Acta Oto-Laryngologica 2008; 128: 9, 1004-1010.
4.Karatzanis AD, Prokopakis EP, Chatzakis NS, Bourolias C, Vardouniotis A, Velegrakis GA: Microscopic endonasal surgery of nasal and paranasal cavities. Auris Nasus Larynx 2008; 35: 509-514.

Fountoulakis EN, Chatzakis N, Panagiotaki I, Karatzanis A, Lagoudianakis G, Velegrakis S, Velegrakis GA

Department of Otorhinolaryngology, University Hospital of Heraklion, Crete, Greece

Key words: isolated sphenoiditis, sphenoidotomy, endoscopic transnasal approach.

Corresponding author: Emmanouil N. Fountoulakis, Department of Otorhinolaryngology, University Hospital of Heraklion, 71 110 Voutes Heraklion, Crete, Greece. e-mail: fudulakis@tiscali.it

Chronic inflammatory demyelynating polyradiculoneuropathy in childhood and response to IVIg

Dear Editor,

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an immune-mediated acquired polyneuropathy which is rare in childhood. The prevalence of patients aged under 19 years old with CIDP has been estimated at 0.48/100.000 and it is higher in male children than it is in female ones[1]. Children with CIDP present with subacute onset of symmetric proximal and distal weakness that progress over at least 2 months. The clinical criteria for the diagnosis of CIDP include: a)progressive or relapsing motor and sensory dysfunction of more than one limb and b)hyporeflexia or areflexia that usually involves all four limbs. Moreover, children with CIDP present with elevated cerebrospinal fluid protein, diminished motor and sensory conduction velocities during electrophysiological studies and also typical demyelinating deterioration in possible sural nerve biopsy[2]. The first-line therapy is IVIg, while the alternative choices include corticosteroids, haemodialysis or immunomodulatory agents[2].

This is the case of a 3 year old boy referred in our department afebrile and in good condition. Parents reported weakness after 500 metres of walking, frequent falls, weakness when climbing a ladder. Neurological examination revealed hypotonia of lower limbs and diminished tendon reflexes. Blood tests and biochemical examinations including CPK (Creatine phosphokinase), metabolic screening (plasma ammonia, lactic acid, plasma and urine aminoacids, homocysteine, phyntanic acid), thyroid gland check and gene screening for spinal muscular atrophy were normal, while in the cerebrospinal fluid (CSF) showed raised protein (72mg/dl, normal limits<45mg/dl) and normal oligoclonal bands. Anti-GM1 IgM (Anti-ganglioside M1) serum antibodies were detected, while the rest antibodies against gangliosides (Anti GD1b, Anti-GQ1b, Anti-MAG) and rest immunologic tests (ANA, anti-DNA, ENA, ANCA, ASMA, RF, anti-AChR, anti-MuSK) were negative. Clinical examination (absence of tendon reflexes) and raised CSF protein levels raised the suspicion of a possible demyelinating polyneuropathy. Following spinal and brain Magnetic Resonance Imaging (MRI) were normal and electrophysiologic studies showed distal demyelinating polyneuropathy and confirmed the diagnosis (diminished sensory conduction velocities and prolonged distal latencies of median and ulnar nerves, conduction block in all affected nerves of upper and lower limbs, absent f wave latencies in right ulnar and left peroneal nerves). Intravenous human immunoglobulin (IVIG) in dose 0,4 mg/kg/D for 5 days was initially given, followed by tapering monthly doses of 1 g/kg for 10 months. The patient had a very good response to treatment with IVIg, with acute improvement of muscle weakness within 4 days after the administration. The clinical improvement after each administration of IVIg lasted about 4-6 weeks. The child also started physiotherapy sessions and 10 months later remains completely asymptomatic.

Children with CIDP, as in our case, have a more favorable outcome than adults, with good response to IVIg or corticosteroids[2]. In this paper we want to mention the clinical and laboratory profile of CIDP, and also the necessity of appropriate diagnosis and treatment in a timely fashion, so patients can have a favorable outcome.

References

1. Rabie M, Nevo Y. Childhood acute and chronic immune-mediated polyradiculoneuropathies. Eur J Paediatr Neurol. 2009;13:209-218.
2. Vallat JM, Sommer C, Magy L. Chronic inflammatory demyelinating polyradiculoneuropathy: diagnostic and therapeutic challenges for a treatable condition. Lancet Neurol. 2010;9:402-412.

Pavlou E[1], Gkampeta A[1], Tsaleras G[2], Tsotoulidou V[1], Athanassiadou F[1]

[1] 2nd Department of Pediatrics, Aristotle University of Thessaloniki, 'AHEPA' Hospital, Thessaloniki.
[2] 1st Department of Neurology, Clinical Neurophysiology Laboratory, Aristotle University of Thessaloniki, 'AHEPA' Hospital, Thessaloniki.

Key words: chronic inflammatory demyelinating polyneuropathy, children.
Corresponding author: Pavlou E, e-mail: eepav@yahoo.gr

ANA: Anti-nuclear antibody, **anti-DNA**: Antibody to Deoxyribonucleic Acid, **ENA**: Antibody to Extractable Nuclear Antigen, **ANCA**: Anti-neutrophil Cytoplasmic Antibody, **ASMA**: Antismooth Muscle Antibody, **RF**: Rheumatoid Factor, **anti-AChR**: Anti-acetylcholine receptor antibodies, **anti-MuSK**: Antibodies to muscle-specific receptor tyrosine kinase