Cascade Filtration in Stiff Person Syndrome: case report

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BACKGROUND

Stiff-person syndrome (SPS) is a rare acquired, autoimmune neurological disorder characterized by fluctuating muscle rigidity in the trunk and limbs as well as increased sensitivity to noise, touch and emotional distress which can result in muscle spasms. SPS is often associated with autoimmune diseases including Graves' disease, Hashimoto's thyroiditis, pernicious anemia and type I diabetes mellitus. Autoantibodies reactive to 65 kDa glutamic acid decarboxylase (GAD65), the enzyme responsible for the synthesis of γ-aminobutyric acid (GABA) in the brain and pancreatic islet cells, are present in the serum in up to 90% of patients with SPS. These antibodies block GABA synthesis. The paraneoplastic form of the syndrome is associated with autoantibodies to the 128 kDa synaptic protein amphiphysin.

Treatment consists of immune therapies, anti-anxiety medications, muscle relaxants, anticonvulsants and pain relievers. High-dose IVIG (2 gm/kg per month) is effective in relieving symptoms of stiffness and spasticity, and in reducing the titer of anti-GAD65 antibodies. Other immunosuppressive treatment, such as rituximab, is often considered when traditional immune therapy and antispasmodics have been ineffective. Therapeutic Plasma Exchange (TPE) can deplete normal immunoglobulins when enough plasma volumes are exchanged in a brief period. TPE may be considered, in addition to standard drug therapy, if the patient is unresponsive to conventional therapy. Cascade Filtration (CF) is a technique used for the selective removal of specific molecules from the blood. CF consists of a first separation of the plasma from the blood through a cell separator, the plasma will then be passed through a special equipment, plasmapher Apherlungs, into a fractionator filter which will eliminate the target molecules. Small molecular weight components such as albumin, are returned to the patient.

METHODS

We present the case of 55-year-old female patient who suffering from Stiff Person Syndrome. With the aim of removing from the blood the anti-GAD antibodies that are excessively produced due to the pathology, the patient was treated with 47 standard plasma exchange procedures from 2019 to 2021; then, he had 6 CF procedures, using the Evaflux 2A20 filter (Kawasumi).

RESULTS

Blood therapy sessions were conducted every 21 days and she was treated of at least one blood volume (approximately 3.5L). The anti-GAD antibody titer (90-115 U/ml) remained constant during treatment with standard procedures, it showed a slight reduction (79 U/ml) after the 6 procedures with CF. All procedures performed were safe and had no adverse effects on the patient. The patient's outcome was positive.

CONCLUSION

The CF treatment seems to reduce the concentration of anti-GAD antibodies from the blood, was simple and safe, allowing an improvement in the patient's quality of life and a benefit on the costs of the procedures as it does not use albumin.