Case Reports

A Young Female Patient of Neuromyelitis Optica Presenting with Hypocalcemic Tetany

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Abstract

Neuromyelitis optica, a variant of multiple sclerosis, presenting with hypocalcemic tetany is an unusual presentation. We report here a case of 25 years old female who was a case of neuromyelitis optica and had hypocalcemic tetany as the initial presentation among others. The case is interesting in that the hypocalcemic tetany was not coincidental. The patient had low vitamin D status and probably, this was correlated etiologically to neuromyelitis optica. Vitamin D has immunomodulatory effect and low vitamin D status has been implicated in the etiology of autoimmune diseases such as multiple sclerosis, rheumatoid arthritis, insulin-dependent diabetes mellitus, and inflammatory bowel disease.

Introduction

Neuromyelitis optica (NMO), also known as Devic’s disease or Devic’s syndrome, is an autoimmune, inflammatory disorder in which a person’s own immune system preferentially attacks the optic nerves and spinal cord producing optic neuritis and myelitis. The patient in our case report had low vitamin D status. Vitamin D has immunomodulatory effect and low vitamin D status was probably one of the factors that precipitated the attack of NMO in our case report.

Case Report

The patient, a 25 year old female, a housewife who had been married for the last two years had a history of acute onset, painless, blurring of vision involving right eye in October 2009. Left eye was apparently alright. She was put on methylprednisolone outside to which she responded. Then in the month of November 2009, she developed subacutely, slowly progressive, daily persistent tingling sensation in all the four limbs with intermittent stiffening of the individual limbs. The episodes were precipitated by neck flexion (Lhermitte sign). Then she developed superimposed carpopedal muscle spasms which were more frequent and dramatic, and on occasions induced by hyperventilation. In the mean time, she again developed blurring of vision this time involving left eye and diplopia. Ophthalmological examination documented retrobulbar neuritis involving left eye. She was put on 3 day methylprednisolone therapy to which she responded by improvement in the vision. The carpopedal spasms, isolated limb stiffening and tingling sensation persisted occurring with more intensity and frequency.  

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frequency in all four limbs, often precipitated by neck flexion and hyperventilation. She presented to our unit on 22/01/2010, got admitted and was assessed.

Past history was negative for HTN, DM, PTB, STD, leprosy, vasculitis, substance abuse/exposure but she had a history of rheumatoid arthritis.

Neurological examination revealed normal HMF, no apparent CN palsy, normal fundus, Lhermitte’s sign +, motor deficit of 4/5 in ULs distally, tone LLs > ULs, DTRs brisk LLs, absent ULs, planters ↑↑, no sensory deficit, normal coordination and gait. Trosseau’s sign and Chevstok’s sign were negative. Other systemic examination was apparently normal.

On laboratory examination, CBC documented leucocytosis, microcytic hypochromic anemia, urine analysis documented pus cells, RFT was normal, LFT was normal, BSL was normal, serum calcium was 8.8 mg/dl, C reactive protein was positive 19.2 mg/dl, ANA positive, RA factor positive, anti-ds-DNA negative, serum vitamin B 12 was 213 pg/ml (N), serum 25-OH vitamin D3 was 14.94 ng/ml (low normal) (11.1 – 42.9 ng/ml).

CSF examination documented a clear, colorless fluid with no cobweb or coagulum, protein- 36 mg/dl, sugar- 49 mg/dl, RBCs- 100/cu. mm, WBCs- <10 lymphocytes/cu. mm, AFB/Gram stains – negative.

On neuroimaging, MRI cervical spine documented diffuse patchy demyelination involving medulla and spinal cord upto C5 level (Figs. 1, 2). MRI thoracic spine was normal. MRI brain (gadolinium enhanced) was normal.

VEP study documented P-100 latency-prolonged in left eye, normal in right eye; P-100-N-75- A MP reduced in both eyes suggestive of b/l optic axonopathy (Fig. 3).

Discussion

Neuromyelitis optica (NMO) is considered as a clinical variant of multiple sclerosis (MS). Whereas MS is a CD4+ T-cell mediated autoimmune disease, NMO is a humorally mediated autoimmune disease. The serum autoantibody marker, neuromyelitis optica-immunoglobulin G (NMO-IgG), appears specific for NMO.

