Cervical Dystonia Mimicking Dropped Head Syndrome

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OBJECTIVES

Dropped Head Syndrome (DHS) is a rare condition characterized by severe weakness of the neck extensor muscles associated with kyphotic deformity of the cervico-thoracic spine. Many diseases may be linked with DHS and differential diagnosis is often complex.

METHODS AND MATERIALS

We describe the case of a 58-year-old man with a history of dropped head since one year. On clinical examination we found a severe cervical kyphosis (Figure 1), weakness and hypotonia of the neck extensor muscles, and bilateral ptosis (Figure 2) (present since when he was adolescent). The cervical posture could be easily corrected by passive extension of the head (Figure 3).
He underwent complete blood test, cervical magnetic resonance imaging (MRI), electromyography (EMG) and muscular biopsy.

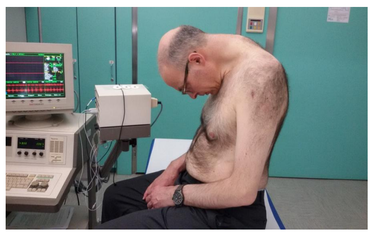


Figure 1 – Severe cervical kyphosis

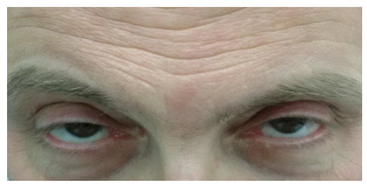


Figure 2 – Ptosis

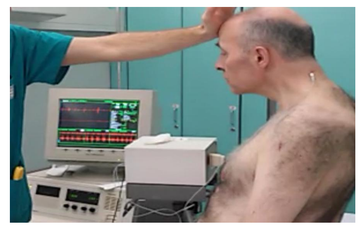
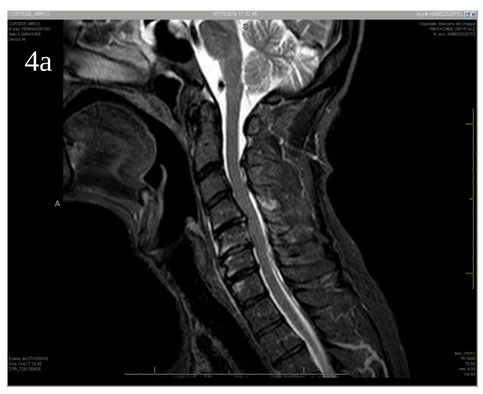


Figure3 – Correction of cervical posture with passive extensione of the head

RESULTS

Blood test revealed: slight increase of CPK value (363 U/L), acetylcholine receptor antibodies test was negative.
MRI showed spondylotic alterations, reversal of the physiological lordosis C4-C5 with reduction of the spinal canal diameter without myelopathy (Figure 4a), mild muscular atrophy and fat infiltration in the long neck muscles were observed (Figure 4b)
EMG revealed myopathic signs on the long neck muscles and fibrillation on deltoid muscles; repetitive nerve stimulation was normal. Kinesiological EMG showed persistent contraction of the sternocleidomastoid and scalene muscles upon active neck extension (Figure 5).
The biopsy of the deltoid muscle showed only rare type 1 transition fibers.
No response was observed after combined treatment with pyridostigmine and prednisone.
Treatment with botulinum toxin type A (BTX), on the anterior scalene and sternocleidomastoid muscles, resulted in an important clinical improvement (Figure 6a e 6b) , followed by transient disappearance of the contraction of the mentioned muscles. EMG analysis showed an electromyographic improvement (Figure 7).



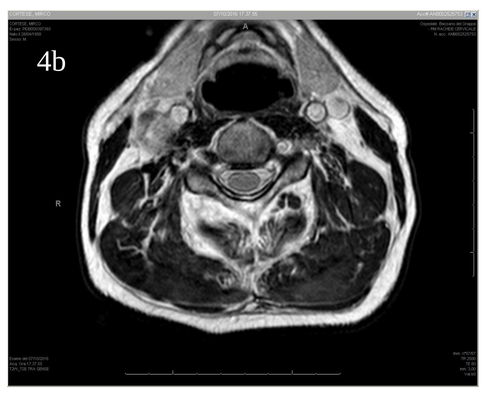
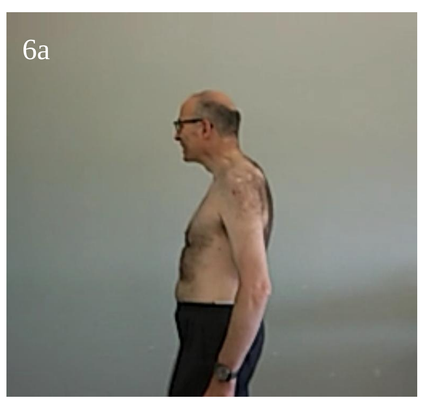


