VIDEO CASE SOLUTION

Solution: ALS

Last month's case presented dysarthria and positive myasthenia serology.

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A 75-year-old man presented with six-month history of speech difficulty, dysphagia, and fatigue that was more pronounced in the evenings. There was no diplopia or ptosis. He developed progressive dyspnea and lost 10 pounds over six months. Repetitive nerve stimulation of the left spinal accessory nerve revealed 15 percent decremental response. Binding AChR antibody titer was increased to 0.5nml/L. He was diagnosed with MG and was transferred to our hospital for plasmaphoresis. Examination is shown. He also had hyperreflexia and mild proximal weakness in the arms.

Mild elevation of AChR antibodies in this case:

1. Excluded the diagnosis ALS
2. Indicated coincidental MG and ALS
3. Was an artefact
4. Along with positive RNS, excluded ALS
5. Is reported in ALS

The correct answer is Number 5.

EMG was done in the hospital and revealed denervation of the tongue and upper extremities muscles and thoracic paraspinal muscles.
AChR antibody titer is about 90 percent specific for MG. False positive mild elevation is reported in patients with

- Thymoma without MG.
- Family history of MG.
- Exposure to snake toxin
- ALS.

The patient in this case met the diagnostic criteria of ALS, and the course of the disease was as expected for ALS. The presence of AChR antibodies did not change the prognosis.
These antibodies suggest neuromuscular junction involvement in ALS. Decremental response occurs in 15 percent of ALS patients due to dysfunction of presynaptic calcium channels.
Although antibody titer is usually below 0.5, titers as high as 50nmol/L nm are reported in ALS.

Okuyama Y, Mizuno T, Inoue H, Kimoto K. Amyotrophic lateral sclerosis with anti-acetylcholine receptor antibody. Intern Med. 1997 Apr;36(4):312-5.
Thomas M, et al.False positive immunoassay for Acetylcholine receptor antibody in ALS. NEJM. 1980;302;868.