DHS was associated with severity of myopathy with the occurrence of two cases of respiratory distress [19]. In this study, DHS was associated with dermatomyositis (three of five cases) and overlapping myositis (one case with scleroderma and one case with Sjogren's syndrome) [19]. In contrast to our series, the evolution was favorable in all cases under combination of corticosteroids, immunosuppressant, and intravenous immunoglobulins, while noting that corticosteroid treatment alone was ineffective [19].

This series gave us a better insight into DHS, which had not previously been studied in our setting. We studied the etiologies associated with this syndrome as well as its prognosis in an internal medicine department.

Thus, this series constitutes a study that should be extended to a larger number of patients.

Conclusion

DHS, in our recent experience, is associated with female gender, young age and severe muscle deficit.

Inflammatory myopathies were etiologies the most of time. The prognosis was guarded with cardiac and pulmonary complications.

Collaborative studies are needed to better define semiological and prognostic value of DHD in our regions.

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