A 15-year old female was referred for evaluation of a left foot drop and left lower limb muscle weakness. The symptoms had reportedly started 1 year ago, evolved over few weeks and were further accompanied with left plantar dysaesthesia and numbness, gradually including the posterior - outer part of the left calf.

Personal history included frequent episodes since infancy with fever, gastrointestinal infections and bronchitis, as well as recurrent ear infections and tonsillitis. At the age of 4 she underwent tonsillectomy and adenoidectomy. She also suffered from frequent urethritis, pharyngitis and skin infections. At the age of 7, laboratory investigation showed combined IgA and IgG hypogammaglobulinemia while the rest of the investigation was reported to be within normal limits (common variable immunodeficiency - CVID). The patient received immunoglobulin replacement therapy (0.5gr/kg/month) over a period of 6 years. During treatment she remained free of infections and all immunoglobulin class values returned to normal range.

Upon neurological examination on admission the patient exhibited reduced left ankle reflex, reduced muscle strength on knee flexion (4/5), plantar flexion (4/5) and dorsiflexion (2/5). There was reduced sensation of the dorsal surface of the foot and the outer surface of the calf. The rest neurological examination was normal. Serology including CRP, ESR and serum albumin and CSF examination including screening for viruses, showed normal findings, however, reduced IgG (684 mg/dl, reference range 751-1560) as well as increased IgE (289 mg/dl, reference range 10-100) were found. Serum free kappa chains was also reduced (471 mg/dl, reference range 629-1350).

Nerve conduction studies showed total absence of the left sural and superficial peroneal nerve action potentials. The compound muscle action potentials of the left common peroneal and posterior tibial nerve were severely reduced indicating substantial axonal loss. A mild prolongation of the distal motor latencies, as well as a minor slowing of motor nerve conduction velocities in the above nerves, were consistent with the degree of axonal loss. Electromyographic sampling showed severe denervation confined to the territory of the left sciatic nerve. MRI examination of the pelvis-hips revealed thickening of the left sciatic nerve (Figure 1).

The patient was treated with a 5 days course of intravenous immunoglobulin (IVIG), followed by a monthly single injection. After 3 months she had significant improvement regarding dysaesthesia and walking difficulty. Muscle strength increased on knee flexion (5/5), as well as on plantar flexion (5/5) and dorsiflexion (4/5).

Discussion

Hypogammaglobulinemia has rarely been linked with neuropathy [1-3]. To our knowledge, this is the second report that manifests evidence of immune-mediated neuropathy in the presence of hypogammaglobulinemia [4].
In a series of pediatric patients with sciatic neuropathy based on etiology, the most frequent cause was structural stress of the nerve, either due to trauma, operational procedures or prolonged compression / immobilization [5]. Less prominent causes include tumor and vascular events, whereas in a small number of patients the neuropathy was idiopathic. Interestingly, in 3 patients the etiology was unknown, but symptoms’ precipitation was attributed to viral infection.

Given the fact that our patient did not report previous trauma or operation on the pelvic area, traumatic etiology of sciatic neuropathy was considered unlikely. Based on the previous history of the patient, the subacute onset of symptoms and the impressive response to IVIG treatment, we consider the immune mediated mechanism (possibly autoimmunity) as the most likely scenario.

References