Pediatric aPL related motor neuropathy and IVIG

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INTRODUCTION

In the last years there are an increased number of children affected by underlying autoimmune diseases associated with neurological manifestations, as the first clinical sign. In most cases the timing and the type of immunomodulant therapy is important to consider to have a successfully outcome.

We report a paediatric clinical case characterized by the onset of acute motor polyneuropathy in presence of persistent LAC positivity successfully treated with IVIG.

CASE

A healthy, nine-year-old girl progressively presented with gait disturbance, frequent falls, difficulty in jumping and rising from the ground. Neurological examination showed mild muscular weakness and absent tendon reflexes at the lower limbs, incomplete Gowers sign, difficulty walking on heels, jumping and instable static and dynamic balance.

METHODS AND RESULTS

Laboratory assessment

Autoimmunity	ASMA, AMA, ENA, ANA, anti Nucleosome, anti Centromere, anti Jo1, p ANCA/c ANCA, anti sDNA, anti GAD, anti SCL 70, anti Citrulline, Anti endomysial and anti transglutaminase, Antiganglioside and aquaporin antibodies, Cryoglobulins, Immune Complexes, C3, C4	Neg
aPL	Lupus anticoagulant, Anti β 2 glycoprotein antibodies, Anticardiolipin antibodies, Antiprothrombin antibodies	LAC + drVV ratio +
Onco-neural Antibodies	Anti Yo, Anti Ri, Anti Hu, anti CCU2, Anti AGNA, anti Me	Neg
Serum exams	Erythrocyte sedimentation rate, C-reactive protein, ASO titer, Ammonium, Serum lactate, Serum ceruloplasmin, Copper serum and urine levels, rheumatoid factor, Serum free thyroid hormones, CPK, Cromogranina A, neuron specific enolase, CEA	Neg
Anti-neuron antibodies	anti NMDAR, anti GABA b, anti Lgi-1, Anti CASPR 2	Neg
Sierology	Herpes, Cytomegalovirus, Enterovirus, Parvovirus, Epstein Barr virus, mycoplasma, TORCH, varicella, measles, adenovirus, mumps, echovirus, influenza A and B, Coxsackie, Borrelia burgdorferi, Parvovirus B19 IgM and IgG	Neg
Hereditary clotting exams	antithrombin, protein C and protein S, factor V Leiden, prothrombin 20210A gene mutation, polymorphism of methylentetrahydrofolate reductase	Neg

- ✓ Brain and medulla magnetic resonance imaging: Negative
- ✓ Electroneurography showed findings compatible with motor neuropathy

Dysimmune polyneuropathy

Intravenous immunoglobulin treatment (IVGI) at the dose of 0.4 mg/kg/day for 5 days

Rapid improvement after IVIG

IVIG treatment	Hammersmith Functional Motor Scale (HFMS)	North Star		6MWT
		10 mt	Rise from floor	
Before IVIG	56/66 (squat difficulty)	8,70 sec	9,38 sec (Incomplete Gowers)	323 mt
1 Month after IVIG	64/66 (mild diff squat)	5,65 sec No double stance	2,21 sec (High kneeling)	489 mt
2 months after IVIG	65/66 (incomplete squat)	5,52 sec with double stance	5,52 sec (High kneeling)	517 mt
4 months after IVIG	66/66 (complete squat)	3,23 sec with double stance	2,48 sec (High kneeling)	571 mt

CONCLUSIONS

This young girl developed a motor polyneuropathy in presence of persistent LAC positivity resembling a non-thrombotic clinical manifestation aPL related that are common in pediatric population. Our findings suggest the importance:

- ✓ To consider aPL antibodies as a *Reading key* underpinning a more specific dysimmune mechanism.
- ✓ To check aPL in children with a rapid onset of neurological signs in order to provide the beneficial use of IVGI in the treatment of pediatric aPL neurological conditions.

References:

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