Neuropsychiatric lupus in a Nigerian teenager

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Abstract

This case report highlights the occurrence of neuropsychiatric lupus in a teenager with Systemic Lupus Erythematosus (SLE). The diagnosis of SLE was made based on the American College of Rheumatology (ACR) classification criteria. She presented with a history of fever, polyarthralgia, malar rash, pharyngitis, abdominal pain, discharging ears with reduced auditory function. She also had a history of fatigue, myalgia, weight loss, facial swelling, cough and hair loss. She had persistent headache, anxiety, confusion and generalized tonic clonic seizures. Essential findings on examination were those of distress on account of pain and difficulty with breathing, bilateral inguinal lymphadenopathy and a pulse rate of 118 beats/min.

Her laboratory results revealed a positive anti nuclear antibody (1:>5120) with a fine speckled pattern, positive anti double stranded DNA (>300 IU/ml), anaemia (Hb 7g/dl; PCV 21%), mean corpuscular volume of 66Fl, white blood cell count of 3.5 x 10^9, a normal platelet count, a negative rheumatoid factor, erythrocyte sedimentation rate of 123mm/HrWestergren. She had proteinuria (1+) with an essentially normal serum, electrolytes urea and creatinine.

Key words: Systemic lupus erythematosus, Neuropsychiatric, Anti nuclear antibody

Introduction

Systemic Lupus Erythematosus (SLE) is an autoimmune disorder with multisystem involvement resulting in significant morbidity and mortality. Childhood-onset SLE (cSLE) is a rare disease with an incidence of 0.3-0.9 per 100,000 children-years and a prevalence of 3.3-8.8 per 100,000 children with a higher frequency reported in Asians, African Americans, Hispanics and native Americans[1-3]. It represents 10-20% of all SLE cases, and is associated with higher disease severity than adult-onset SLE[1]. Hiraki *et al*[2] noted that non-Caucasian ethnicity is associated with increased childhood onset SLE disease prevalence and that non-Caucasian patients were significantly younger and more likely to have nephritis.

Most studies report a median age of onset of cSLE between 11-12 years. As in adult onset SLE, approximately 80% of patients with cSLE are females[4, 5]. As in adults the diagnosis is made based on the fulfillment of 4 or more of the American College of Rheumatology classification criteria for SLE[6].

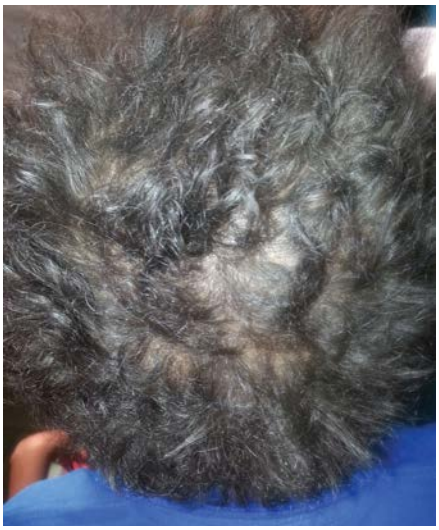
The clinical features in SLE are protean ranging from constitutional symptoms such as fever, anorexia, weight loss to neuropsychiatric involvement. Reports of childhood onset lupus from sub-Saharan Africa are scanty in spite of increasing reports of adult SLE[7]. In a study on juvenile SLE cases in Nigeria, 12 patients were seen over a 4 year period with all patients having haematological, constitutional symptoms and a positive anti nuclear antibody[7]. The only other report of juvenile SLE from Nigeria is a 2007 study of 11 children seen in a paediatric nephrology clinic[8]. Based on the rarity of this presentation we report a case of neuropsychiatric lupus occurring in a teenager.

Case report

The patient was a 13-year-old female student, who was apparently well until about a year prior to presentation when she developed high-grade fever, generalized joint pains and rash. Joint pains affected the wrist, knees, and the small joints of the hands with no associated swellings or stiffness. She observed a sudden appearance of non-pruritic rash on her face, sparing the nasal-labial area (malar-rash). The distribution of the skin lesions also involved the trunk and extremities. There was a history of pharyngitis, abdominal pains and bullous lesions appearing on the limbs. About two months prior to presentation, she developed bilateral, discharging ears which was initially painful. This has resulted in a decline in her auditory function.

