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Guillain–Barré Variant as primary presentation of systemic lupus erythematosus and class V lupus nephritis

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ABSTRACT

Any organ of the body can be affected in systemic lupus erythematosus (SLE) which is an auto immune. Patients can present with clinical features of mild joint and skin involvement to life-threatening kidney, hematologic, or central nervous system involvement. Peripheral neuropathies in SLE is not an uncommon presentation but Guillain–Barré syndrome (GBS) is considered as an unusual and one of the least neuropsychiatric syndromes in SLE. In this case report we are aiming to report a rare association of GBS as an initial presentation for an SLE in a male patient. A 46-year-old male patient present with overlapping symptoms and required admission to intensive care unit. He was diagnosed initially as case of GBS treated with Intravenous immunoglobulin (IVIG) for five days with improvement. Patient developed overlapping symptoms of fever, palindromic polyarthralgia with active urine sediment, and proteinuria lead to diagnosis of class V lupus nephritis. Prednisolone and Mycophenolate mofetil given and he received rituximab injection for extra renal manifestation of SLE.

Keywords: Guillain–Barré syndrome (GBS), systemic lupus erythematosus, Lupus nephritis, Intravenous immunoglobulin (IVIG) Acute Inflammatory Demyelinating Polyradiculoneuropathy.

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INTRODUCTION:

Systemic lupus erythematosus (SLE) is auto immune disease of multi-organ. Neuropsychiatric involvement of SLE is broad including central and peripheral nervous systems. Peripheral nervous system can be involved in the form of acute demyelinating polyradiculoneuropathy, GBS, mononeuropathy, polyneuropathy, myasthenia gravis, and others. SLE is more prevalent in women than men with ratio 2.0: 1 to 1.5: 1, which is has been attributed in part to an estrogen hormonal effect [1] and it varies by age and race [2]. SLE diagnosis can be a challenge due to its variable presentation. The incidence and prevalence of neurologic and psychiatric symptoms among patients with SLE vary greatly and studies show that approximately 1/3 to 1/5 of SLE patients report neurologic or neuropsychiatric symptomatology [3-4]. GBS as initial presentation of SLE is considered a rare and one of the least neuropsychiatric syndromes in SLE [5]. Kidney involvement is common in (SLE). Prevalence of membranous nephropathy is 10 to 20 percent in patients diagnosed with lupus nephritis [6]. SLE with silent clinical feature can be diagnosed with help of serologic finding for some antibodies (anti-double-stranded deoxyribonucleic acid [anti-dsDNA] and anti-Smith [anti-Sm]). Here we report the first case of Guillain barre variant as an initial presentation of SLE in the state of Qatar.

Case Report:

A 46-year-old male patient known case of bronchial asthma off medicines for five years. On 21 of July 2022 brought by EMS to Hazm Mebaireek General Hospital, Doha, Qatar emergency due to traumatic fall associated with head injury. Patient gave history of feeling very weak particularly in legs, when he tried to get on the bed he could not balance, and he fell backward. Patient spiked a 38C fever in emergency and his medical history revealed that he was discharged from hospital two weeks back as case of community acquired pneumonia. Patient has history of recurrent chest infection in last two months with negative septic work. Blood investigation on admission came unremarkable except low serum albumin. Based on his previous admission and the present chest x ray finding he was admitted as case of pneumonia and head trauma. CT scan head was unremarkable. Patient seen by pulmonologist, and he advised for bronchoscopy, but unfortunately patient desaturated before bronchoscopy and he was transferred to the intensive care unit where he was intubated. Chest x ray revealed bilateral lower lobe infiltrate and pleural effusion. Patient extubated after three days and step down to medical floor. On examination, he looked ill, febrile, and vitally stable. Patient developed new complain of inability to chew solid food and weakness all over body and neck and he was unable to sit without assistant help. He reported intermittent diplopia on the lateral gaze for the last three weeks associated with blurring of vision, not previously mentioned by him. Upper and lower limbs examination showed no muscle wasting or fasciculation. However, power was found to be grade three in both lower limbs and grade four in both upper limbs. Hypotonia was also noticed in his four limbs while hyporeflexia was confined to his ankles and knees along with bilateral down going plantar reflex. He has partial right third cranial nerve palsy with binocular diplopia more on the left gaze, no nystagmus and normal pupil, mild ptosis with overactive frontalis muscles. The possibility of Guillain Barre syndrome and third nerve palsy could be due to demyelinating mononeuritis. Patient was seen by neurologist and empirically treated with iv immunoglobulin and suggested to do lumber puncture, MRI head and nerve conduction study. MRI head revealed mild chronic microangiopathic changes. Electrodiagnostic study confirmed the diagnosis of acute motor and sensory axonal neuropathy variant of Guillain Barre syndrome. Patient remain febrile and he developed palindromic polyarthralgia. His blood investigation revealed pancytopenia. Serologic test for ANA, anti-RO, anti-LA, RNP were positive with negative anti

