Two cases of neck extensor myopathy responding to IGIV

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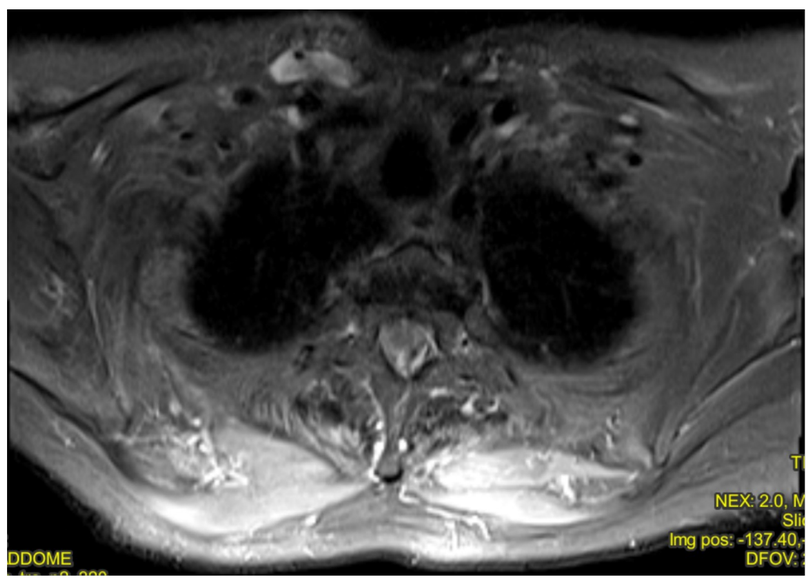
BACKGROUND: The dropped head syndrome is a condition in which patient develop a myopathy affecting only or predominantly neck extensor muscle. The cause is not clear but can be associated with different diseases that can be classified in Neurological (Amyotrophic lateral sclerosis, Parkinson disease, Cervical myelopathy, CIDP, Myasthenic syndrome) Muscular inflammatory (Polymyositis, Scleromyositis) and non-inflammatory (Nemaline myopathy, Inclusion body myopathy, Mitochondrial myopathy, Fascioscapulohumeral dystrophy) and secondary myopathy to internal disease . The cause of this condition is still unclear, and the therapy is not defined.

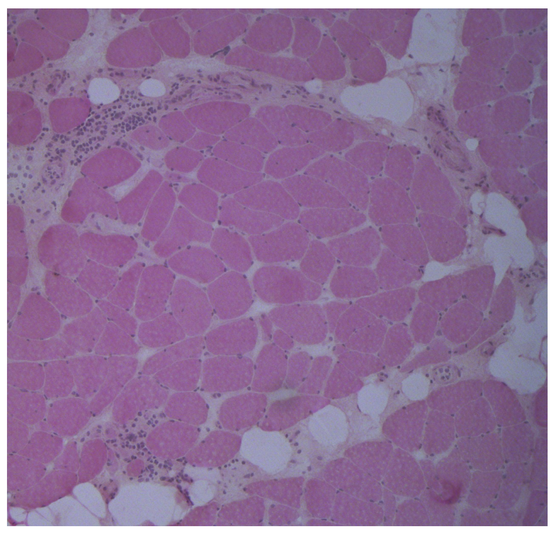
OBJECTIVE: Describe two patients with neck extensor myopathy and secondary proximal arms weakness, responding to immunomodulating treatments.

METHODS AND PATIENTS: The patients present a history of progressive subacute weakness of neck extensor muscles and subsequent onset of proximal arms weakness. Both subjects underwent: routine laboratory studies (all in normal range) electrodiagnostic testing (ENG and repetitive nerve stimulation negative), anti-AChR and anti-MUSK antibody test (negative) and a total body TC (negative), in order to exclude the most frequent causes of dropped head.

Patient 1:
A 67 years old woman whose exams showed a moderate hyperCKemia (403 U/L), positive ANA (> 1:320) and ANCA P, altered EMG (presence of positive potential and fibrillation potential in left splenius and bilateral cervical paraspinal muscles) and muscle MRI (atrophy of the paraspinal muscles). The biopsy (deltoid muscle) was characterized by active inflammatory signs with chronic degeneration).



Patient 2:
A 68 years old woman underwent the dosage of CPK (418 U/L), ANA (> 1:320), ANCA P (Positive), PR3 ANCA (3,15 RU/ml), cervical MRI (diffuse spondylarthrosis), EMG (signs of myogenic damage), muscle MRI (hyperintensity of the trapezius and atrophy of paraspinal dorsal muscles) and biopsy of brachial biceps (modest signs of muscle impairment).



RESULTS: we observed a strength improvement in both patients after the first cycle of intravenous immunoglobulin. The patients were able to keep the head upright, with an increase of, at least, one point on MRC scale. The first patient requires a maintenance dose of 0,2 g/kg every month, while the second patient performs monthly infusions at the full dose of 0,4 g/kg.

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