Dropped head syndrome (DHS) is characterized by severe weakness of the cervical paraspinal muscles that results in the passively correctable chin-on-chest deformity. DHS is most commonly associated with neuromuscular disorders.

**Patient and Methods**

A 72-year-old man complained of sudden onset of neck pain followed, after a 15 day period, by weakness of neck extensor muscles and dysphagia causing ab-ingestis pneumonia. Since many years he presented a nuanced gait disturbance and diffuse muscle wasting.

Neurological examination showed the presence of dropped head and weakness/atrophy of the upper limb proximal muscles (MRC 4-/5). EMG analysis showed a mixed neurogenic and myogenic pattern at both the upper and lower limbs. Deltoid muscle biopsy was performed.

In the next two months, the patient experienced a gradual spontaneous improvement until he was able to hold his head upright again and no longer complained of dysphagia.

**Results**

Deltoid muscle biopsy showed severe myopathic changes with prominent features of mitochondrial respiratory chain dysfunction (over 80 COX-negative fibers), many fibers with subsarcolemmal accumulation of mitochondria and ragged blue fibers. Rare small inflammatory infiltrates, scattered fiber atrophy and increase in lipid droplets were also observed. No MHC I expression in muscle fibers was present.

The sequence analysis of the 22 tRNAs in mtDNA did not show any variation. The remaining mtDNA sequencing and respiratory chain biochemical assay are ongoing.

**Discussion**

Dropped head has been frequently reported in a wide range of neuromuscular disorders, including Motor Neuron Disease, Polymyositis and Myasthenia Gravis. It has been only rarely described in mitochondrial myopathies and it is quite infrequent as a presenting feature.

Muscle biopsy in our patient showed a picture suggesting a primary mitochondrial myopathy and the degree of respiratory chain impairment seems to exclude age-related mitochondrial changes.

This is the first reported case of DHS in a patient with a mitochondrial myopathy having a spontaneous recovery. Spontaneous recovery in metabolic myopathies is very unusual. We may assume a temporary respiratory chain failure in presence of an increased energy demand due to an intercurrent unknown event.

It is well known that axial muscles have higher metabolic requirement because of their role in postural maintenance.

**Conclusions**

Our report should induce to consider mitochondrial myopathy in the differential diagnosis of dropped head syndrome. The role of mitochondrial abnormalities and the mechanisms underlying spontaneous recovery in this patient remain to be explained.