

Neuroboriellosis and Associated Myoclonus in a Patient with Kartegener's Syndrome

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Abstract: Kartegener's Syndrome is an autosomal recessive disease with primary ciliary dyskinesia. It has a triad of bronchiectasis, dextrocardia or situs inversus and chronic sinusitis. Lyme disease is a multisystem illness. It is referred to as neuroboriellosis if neuropsychiatric involvement is predominant. Myoclonus is a rapid and short hyperkinetic motion disorder. Its secondary etiology is located in the subset of post infectious disease. In this case report we are presenting a female patient of 38 year old known to have Kartegener's Syndrome who had myoclonic contractions. In the etiological work up, no other finding other than acute infection with *Borrelia burgdorferi* was found. Lyme disease is a rare etiological factor in involuntary movements, which directed us to prepare this case report.

Keywords: Myoclonus; Lyme disease; Neuroboriellosis; Kartegener's syndrome

Abbreviations : SEP: Somatosensory Evoked Potential; MRI: Magnetic Resonance Imaging; EEG: Electroencephalography; CT: Computed Tomography; MS: Multiple Sclerosis; ENT: Ear-Nose-Throat; HRCT: High Resolution Computed Tomography

Introduction Kartegener's Syndrome is an autosomal recessive disease with primary ciliary dyskinesia. It has a triad with bronchiectasis, dextrocardia or situs inversus and chronic sinusitis [1]. Lyme Disease (LD) is an infectious disease caused by *Borrelia burgdorferi*. This infectious agent is a spirochete transmitted by tick bites [2]. Being a multisystem disease, LD manifests itself with dermatologic, skeleton-muscular and neuropsychiatric symptoms [3]. Atypical findings can be absent in neuroboriellosis. These are, acute idiopathic polyneuritis, urinary retention, hyponatremia, sensory deficits, visual hallucinations and constipation [4]. Myoclonus is a rapid and short hyperkinetic motion disorder. It is divided into three groups according to etiology; physiological, essential and epileptic. Secondary myoclonus may result from cortical or subcortical disease. Among the secondary reasons,

postinfectious encephalitis belongs to the subset of cortical reasons [5].

In this case report we are presenting a 38 year of female patient known to have Kartegener's Syndrome who had myoclonic contractions. In the etiological work up no other finding than acute infection with *Borrelia burgdorferi* was found. Lyme disease is a rare etiological factor in involuntary movements, which directed us to prepare this case report.

Case Report A 38-year-old woman living in the village working as a farmer, admitted to the outpatient neurology clinic with myoclonic contractions which increased gradually for the last 4 years. She has been followed for Kartegener's Syndrome (bronchiectasis, situs inversus and chronic sinusitis) for 20 years. Her solid organs located in the abdomen and thorax on the opposite location. She has been followed for Bronchiectasis and pansinusitis by the Clinics of Pulmonary Medicine and ENT. She had immotile cilia and anosmia and she had been treated for Pneumonia caused by *Pseudomonas Aeruginosa*. In the ultrasound imaging of the abdomen, it was seen that the solid abdominal organs were located on the opposite side. The patient has been experiencing numbness on the right foot which was relieved by motion, right knee pain and difficulty in standing up for approximately 5 years. She also had stabbing type back pain and pain that was aggravated by deep inspiration. The patient experienced fasciculations in her body, legs and sometimes in her face which was sometimes accompanied by a brief jerk. These involuntary movements happened while she was asleep. She was admitted to a neurology outpatient clinic but no pathological finding was discovered in the EEG. She was prescribed valproic acid for involuntary movements. The patient experienced partial relief. Acute pansinusitis and otitis were the only pathological findings in the cranial CT and MRI . Her treatment was modified with the addition of levetirasetam and the cessation of valproate. The patient experienced partial relief with the new treatment as well and for the last year she had 20 daily involuntary movements all through her body. She has been experiencing knee pain and occasionally fever.

She was admitted to our clinic. Synchronized EEG and videomonitorization was utilized but no epileptiform activity was observed during the myoclonic contractions. Posterior Tibial SEP and median SEP was normally range. Extended routine laboratory workup was done. When the acute phase reactant results were analyzed retrospectively increase in CRP, ESR and WBC values secondary to chronic infection was noticed. Nothing special was noticed in Vasculitis tests, Lupus anticoagulant, ANA, ds-DNA. Her liver function tests were within the normal range. She had normal seruloplasmin levels and she had no Kaiser Flesher Ring in the ocular examination, thus Wilson's disease was ruled out. For the investigation of LD, *Borrelia burgdorferi* antigens came out to be Ig M (+) and Ig G (+) in the Elisa and Ig M (+) and Ig G (+) in the Western Blot. She was internalized in the Infectious Disease Clinic. She had no history of tick bite. Ceftriaxone 2x1 gr iv and tetracycline 2x100 mg iv treatment was initiated. Posttreatment 3rd month *Borrelia burgdorferi* antigen values were Ig M (-), Ig G (++), and the 5th month antigen values were Ig M negative, Ig G positive (+) in the Elisa. No change in the involuntary movements was observed after the antibiotherapy but the acute phase reactants decreased.

Conclusion

In conclusion, in patients presenting with atypical symptoms like progressive myoclonic jerks, extensive muscle and joint pain infectious and treatable etiologies like LD could be thought in the differential diagnosis.

Informed Consent

Written informed consent was obtained from patient who participated in this study