Figure 1: Dyspigmented lesions on the face

Figure 2: Lupus hair with alopecia



She has had a history of fatigue, myalgia, weight loss, significant hair-loss, facial swelling and cough which is non productive and non pleuritic. She had been experiencing persistent headache which is dull in nature, acute confusion and anxiety. She had some episodes of generalized tonic-clonic seizures while on admission, sudden weakness of the upper and lower limbs a day prior to presentation and attained menarche on admission.

On general physical examination, she was chronically ill looking, in distress due to pain and difficulty with breathing and wide-spread dyspigmented lesions on the face and trunk. She was warm to touch (37.1^{o}C), pale, anicteric, acyanosed, facial puffiness with bilateral inguinal lymphadenopathy. She was mildly dehydrated with bilateral pitting pedal edema.

Her pulse rate was 118 beats per minute; irregularly irregular with apex beat at the 4th left intercostal space, mid-clavicular line. Heart sounds S1, S2, S3 were heard with respiratory rate of 32 cycles per minute. Her breath sounds were broncho-vessicular with SPO_2 of 88%. There was generalized abdominal tenderness. She was conscious and oriented in person but not in place and time.

Her laboratory results revealed a positive anti nuclear antibody (1:>5120) with a fine speckled pattern, positive anti double stranded DNA (>300 IU/ml), cardiolipin Ig M and IgG (1.7 NPL/ml, 3.0GPL/ml respectively), anaemia (Hb 7g/dl, PCV 21%), mean corpuscular volume of 66Fl, white blood cell count of 3.5 x 10^9, normal platelet count (376×10^9/L), a negative rheumatoid factor (4.2 IU/ml), erythrocyte sedimentation rate of 123mm/HrWestergren, CRP <2.0mg/l. She had proteinuria (1+) with an essentially normal serum electrolytes, urea and creatinine. She was placed on pulse methyl prednisolone for 3 days and then oral prednisolone, hydroxychloroquine, azathioprine, phenytoin infusion and oral phenytoin thereafter. Other medications administered were; ceftriaxone, co-amoxiclav, omeprazole, lisinopril, and otomed ear drops. She was transfused with two units of blood. She was discharged on the nineteenth day on admission having improved remarkably.

Discussion

The reported case has SLE having had more than four (polyarthralgia, hair loss, anaemia, leucopenia, seizures, malar rash, a positive anti nuclear antibody and double stranded DNA) of the American College of Rheumatology classification criteria[6]. The exact aetiology is unknown but the interactions between immune complexes, autoantibodies, genetic, drugs and environmental factors do play a significant role in causing inflammation and eventually damage to the organs and systems[9].

The age of the patient (13 years) is in keeping with most studies which have reported a median age of onset of cSLE between 11-12 years[10]. The female preponderance seen in other reports is highlighted in this case[4,5]. Fevers, lymphadenopathy, rash, renal dysfunction, neurological and haematological disorders and polyarthralgias have been described[11].

She had constitutional symptoms which are frequently recounted in patients with childhood onset lupus[8]. She experienced some neuropsychiatric manifestations (seizures, headache, confusion and anxiety). The central and peripheral nervous systems can be involved with 19 distinct neuropsychiatric lupus (NPSLE) syndromes described[12]. Up to 65% of childhood SLE patients develop NPSLE at any time during the disease course, and up to 85% of these patients will develop NPSLE within the first 2 years from diagnosis[13,14]. Neuropsychiatric involvement with SLE is at least as common in children as it is in adults, with the former experiencing symptoms especially within a year after diagnosis with SLE (70% vs. 28%)[16]. Neuropsychiatric manifestations have been reported in 29-44% of paediatric patients with SLE[17]. Anaemia is not uncommon in childhood SLE as seen in this case. Cytopenias are common in cSLE, with more than 50% of patients presenting a decrease in at least one cell line[4,16].

Anti Nuclear Antibody (ANA) was positive with a significantly high titre. The commonest autoantibody is the ANA which is present in more than 95% of cSLE[10]. Anti-ds DNA antibodies are highly specific for SLE, and are present in about 61-93% children with active disease, especially active nephritis[18]. This serological marker was positive in the reported case with a significantly elevated titre as well as the presence of proteinuria. The use of corticosteroids and hydroxychloroquine have shown excellent results in control of the disease[18] as in the reported case.

Conclusion

Systemic lupus erythematosus has protean manifestations. Neuropsychiatric lupus in children has been rarely reported in Nigeria. A high index of suspicion is imperative in making a diagnosis and the institution of effective aggressive therapy is rewarding.

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