dsDNA. Rheumatologist consulted to rule out mixed connective tissue disease. According to the rheumatologist, the patient has anti-nuclear antibodies, RNP positive but does not fulfil the clinical criteria for the diagnosis of mixed connective tissue disease. He has borderline rheumatoid factor and significant anti-cyclic citrullinated peptide (anti-CCP) positive. Due to palindromic symptoms of polyarthralgia, he was started on prednisolone and azathioprine. Nephrologist were consulted because of active urine sediment and low serum albumin. Ultrasound guided renal biopsy in presence of 24-hour urine protein 1.9 gm/day with microscopic hematuria, low C3 complement, and low anti-phospholipase A2 receptor (PLAR2) antibody was performed. Renal biopsy revealed membranous lupus nephritis, ISN/RPS Class V with activity index:1/24, chronicity index: 1/12. We changed azathioprine to mycophenolate mofetil and given IV Rituximab 1gm every two weeks for two doses for extrarenal manifestations of lupus. Patient symptoms subsided and he was discharged with follow in nephrology and rheumatology clinic.

Discussion:

SLE is more common in female and affects multi organ in the body such as joints, kidney, heart, central and peripheral nervous system, blood, skin, and others.[7] The incidence of lupus nephritis is higher in black patients with SLE (34 to 51%), Hispanic patients (31 to 43%), and Asian patients (33 to 55%) than it is in White patients (14 to 23%).Our patient is a male and from Africa. Involvement of central nervous system in SLE is rare but vascular related manifestations (stroke, transient ischemic attack and venous thrombosis), cognitive (delirium and dementia), headache, psychiatric disorder (psychosis, mood disorder, anxiety) and seizer are more common[8] . In the aggregate, studies report that approximately one-third to one-half of SLE patients report neurologic or neuro-psychiatric symptoms [9]. In a study done by Hanley JG et al, 572 patients with SLE were reviewed for neuropsychiatric manifestations. Around 28% of them had at least a single neuropsychiatric event within five months of the diagnosis; however, only 19%-38% were attributed to SLE [10]. The prevalence of GBS is less than 2% in SLE but it can be fatal due to respiratory failure and autonomic disturbances [11]. Neuropsychiatric events can precede, occur concomitantly with, or follow the diagnosis of SLE. However, most events are accompanied by other SLE disease activity and occur close to the time of diagnosis [12]. GBS is an autoimmune demyelinating polyneuropathy affecting peripheral nerves and usually occurs after episodes of upper respiratory tract or gastrointestinal infections [13] . Campylobacter jejuni, mycoplasma pneumoniae, influenza virus, Epstein Barr virus, hepatitis, HIV and other organisms were found to be associated with GBS [14]. The prevalence of GBS increase with age [15]. Our patient presented with symptoms suggestive of GBS at the age of 46. He had experienced symmetrical ascending mu-scle weakness involving both upper and lower limbs. It was gradual and reached the maximum on the fifth day. There was no sensory loss but associated with partial right third cranial nerve palsy with binocular diplopia. Weakness can be somewhat asymmetric, and sensory loss can also be variable, rarely presenting with a pseudo-sensory level suggesting myelopathy. Facial nerve involvement occurs in up to 70% of cases, dysphagia in 40%, and rarely (5%) patients may develop ophthalmoplegia and ptosis [16]. Involving cranial nerves only in a form of Miller-Fisher (MFS) variant was also reported, but in our patient it does not fulfill miller fisher criteria [17]. Few similar cases have been reported among SLE patients [15-16]. Diagnosis of GBS is clinical and supported by many investigations including cerebrospinal fluid (CSF) analysis; with high protein level and normal cell count, but it can be negative in the first week in about half of patients. Polyneuropathy or polyradiculopathy on nerve conduction studies are diagnostic features [18,19]. Acute Inflammatory demyelinating

polyneuropathy (AIDP) is the most common subtype of GBS which is the same for our case[20]. This patient presented with a previous history of recurrent chest infection which can be organizing pneumonia in presence of positive Anti-Ro antibodies, rheumatoid factor, anti-cyclic citrullinated peptide (anti-ccp) and high ANA titer indicate mixed connective disease. The diagnosis of SLE is based on both clinical features and laboratory results. In this patient proteinuria, microscopic hematuria with low complement and high ANA titer determined lupus nephritis, proved by renal biopsy as membranous lupus nephritis, ISN/RPS Class V. Patient received IVIG with significant improvement. Prednisolone 40 mg tabs daily and mycophenolate mofetil 1000 mg tabs twice daily were started. This treatment was effective, and his condition had improved significantly with power grade 5 in all limbs during his follow up visit. Variant of Guillain–Barré syndrome and Miller-Fisher (MFS) as the initial presentation of SLE has been reported in only a few cases.[21]

Conclusion:

Guillain–Barré syndrome (GBS) is a rare manifestation of SLE. We report variant of Guillain–Barré syndrome as the initial presentation of SLE which is reported in literature with only few cases. Our patient developed overlapping symptoms of mixed connective disease and lupus nephritis. His condition had significantly improved with the use of IVIG, Mycophenolate mofetil and corticosteroids.

Conflict of interest: None

The manuscript has been seen and approved by all authors and it is not under consideration for publication elsewhere in any language.

Patient had given his consent for the case report to be published.

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