LETTER TO THE EDITOR

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Myotonic Discharges in Anti-MuSK Myasthenia

Keywords: Myasthenia, Anti-MuSK, Myotonia

A 63-year-old woman of Malaysian–Chinese origin presented with subacute progressive dysarthria and dysphagia. She had a history of left stage II Her2-positive breast cancer with metastasis only to an adjacent lymph node, treated with mastectomy, chemotherapy, and radiation the year before presentation. She was only on Herceptin at the time of neurological symptoms, which was discontinued shortly thereafter. She was not on a statin. Over the subsequent 8 months her dysphagia progressed, requiring gastrostomy tube placement. A trial of pyridostigmine was negative. She developed dyspnea and required nocturnal ventilator assistance, but was not intubated. She received a course of intravenous immunoglobulin, without benefit.

MRI brain was performed twice, 7 months apart, and was normal. CT of the chest, abdomen, pelvis, and neck was unremarkable. Bloodwork, including B12, thyroid-stimulating hormone, and creatinine kinase, was normal. Acetylcholine receptor antibody (AchRAb) testing was negative, including LGI1 and CASPR2. CSF examination was normal. Needle electromyography (EMG) demonstrated myotonic discharges in the left extensor indicis and tibialis anterior, and repetitive stimulation of ulnar nerve recording from abductor digiti minimi was normal. Muscle biopsy of the right tibialis anterior showed type 1 muscle fiber atrophy without other abnormality. Genetic testing for myotonia congenita, and paramyotonia.5 However, these disorders were ruled out, as were other plausible causes of myotonia.

On presentation to our institution 1 year after symptom onset, she had diffuse cachexia without fasciculations, and dyspnea at rest. Speech was unintelligible owing to flaccid dysarthria, and the patient communicated in writing. She denied any fluctuation throughout the day or over time. She had bilateral ptosis, worse on the left, with fatigability. She had bilateral adduction paresis and horizontal diplopia in all directions of gaze. She had bilateral facial weakness, and electrical myotonia. Testing for anti-MuSK myasthenia should be considered, in order to make a prompt diagnosis, and critical revision of the manuscript.

The authors have nothing to disclose.

STATEMENT OF AUTHORSHIP

RAJ was involved in patient management and writing of the manuscript. GI, AG, and CDK contributed to patient management and critical revision of the manuscript.

SUPPLEMENTARY MATERIALS

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REFERENCES