# Aetiology of Proximal Weakness among Adult Sudanese patients

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**Abstract**

**Objective**: To determine the aetiology of proximal myopathy among adult Sudanese patients seen in Elshaab Teaching Hospital.

**Methods:** This is a descriptive cross sectional hospital based study conducted in Elshaab Teaching Hospital, during the period from January 2004 – September 2005. 100 adult Sudanese patients with proximal myopathy were reviewed, detailed history and proper clinical examination was done by the authors.

**Results**: The most frequent cause of proximal myopathy was found to be muscular dystrophy which accounted for 30% of the cases, followed by myasthenia gravis 20%, polymyositis and dermatomyositis 14%, Guillain Barre 8%, diabetes mellitus 5%, connective tissue diseases 5%, thyrotoxicosis 3%, chronic renal failure 3%, malignancy 2%, drugs (steroids and chloroquine) 2%, alcohol 2%. Spinal muscular atrophy, hypokalaemia, and hypocalcaemia each accounted for 1%.

**Conclusion:** The study showed that the incidence of proximal myopathy is more common among females. Proximal muscle weakness involved the lower limbs more than the upper limbs.

Key words: myopathy, Guillain Barre, dermatomyositis, polymyositis, hypokalaemia, atrophy.

Myopathy is a disease of striated muscle. Clinically the patient generally experiences weakness or complete paralysis of the affected muscle. Acute inflammatory myopathies can be part of the manifestations of infections[1]. Acute inflammatory polyneuropathy, Guillain Barre Syndrome, porphyria, diphtheria usually have sub acute onsets. Myasthenia Gravis, severe polymyositis and dermatomyositis must be considered in the differential diagnosis of subacute weakness of muscles. Weakness from endocrine disorders and certain muscle toxins may also develop sub acutely. Proximal weakness evolving over months may be a part of manifestations of polymyositis, dermatomyositis or endocrinopathy[2]. When the course has extended for a year or more, muscular dystrophies, spinal muscular atrophy, and myasthenia gravis has to be suspected.

**OBJECTIVE**: To study the etiology of proximal myopathy among adult Sudanese patients attending neurology department in EL Shaab Teaching Hospital Khartoum Sudan in the period from January 2005 to December 2005.

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**METHODOLOGY**: This is a descriptive cross sectional hospital based study .It was conducted at Elshaab Teaching Hospital, Khartoum Sudan which is a 240 bedded hospital with cardiology, respiratory, neurology and an intensive care units. The study population included adult Sudanese patients with proximal weakness referred to the hospital from different parts of Sudan. One hundred patients were included in the study. Detailed history and proper clinical examination was done. Urine analysis, complete haemogram, blood urea, serum potassium, CPK, muscle biopsy, thyroid function test, F.B.S, P.P.B.S., Tensilon test and EMG were done.

**DATA ANALYSIS**: Data were analyzed using Statistical Package for Social Science (SPSS). The results were expressed as tables.

**RESULTS**: Out of 100 patients included in the study 60% were females. The majority of the patients were younger than 45 years [Table 1]. Facial muscle weakness was found in 4%, dizziness in 6%, convulsions in only 1%, whereas fever, skin rash, polyuria and polydepsia were found each in 5% of patients. Rest of the symptoms was shown in table2.

Table 1: Relationship between sex and age in 100 Sudanese patients with proximal myopathy.

| Age | Female | Male | Frequency | % |
|---|---|---|---|---|
| 16 – 25 | 16 | 14 | 30 | 30 |
| 26- 35 | 09 | 08 | 17 | 17 |
| 36 - 45 | 13 | 02 | 15 | 15 |
| 46 – 55 | 10 | 07 | 17 | 17 |
| 56 – 65 | 07 | 06 | 13 | 13 |
| > 66 | 05 | 03 | 08 | 08 |
| Total | 60 | 40 | 100 | 100 |

Table 2: Some of the symptoms of the patients:

| Symptoms | Frequency | Percentage |
|---|---|---|
| Visual disturbance and diplopia [each] | 09 | 09 |
| Dropping of upper eyelid | 13 | 13 |
| Difficult mastication and swallowing [each] | 22 | 22 |
| Nasal regurgitation | 12 | 12 |
| Abnormal speech | 14 | 14 |
| Frequent falls | 66 | 66 |
| Not able to get up from sitting | 86 | 86 |
| Use his hand to climb up himself when getting up | 29 | 29 |
| Not able to comb hair | 85 | 85 |
| Not able to lift objects | 78 | 78 |
| Not able to climb stairs | 80 | 80 |
| Muscle pain | 14 | 14 |
| Joint pain | 13 | 13 |
| Weight loss | 80 | 80 |
| Loss of appetite | 25 | 25 |
| Constipation | 24 | 24 |

Myasthenia gravis and limb girdle muscular dystrophy were the most frequent myopatheis among our patients. Backer disease, diabetes mellitus and connective tissue disease affect 5% each while Facio-scopuls-humeral dystrophy, Spinal muscular atrophy, Hypokalaemia and Hypocalcaemia were found each in only 1% of the patients. Rest of the diagnoses was shown in table 3.