Figure 4 – MRI:
a) reduction of spinal canal diameter
b) mild muscular atrofy with fat infiltration



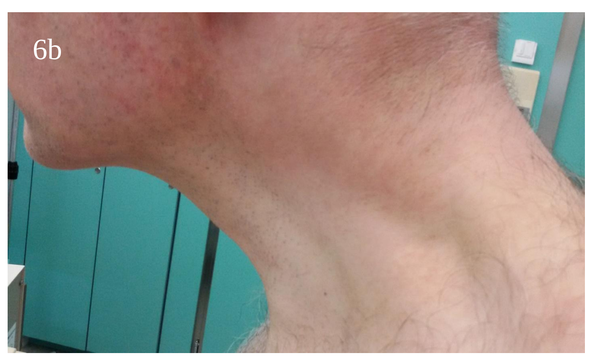


Figure 6a e 6b – Improvement of posture of the neck e reduction of muscle contraction after botulinium toxin treatment

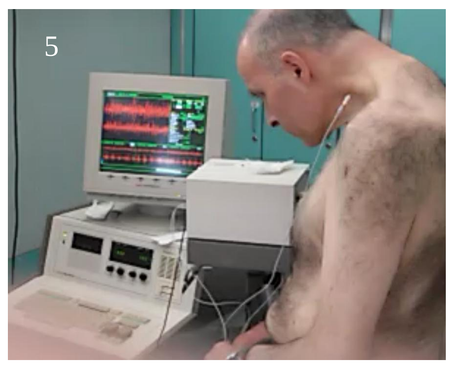


Figure 5 – Contraction persistent of scalene muscle during neck extension

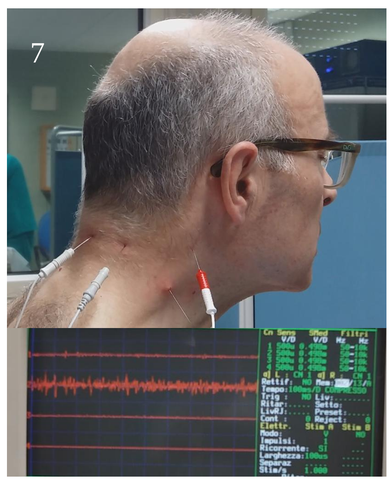


Figure 7 – Electromyographic improvement after treatment with BTX

1 ch splenius
2 ch ext neck
3 ch scalenus
4 ch scm

DISCUSSION

Our findings of myopathic signs, muscle dystonia and cervical spondyloarthrosis are part of the typical pattern for DHS. The muscular biopsy did not show significant myopathic findings, neither the EMG neither the MRI were compatible with a myopathic involvement. The hypothesis of a myopathy is thus to be excluded.
Motor neuron disease was considered, but it is unlikely because of the following findings: lack of involvement of other muscular segments, no pyramidal signs, lack of chronic reinnervation on EMG, biopsy results and the treatment response with BTX.

CONCLUSIONS

The primary cause of DHS in our patient is likely due to cervical spondyloarthrosis, that induced focal dystonia, leading to the myopathic pattern, compatible with neurophysiological hypotrophy. Similar hypotheses have been suggested in the literature. Clinical follow-up will be crucial to monitor disease progression and better understand of its pathophysiology.

REFERENCES

[1] AR Martin, R Reddy, MG Fehlings, Dropped head syndrome: diagnosis and management, EB Spine Care Journal, 2011, vol 2, 41-47.
[2] A Rahimizadeh, HF Soufiani, S Rahimizadeh, Cervical spondylotic myelopathy secondary to Dropped Head Syndrome: report of a case and review of Literature, Case Reparts in Ortopedics, 2016, 1-7.