Case Report

Systemic Lupus Erythematosus Masquerading as Disseminated Tuberculosis: Case Report in a Nigerian Adolescent

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Abstract

Systemic lupus erythematosus (SLE) is a connective tissue disorder whose manifestations may mimic other common chronic diseases in children. In developing countries, its diagnosis is often delayed or missed leading to delay in instituting appropriate treatment and invariably high mortality. We report the case of A. B who was a 13-year-old girl referred from a peripheral hospital with chronic cough, weight loss, and dyspnea. She had signs of heart failure and developed depression as well as oliguria. The patient also had pleural effusion, but aspirate result was negative for *Mycobacterium tuberculosis* and cytology. She commenced antituberculous drugs and dexamethasone with other supportive care but died after 19 days on admission. Serum assay was positive for antinuclear antibody. SLE is a potential masquerader of chronic diseases such as tuberculosis. Delay in diagnosis and treatment is associated with poor outcome; hence, there is a need for high index of suspicion for early diagnosis with prompt initiation of appropriate treatment.

Keywords: Adolescent, systemic lupus erythematosus, tuberculosis

Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem inflammation of blood vessels and connective tissue.[1] The specific cause of SLE is unknown, but multiple factors are associated with the development of the disease, namely genetic, ethnic, hormonal, and environmental factors.[2]

More than 90% of cases of SLE occur in female, usually starting at childbearing age.[3] The male: female ratio before puberty is 1:3, but after puberty, it increases to 1:9.[1] The manifestations of SLE are mediated by circulating immune complexes in various tissues or the direct effects of antibodies on cell components.[2] The new classification by the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) requires an antinuclear antibody (ANA) titer of at least 1:80 or an equivalent positive test in addition to at least one clinical criterion and 10 or more points as diagnostic criteria for SLE.[4]

The course of the disease is milder with higher survival rate in persons with isolated skin and musculoskeletal involvement than in those with renal disease and central nervous system (CNS) disease.[5,6] SLE carries a highly variable prognosis in different patients; the natural history of the disease ranges from relatively benign disease to rapidly progressive and even fatal disease.[7] Delay in diagnosis as well as presence of renal or CNS disease is associated with high mortality.

Case Report

A. B was a 13-year-old girl who was referred to our facility from a private teaching hospital with 4-month history of intermittent cough and fever. She also had progressive weight loss with dyspnea, leg swelling, and chest pain. The results of the investigations from the referring center are shown in Table 1. She had pericardiocentesis that yielded a culture-negative effluent; she was being managed for pulmonary tuberculosis with effusions and was commenced on antituberculous regimen with cefuroxime before she was referred to our facility.

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At presentation, she was febrile and had clinical features of pleural effusion with congestive cardiac failure. She was assessed to have disseminated tuberculosis in heart failure, to rule out connective tissue disorder. Investigations showed elevated erythrocyte sedimentation rate, azotemia, leukocytosis with toxic granulation, proteinuria with glycosuria, as well as evidence of right-sided pleural with pericardial effusion [Table 2 and Figure 1]. She recommenced the antituberculous drugs and also ceftriaxone, short course of frusemide, fresh frozen plasma, and dexamethasone; she had closed tube thoracostomy drainage [Figure 2] with subsequent intrapleural injection of streptokinase on the 13th day into admission. The patient developed irrational talk, headache, and features of depression on admission; she later became unconscious and oliguric shortly before demise. She was commenced on supportive management for acute kidney injury but died 6 h after, although the total duration of admission was 19 days. Her autopsy showed widespread fibrotic lesions in the lung, but no giant cell or granuloma was seen. She had positive ANA test, but the result was retrieved after demise.

Discussion

Juvenile SLE is scarcely reported in sub-Saharan Africa despite the increasing reports of the adult form. Adelowo *et al*. posited that the juvenile type accounts for about one in five cases of SLE in a study from South West Nigeria.[8] It accounts for 24.6% of pediatric rheumatic diseases in a study by Olaosebikan *et al*. in Lagos State, Nigeria.[9] Delay in diagnosis from symptom onset to diagnosis ranges from 1 month to 3.3 years.[10] This is due to its similar manifestation with other chronic tropical conditions such as tuberculosis.