Table 3: Causes of proximal myopathy among 100 Sudanese patients.

| **The diagnosis** | **Frequency** | **Percentage** |
|---|---|---|
| Duchenne | 04 | 04 |
| Limbgirdlemusculardystrophy | 19 | 19 |
| Motor neuron disease | 04 | 04 |
| Myasthenia gravis | 20 | 20 |
| Guillain Barre syndrome | 08 | 08 |
| Polymyositis and dematomyositis [each] | 7 | 07 |
| thyrotoxicosis and CRF [each] | 03 | 03 |
| Malignancy, drugs and alcohol [each]. | 02 | 02 |

The study showed that 79% of our patients had chronic symptoms. Family history of proximal myopathy was detected in 19% of the studied group. The vast majority of the patients had normal sensations and coordination [table 4]

Table 4: Upper and lower limbs examination findings:

| Clinical finding | Frequency in upper limbs | Frequency in lower limbs |
|---|---|---|
| Fasiculation and tremors [each] | 03 | 03 |
| Wasting | 15 | 15 |
| hypertrophy of deltoid muscle | 09 | 09 |
| hypotonia | 38 | 36 |
| Hypertonia | 02 | 00 |
| Hyperreflex | 02 | 02 |
| Hyporeflex | 34 | 34 |
| normal sensation | 95 | 93 |
| Normal coordination | 85 | 69 |

Associated skeletal deformities (Kyphosis, Scoliosis, and Kyphoscilosis) were seen in 10% of the patients, where as 19% had winging scapulae due to weakness of serratous anterior, pectoralis minor, trapezius muscle resulting in instability of scapular and shoulder joint.

Creatine kinase was high in 66%, RBS> 126 in 6% while potassium and calcium were low in 1% of the patients. Thyroid function test consistent with thyrotoxicosis was seen in 3% and tensilon test was positive in 20%. Hgh CSF protein was found in 4%. EMG showed myopathic changes in 92%, muscle biopsy revealed an evidence of myopathy in 46%,

The frequency of diagnosis according to classification of muscle diseases showed that 30% of our patients had genetic disorders, 20% neuromuscular (myasthenia), 22% inflammatory disorders, 16% metabolic disorders, 5% autoimmune and 7% had other causes.

**DISCUSSION:**

The study showed that the incidence of proximal myopathy is more common among female, this is probably due to the fact that diseases like myasthenia gravis, thyroid diseases, polymyositis and dermatomyositis, which constituted 37% of all cases with proximal myopathy in the study, are more common among female. Almost 95% of patients with myasthenia gravis, thyrotoxicosis, dermatomyositis and dermatomyositis were females; this is similar to the results reported elsewhere[3-5]. The peak incidence was observed in individual aged 16 -45 accounting for 62%, which is the most active sector of the community, this goes with what was mentioned in the literature[6]. Patients with proximal myopathy showed chronic onset of presentation in 79%, this may be because most of the causes of proximal myopathy like muscular dystrophy, and myasthenia gravis have chronic and progressive course[7-8]. Some diseases presenting with proximal myopathy like myasthenia gravis and thyroid diseases have increased incidence among relatives of the patients[6, 9]. This is supported by the finding of family history of similar condition in 18% of our patients. Unlike what was found in UK and USA, most frequent causes of proximal myopathy in our population were muscular dystrophy followed by myasthenia gravis, polymyositis and dermatomyositis[10,11]. Wide variety of clinical presentations of proximal myopathy were encountered including frequent falls [66%] which was similar to what was mentioned in the literature[12]. In consistent with other reports proximal muscle weakness involving the pelvic girdle was seen in 86%, where as that involving the shoulder girdle was seen in 70%[1,2,10]. Myopathy with muscle pain seen in all patients with polymyositis and dermatomyositis was similar to a study done by Dalakes and Narayana[13,14]. The classical presentation of limb girdle and Fascioscabulohumeral muscular Dystrophy which was seen in 10% and 19% of our patients respectively differs from what was reported by Panegyres who found a lesser percentage skeletal deformities and winging of scapulae. This can be explained by the fact that limb girdle and Fascioscabulohumeral muscular Dystrophy constitute 20% of our patients[15]. Proximal

myopathy attributed to abnormal gate in 66%, while 20% were completely bed ridden due to severe weakness. This contrasts the findings reported by others[15]. However, it can be explained by the late presentation in our patients. Similar to earlier reports, CPK was found to be high in 66% of the study group including all patients with Duchenne and Beaker muscular dystrophy[1,12]. The higher characteristic EMG features seen in our patients with proximal myopathy [92%] when compared with other's [55%], is probably due to the late presentation of our patients[4]. Similar to Christopher findings, muscle biopsy supported the diagnosis of muscular dystrophies in 46% of the patients[5].

## CONCLUSION:

The study showed that the incidence of proximal myopathy is more common among females. Proximal myopathy has a wide range of causes and had involved the lower limbs more than the upper ones. Muscular dystrophy, myasthenia gravis and limb-girdle muscular dystrophy had dominated the clinical presentation.

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