Peripheral Neuropathy as the Initial Presentation of Systemic Lupus Erythematosus

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ABSTRACT

SUMMARY

Systemic lupus erythematous (SLE) is a chronic and fatal disorder. Prior to now, SLE was said to be uncommon among blacks however recent findings are invalidating such opinions. Furthermore, peripheral neuropathy manifestation is rarely noted among Nigerian SLE patients, most especially as the first manifestation. We report this case with peripheral neuropathy manifestation as a major and initial composite symptom as learning point and also encourage high index of suspicion in this environment.

Keyword: SLE, Peripheral neuropathy

INTRODUCTION

Systemic Lupus Erythematosus(SLE) is a chronic, fatal and severe multi-systemic autoimmune connective tissue disease with multifactorial aetiology and a relapsing and remitting course .[1-5] SLE is reported to have a high prevalence in black Americans but under reported in Black Africans.[2] Before now it said to be rare in West Africa and before 1960 it was unknown in Sub Saharan Africa.[6] Although, the disorder is a heterogeneous entity, peripheral neuropathy is an uncommon presentation especially among Blacks which have cutaneous, manifestation as the commonest manifestation of the disease.[7]

The commonest manifestation reported in Nigeria was polyarthritis and polyathralgia, 2 the commonest neuropsychiatric systemic lupus erythematosus (NPSLE) manifestations in SLE are stroke, seizures, cognitive dysfunction and psychosis.[8,9]

Reporting this case is to encourage high index of suspicion in view of the fatal, variable and unpredictable course of disease.[1] It is necessary to remind ourselves that peripheral neuropathy(PN) can be a predominant and initial presentation of SLE.

CASE PRESENTATION

19 years old single tailor of Yoruba tribe presented at our centre on account of 2 months history of glove and stocking pattern of tingling sensation on both legs and difficulty in walking. The sensation was said to be of insidious onset and severe enough to impair her sleep. There was also low back pain, recurrent low grade fever, fatigue, weight loss and poor appetite. There was also joint swelling and pain, effort intolerance and easy fatigability.

There was no alopecia, skin rash, or oral ulcers. There was no photosensitivity or history suggestive of Raynaud phenomenon. There was no history change in urine volume and facial swelling.

There was no history of previous bowel surgery. She does not ingest alcohol or smoke cigarette.

She was found to be in obvious painful distress when first seen, pale, anicteric with multiple macular and patches on the face. There was no significant lymphadenopathy, no finger clubbing, half and half nail, mild pedal oedema.

She was conscious, oriented in time, person and place, cognition was intact, cerebellar sign was absent, there was weakness of small muscle of

hands, high stepping gait and feet and bilateral impairment of light touch in stocking pattern but no loss of vibration & joint position sensation. The assessment of power was 4 globally. There was inverted supinator jerk (left more than right) and absent ankle jerk while musculoskeletal examination revealed tender swollen ankles only. There was normal chest and abdominal finding.

INVESTIGATIONS

The erythrocyte Sedimentation rate (ESR) was 80mm/hr at admission. Anti-nuclear antibodies(ANA)(1:320) was positive while rheumatoid factor was 6.4IU/ml (normal <14.0 IU/ml) and Anti-cyclic citrullinated peptide (Anti-CCP) was negative. The x-ray of chest and pelvic were normal. The serum sodium, potassium, chloride, creatinine and bicarbonate were normal. The plasma blood sugar was normal. White cell count and platelet count were normal. Result of Human Immunodeficiency Virus screening was negative. The presenting packed cell volume(PCV) was 29% with thrombocytopenia. The mean corpuscular volume(MCV) was 82.1fL while mean corpuscular haemoglobin(MCH) was 25.6pg.

DIFFERENTIAL DIAGNOSIS

Acute inflammatory demyelinating polyradiculopathy, cervical spondylosis with radiculopathy and vasculitis.

TREATMENT

Patient was given various analgesic, pregabalin and commenced on standard SLE regimen.