Low vitamin D status has been implicated in the etiology of autoimmune diseases such as multiple sclerosis, rheumatoid arthritis, insulin-dependent diabetes mellitus, and inflammatory bowel disease. Experimentally, vitamin D deficiency results in the increased incidence of autoimmune disease suggesting a link between vitamin D levels and autoimmunity. Vitamin D is a modulator of calcium homeostasis but it also plays an important role in T cell homeostasis during the course of multiple sclerosis. Geographical variation in the incidence and prevalence of MS also has been attributed to the ‘latitude effect’ - being higher at higher latitudes and lower at lower latitudes. Ultraviolet radiation from sun is the most important source of vitamin D in most individuals, and low levels of vitamin D are common at high latitudes where sun exposure may be low, particularly during winter months.

Munger KL et al4 showed that the women who used supplemental vitamin D had 40% lower risk of MS, compared with the women who did not use vitamin D supplements.

The criteria used for diagnosis were those suggested by Wingerchuk DM et al 20065 which require two absolute criteria plus at least two of three supportive criteria being:

Absolute criteria:
1. Optic neuritis
2. Acute myelitis

Supportive criteria:
1. Brain MRI not meeting criteria for MS at disease onset
2. Spinal cord MRI with contiguous T2-weighted signal abnormality extending over 3 or more vertebral segments, indicating a relatively large lesion in the spinal cord
3. NMO – IgG seropositive status.

The patient reported in the case study therefore was a case of Neuromyelitis Optica and had a deficiency of vitamin D which probably precipitated the attack. The carpopedal spasms which were documented in our presence were probably due to hypocalcemia induced by vitamin D deficiency. By the time she reported to us, she had already been on calcium supplementation. The serum calcium level documented though was within normal limit, the serum 25(OH) vitamin D was low normal (14.94 ng/ml).

In one study,6 the mean 25(OH) vitamin D levels were 19 ng/ml during relapse and 24 ng/ml during remission of multiple sclerosis. In the findings presented by Allison Drake, a researcher with the Jacobs Neurological Institute at the State University of New York (SUNY) Buffalo School of Medicine and Biomedical Sciences, at the Consortium of Multiple Sclerosis Centers

Fig. 3: VEP study showing P-100 latency: prolonged in left eye, normal in right eye. P-100 – N-75: amplitude reduced in both eyes → Bilateral optic axonopathy.
First Crimean-Congo Hemorrhagic Fever Outbreak in India

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Abstract
Crimean-Congo hemorrhagic fever (CCHF) has not been reportedly previously from India. Initial clinical features of dengue fever and CCHF are similar and it is very difficult to differentiate and diagnose CCHF. Common clinical features of CCHF include; high grade fever with chills, headache, body ache, myalgia, vomiting, abdominal pain, weakness and bleeding from multiple sites. Laboratory investigations showed cytopения, raised prothrombin time (PT) and activated partial thromboplastin time (aPTT), raised creatinine phosphokinase (CPK) and lactic dehydrogenase (LDH) as well as altered liver and renal functions. Patients with above symptoms can rapidly progress to bleeding from multiple sites and death compared to dengue fever. It is crucial to recognize CCHF at early stage to institute ribavirin treatment and also to prevent nosocomial spread of disease to health care workers. We are describing first four cases of recent CCHF outbreak in Ahmedabad.

Introduction
Crimean-Congo hemorrhagic fever (CCHF) virus is an enveloped RNA virus of the family Bunyaviridae (genus Nairovirus) and often causes severe viral hemorrhagic fever. It is transmitted either through bite of the tick vector, mainly Hyalomma spp., or via direct contact with blood or tissues of viremic animals or humans.⁷⁻¹² Human-to-human transmission is believed to be occurring, although the infectivity of the virus by ordinary contact appears to be very low.⁴⁻⁶ History of tick bite, high-risk occupations, having contact with livestock, living in a rural area and older age are risk factors identified by investigators for acquisition of CCHF.⁶⁻⁸ Meat consumption is not usually a risk factor for CCHF. A wide variety of vertebrates like cattle, goats, donkeys, horses, etc., along with smaller wild life species like hares and hedgehogs act as a reservoir for the virus.¹ CCHF has the potential to cause community acquired and nosocomial outbreaks. Healthcare workers are at increased risk of transmission of CCHF infection while taking care of patients with CCHF. There are several reports of nosocomial outbreak with high mortality among hospital staff.⁹⁻¹²

References
7. Emma Hitt, PhD, Vitamin-D Levels Inversely Correlated With Disability and Disease Progression in Multiple Sclerosis (internet).

Case 1: Index Case
A 32years old woman, housewife, had come to Shalby hospital...