Pediatric SLE usually presents in postpubescent females.[1] The mean ages at lupus diagnosis are 11–12 years.[1,11] There is a 24%–56% concordance rate in monozygotic twins, compared with a 2%–5% risk in dizygotic twins, suggesting genetic tendency.[12] Chronic infections may induce anti-DNA antibodies or even lupus-like symptoms, and acute lupus flares often follow bacterial infections. Estrogen use in postmenopausal women appears to increase the risk of developing SLE. Photosensitivity is clearly a precipitant of skin disease since ultraviolet light stimulates autoantibody production.[13]

Common manifestations of the disease include fatigue, fever, weight loss, lymphadenopathy, and hepatosplenomegaly.[1,7] Fever may reflect active SLE or infection; patients with SLE are considered immunocompromised. Lupus patients may be functionally asplenic and may be at risk for encapsulated bacterial infections.[7]

Renal disease manifests in about 50% of patients with SLE at presentation and is the greatest contributor to morbidity and mortality in pediatric SLE population.[1,7] Neuropsychiatric disease occurs in up to two-thirds of patients; it is the second leading cause of morbidity and mortality in affected population.[1] Headache is the most common CNS finding in SLE, occurring in 72% of children.[14] Pericarditis is the most common cardiac manifestation, while pulmonary involvement may manifest as pleuritis, pleural effusion, and pulmonary hypertension[1,7]

ANA is found in 99% of patients with SLE, although it may be positive in mixed connective tissue disease and dermatomyositis.[1] The anti-dsDNA, on the other hand, is very specific for SLE but less sensitive compared to ANA. It may be found in >75% of patients with pediatric SLE.[1]

Table 1: Tests and results from referring center

Tests	Results
ESR (mm/h, Westergreen)	70 mm/h
Sputum microscopy	*Streptococcus pneumonia*
Sputum GeneXpert	Negative
Echocardiography	Moderate pericardial effusion
Abdominal ultrasound	Moderate ascites

ESR: Erythrocyte sedimentation rate

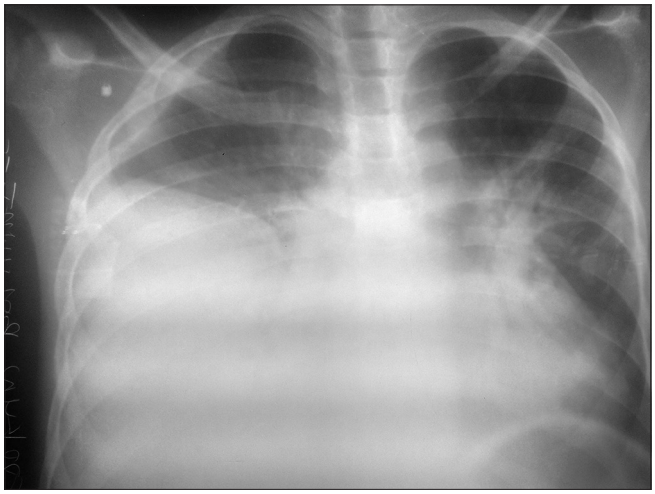

Figure 1: Chest X-ray showing right-sided homogenous opacity (pleural effusion)

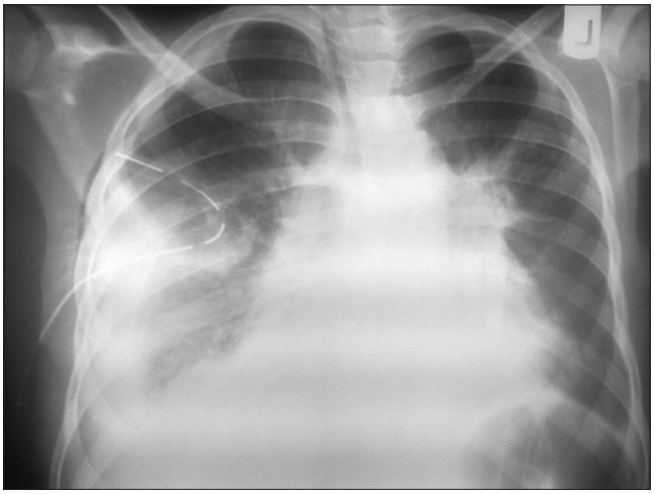

Figure 2: Chest X-ray after insertion of chest tube at site of pleural effusion with cardiomegaly