OUTCOME AND FOLLOW-UP

Using the 2012 Systemic Lupus International Collaborating Clinics (SLICC) SLE criteria, making a diagnosis of SLE requires at least 4 criteria (at least 1 clinical and 1 laboratory criteria) of 17 total (11 clinical and 6 immunological/laboratory criteria) must be met.10 Furthermore patients with an ANA titre >1:40 and characteristic multi-organ system involvement can be diagnosed with SLE without additional testing.4 The patient met the criteria for diagnosing SLE with abnormal antinuclear titre, anaemia, leukopenia, arthritis and neurologic involvement (peripheral neuropathy). An assessment of distal sensori-motor polyneuropathy secondary to SLE was made. The problems identified in the patient included synovitis, fever, fatigue, anaemia, motor deficit and weight loss.

DISCUSSION

Incidence and prevalence of SLE varies globally.[3] The disease is common among Blacks and Hispanics.[2] The disorder is rarely reported among African blacks compared to African Americans.[11]

There is gender disparity with female predominance and ratio as much as 12:1 in childbearing years.[3] General M:F ratio is 4:1 with severity more in blacks than Caucasian whites.[7] This case was also in a female.

This case presented with a predominant and initial presentation of peripheral neuropathy. This is unusual in this environment based on available data although prevalence studies are few in Nigeria.[2,11] In a previous 6 year study of 1,250 rheumatologic cases presenting in a rheumatology speciality centre only 5.28% of such cases were SLE and peripheral neuropathy was not noted.[11] Common presentations that have been reported among blacks include fatigue, seizures, pleuritic chest pain, hair loss, weight loss, mouth and pharyngeal ulcers and cognitive impairment.[5] Although this is not the case in other settings where about 1.4 -5% prevalence of peripheral neuropathy in SLE have been found.[12,13] There are possibilities of other causes in setting of SLE such as chronic alcoholism, hereditary, uraemia, traumatic, diabetes mellitus, infectious and inflammatory causes were noted among their subjects.[13] In the patient in this study, other causes were rule out from history, physical examination and the investigations highlighted above.

Majority of PN in this disease tend to be polyneuropathy in nature.[13,14] While small fibre affection is the most common pattern seen in SLE.[13,15,16] Our case suggests a small fibre neuropathy rather than the large fibre neuropathy due to the normality of vibration and proprioceptive function. The predominant small fibre affectation in SLE is due to upregulation of matrix metalloproteinase-3 and matrix metalloproteinase-9 which causes vessel wall injury.[13,17]

Estimated mean age of presentation of SLE in Nigeria is 33 years and our patient presented at a lower age. Those with PN in a previous series showed those presenting with peripheral neuropathy also present about that age.[2,5,11,12]

There are almost always productions of antibodies and this contributes to the pathogenesis of SLE. The autoantibodies are produced by B cell and they

mediate tissue injuries by many mechanisms.[2] In addition, there is abnormal T cell signalling and gene expression which aid production of procytokines.[2] Antibodies such as antinuclear, anti-Ro, anti-LA, anti-phospholid, anti-double stranded DNA, anti-Sm and anti-nuclear ribonucleoprotein are present many years to varying degree before clinical manifestation of SLE.

Autoantibody play important role in the pathogenesis of neuropsychiatric manifestations in SLE through vascular or neuronal injury, intra-thecal production of inflammatory mediators and accelerated atherosclerosis.[8] The neuronal injury mechanism maybe the most likely in peripheral nerve disease found in SLE. In addition other study has demonstrated high prevalence of PN in SLE patients with positive anti-Sm antibody.[13]

Antinuclear antibody (ANA) is the commonest marker seen in SLE generally and also those with PN features and this was present in our patient.[12] ANA is particularly relevant in initial screening and reported sensitivity and specificity as high as 98.8% and 95.3% respectively .[18,19]

Currently the disease has no cure although it can be managed to improve better outcome and quality of life which can be low.[1] Glucocorticoid alone or with immunosuppressive drug appears to work well with response rate as much as 60-75%.[8] Cyclophosphomide, azathioprine or plasma exchange may be relevant in resistant cases.

This report is limited by inability to do conduction nerve study or electromyography for the patient although such usage have been noted to be aberrant.[13]

LEARNING POINTS

- Peripheral neuropathy is rare in SLE it can occur as first and predominant feature of SLE.
- A high index of suspicion is necessary for diagnosis SLE due to high mortality and morbidity especially among blacks and the fact it is not a common cause of peripheral neuropathy.[1,2,5,20,21]
- SLE has protean manifestations and peripheral neuropathy can occur as part of the symptom.

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