TREATMENT OF NEUROLOGICAL MANIFESTATIONS OF SJOGREN’S SYNDROME

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Neurological manifestations of primary Sjogren’s syndrome appear to be wide ranging. Treatment is recommended for those
with progressive neurological impairment: usually immunosuppressive treatments including cyclophosphamide, steroids, azathioprine, and anti-TNF agents.

We present three cases that didn’t respond to initial steroid treatment but subsequently improved with regular infusions of intravenous immunoglobulins. In all three cases, the initial anti-Ro test was negative, but became positive after 4–11 years, an important feature to recognise in clinical practice.

Case one presented with painful dystonic spasms of the arm and leg, hemi-facial spasm, vertigo, diplopia with skew deviation, and trigeminal neuropathy. Several years later she developed features of systemic mixed connective tissue disease. Case two presented as a facial palsy, sensory trigeminal neuropathy, and spastic quadraparesis. GAD antibody was positive but she did not clinically fit with stiff man syndrome. Case three has had relapsing inflammatory optic neuropathy since the age of 19, with no clear cause initially identified.

These cases highlight the diversity in the neurological manifestations of Sjogren’s syndrome, the importance of not relying on initial antibody testing, and the potential for improvement with intravenous immunoglobulins, a treatment that has less adverse effects than potentially toxic immunosuppressants.
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