**INTRODUCTION**

Stiff-person syndrome (SPS) is a rare autoimmune neurological disorder characterized by rigidity of axial and proximal appendicular muscles, abnormal gait and painful spasms. It may be associated with autoimmune disorders such as insulin-dependent diabetes mellitus, Hashimoto's thyroiditis, pernicious anemia, and vitiligo; moreover, it can develop as a paraneoplastic neurological disease. It has been associated with a growing number of antibodies, such as: anti-GAD, anti-Glycine, anti-GABA-A or GABA-B, anti-Amphiphysin, anti-Ri, anti-Gephyrin.

**CASE REPORT**

-14-year-old male patient

- family members affected by amyotrophic lateral sclerosis (grandmother and maternal uncle)

- he gradually developed: difficulty in opening and closing his hands and a gait disturbance with reduced fluidity of his movements, associated with axial and proximal stiffness, cramps and legs pain. Moreover his clinical history was also characterised by extrasystoles.

-Electromyography showed continuous motor unit activity and the co-contraction of agonist and antagonist muscles.

-Creatinkinase were more than 600 IU/L.

-Brain MRI was negative for pathological findings.

-VGKC antibodies (LGII and CASPR2) were assayed, in particular anti-CASPR2, in the Isaacs syndrome suspicion, with a negative result.

-Anti-neural antibodies (including anti-Amphiphysin and anti-Ri), anti-GAD, anti-pancreatic islet cell, anti-ANA, ENA, ANCA, anti-striated muscle were negative.

-Genetic tests for ALS resulted negative.

**SPS was diagnosed resulting from clinical examination and electromyography, despite the antibodies tested negative, in particular anti-GAD65.**

The patient was treated with benzodiazepines and baclofen with mild response; subsequently he started therapy with intravenous immunoglobulins and steroids, with improvement in his clinical condition. The patient will have to undergo periodic cancer screening and cardiologic follow-up because of the frequent association of SPS with tumors and with autonomic disorders such as cardiac arrhythmia.

**DISCUSSION AND CONCLUSIONS:**

SPS is generally associated with specific antibodies, especially anti-GAD65; however we can also find cases without associated antibodies: this case report highlights the importance that clinical manifestations are essential for diagnosis of SPS.

Beside the immunology tests, electromyography is a very important diagnostic tool and reveals continuous motor unit activity that disappears during sleep and general anesthesia.

**REFERENCES**


3) Harini Sarva et al - "Clinical Spectrum of Stiff Person Syndrome: a Review of Recent Reports" - Tremor and Other Hyperkinetic Movements - 2016 - 6: 340