Stiff-arm syndrome

Figure Limited range of right arm movements

The patient demonstrates the extent of her range of movements when asked to abduct and extend the arm. The posture of shoulder adduction and elbow flexion was minimally overcome by passive manipulation.

A 56-year-old woman experienced progressive stiffness and painful spasms in the right arm leading to a frozen arm within 2 years. She had previously developed cerebellar ataxia (anti-glutamic acid decarboxylase [GAD] antibodies, 1,550 nmol/L; age 52 years). Examination showed marked rigidity and decreased movement range of the right arm with generalized hyperreflexia (figure and video). There were no anti-amphiphysin or other paraneoplastic antibodies. Search for malignancies was negative. Diabetes was ruled out. The patient benefited from diazepam and IV immunoglobulin. Stiff-person syndrome (SPS) affects predominantly the trunk and lower limbs.¹ This case illustrates that the cerebellar ataxia-SPS spectrum of anti-GAD autoimmunity may occur sequentially rather than concurrently, and a stiff arm, a variant of paraneoplastic SPS,² may rarely be a focal form of anti-GAD-mediated (nonparaneoplastic) SPS.

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