PLASMA-EXCHANGE DELAYED PSEUDOCHOREOATHETOSIS IN A PATIENT WITH ANTI-HU NEURONOPATHY

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BACKGROUND

Pseudochoreoathetosis refers to abnormal writhing movements of distal portions of limbs occurring in association with impaired processing of proprioceptive information that alters integration of sensory-motor signals. Any lesion along proprioceptive pathways from muscle spindles and joint receptors to primary somatosensory cortex may produce pseudochoreoathetosis. Nevertheless, pseudochoreoathetosis is most often seen in peripheral neuropathies and in dorsal column or dorsal root disorders, and may occur in association with onconeural antibodies.

We report here a case with pseudochoreoathetosis which occurred several years after the onset of anti-Hu sensory neuronopathy and improved after plasma exchange (PEX).

CLINICAL FEATURES

A 65-year-old woman presented with insidious onset of dysphagia, lower-limb dysesthesia and progressive gait ataxia. Her personal medical history showed hypertension, sensory hypoacusia, esophageal achalasia and recurrent intestinal pseudo-obstructions.

At first neurological examination (2003) she showed generalized areflexia, marked proprioceptive impairment, superficial sensory loss with a glove-and-stocking distribution, and preserved muscle strength. Romberg's sign was present, and the patient displayed broad-based gait and dysmetria of the four limbs.

ELECTROPHYSIOLOGICAL FINDINGS

Nerve conduction velocity study showed severe and generalized reduction of sensory nerve action potentials, associated with slight reduction of sensory nerve conduction velocity and normal motor nerve conduction velocity

NEUROIMAGING

Brain and cervical spine MRI showed dorsal column hyperintensity

BLOOD ANALYSIS

Remarkably high titer of serum **anti-Hu antibody**.

CSF EXAMINATION

Slightly increased protein concentration: 80 mg/dl (n.v. 15.00-45.00)

SCREENING FOR CANCER

Serum neoplastic markers were negative;

Whole-body CT scan and whole-body [18F]-fluorodeoxyglucose PET failed to reveal primary or metastatic neoplasms.

DIAGNOSIS

Subacute sensory neuronopathy/Denny Brown syndrome associated with anti-Hu antibodies.

CLINICAL COURSE

The patient was submitted to PEX with partial improvement of symptoms.

PEX was chronically maintained for almost 6 years, then discontinued (January 2016) due to hypoalbuminemia.

Approximately 3 months after discontinuing PEX, the patient developed involuntary, mostly slow, distal movements of the legs, worsened by eye closure and postural standing, with the features of **pseudochoreoathetosis.**

On this basis, PEX was restarted. Involuntary movements markedly reduced 3 weeks after re-initiation of PEX (5 consecutive sessions) which was chronically maintained for the next 12 months, with persistent reduction of pseudochoreoathetosis.

Electrophysiological control performed 12 months after PEX restart did not show any change of nerve conduction data.

CONCLUSIONS & TAKE-HOME MESSAGES

- Anti-Hu antibodies were present despite of the long-lasting negativity of both serum neoplastic markers and imaging search for neoplasms.
- Symptoms of pseudochoreoathetosis developed 13 years after the onset of sensory polyneuropathy, following interruption of PEX.
- PEX was followed by a marked improvement of psedochoreoathetosys, but not by an improvement of neurophysiological data.
- These features most likely suggest progression of deafferentation with pathologic involvement of dorsal root ganglia occurring as the consequence of the pause of immunotherapy. Further studies are advised, however, to define the mechanisms that may trigger delayed symptoms in anti-Hu neuronopathies.

References

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