Table 2: Tests and results from our facility

Tests	Results
HIV	Nonreactive
Urinalysis	Proteinuria (1+), Glycosuria (1+)
Pleural fluid for microscopy	No growth
Pleural fluid for GeneXpert	No MTB seen
Pleural fluid for cytology	Inflammatory cells seen, no malignant cell
ESR (mm/h, Westergreen)	103 (↑)
Full blood count	
Packed cell volume	35% (N)
White blood cell	12,800 c/mm^3 (↑)
	Neutrophil-82%, Lymphocyte-18%
	Toxic granulation
Serum chemistry	
Sodium (mmol/L)	133 (N)
Potassium (mmol/L)	3.1 (N)
Bicarbonate (mmol/L)	20.0 (N)
Urea (mmol/L)	18.0 (↑) (Day 4: 4.6 [N])
Creatinine (umol/L)	59.0 (N)
Total protein (g/L)	68 (N)
Albumin (g/L)	34 (↓)
Globulin (g/L)	34 (N)
Aspartate transferase (U/L)	11 (N)
Alanine transferase (U/L)	5 (N)
Blood culture	No growth
Chest X-ray	Homogenous opacity suggestive of pleural effusion in the right hemithorax
Chest ultrasound	Right-sided pleural effusion with loculation
Echocardiography	Pericardial effusion, no structural defect

Table 3: 2019 European League Against Rheumatism/American College of Rheumatology Clinical and immunological domains and criteria for systemic lupus erythematosus[4]

Domain	Criteria	Points
Constitutional	Fever	2
Hematologic	Leukopenia	3
	Thrombocytopenia	4
	Autoimmune hemolysis	4
Neuropsychiatric	Delirium	2
	Psychosis	3
	Seizure	5
Mucocutaneous	Nonscarring alopecia	2
	Oral ulcers	2
	Subacute cutaneous or discoid lupus	4
	Acute cutaneous lupus	6
Serosal	Pleural or pericardial effusion	5
	Acute pericarditis	6
Musculoskeletal	Joint involvement	6
Renal	Proteinuria >0.5 g/24 h	4
	Renal biopsy Class II or V lupus nephritis	8
	Renal biopsy Class III or IV lupus nephritis	10
Antiphospholipid antibodies	Anticardiolipin antibodies or	2
	Anti-β2GP1 antibodies or	
	Lupus anticoagulant	
Complement proteins	Low C3 or low C4	3
	Low C3 and low C4	4
SLE-specific antibodies	Anti-dsDNA antibody or	6
	Anti-Smith antibody	

SLE: Systemic lupus erythematosus

The new diagnostic EULAR/ACR criteria have sensitivity of 96.1% and specificity of 93.4%, compared with 82.8% sensitivity and 93.4% specificity of the 1997 ACR criteria[4] [Table 3]. Positive ANA result is cardinal to making the diagnosis of SLE.[4] In line with the new guideline, this index case met more than ten-point criteria to diagnose SLE.

Treatment modality is medical and includes the use of hydroxychloroquine or nonsteroidal anti-inflammatory drugs, whereas severe disease is treated with steroids (methylprednisolone and prednisone) or steroid-sparing antirheumatic drugs (cyclophosphamide, methotrexate, azathioprine, belimumab, and rituximab).[7]

The average 10-year survival rate exceeds 90%;[6] The presence of renal or CNS manifestations is associated with poor prognosis. The high rate of misdiagnosis or delayed diagnosis is probably responsible for high mortality associated with lupus in resource-poor settings.[8,11]

Conclusion

SLE is rarely reported in Nigeria owing to frequent misdiagnosis. This is because of its similar manifestations with other chronic diseases that are endemic in Nigeria and hence the need for high index of suspicion for early diagnosis and prompt treatment so as to keep the mortality low.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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