

Isolated Digital Necrosis in a Young Phototype VI Senegalese, Revealing Systemic Lupus Erythematosus (SLE)

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Abstract

Introduction: Lupus disease has a wide range of clinical manifestations. Finger necrosis is often described during the disease. We report an exceptional case of isolated digital necrosis as a presenting symptom of SLE in a Senegalese of type VI prototype.

Observation: A 33-year-old man, smoker 1 pack/year, had consulted for a 3-year evolving digital necrosis and repeated amputations. On examination, the general condition was altered and the right radial pulse was not palpable. The diagnosis of lupus vasculitis was established in presence of the anti-Sm positive antibodies and ruling out of other vasculitis causes. The evolution was good under corticosteroid, antimalarial and anticoagulant therapy. No recurrence was observed after a three-year following.

Discussion: Digital necrosis is one of the late manifestations of SLE. It occurs rarely as primary or the only manifestation of SLE. Therefore, in our patient, before establishing SLE diagnosis we have ruled out as a priority, systemic scleroderma and Berger disease. The occurrence of this a typical SLE manifestation in an African man makes our observation more unusual.

Keywords: Necrosis; Vasculitis; Systemic lupus

Introduction

Finger necrosis has wide variety of causes. The main causes are systemic scleroderma and arterial disease [1]. It is rarely described in SLE. In fact, digital necrosis affects less than 1% of patients with SLE [2], and they occur mainly in chronic and protracted SLE [3]. In such cases, they are not always bound to an antiphospholipid antibody syndrome [4]. In Africa, lupus vasculitis with isolated finger necrosis is very rarely reported. We report an exceptional case of isolated digital necrosis as a presenting symptom of SLE in a Senegalese of type VI prototype.

Observation

A 33-year-old patient, a professional cook, smoker, 1 pack-year, weaned 3 years ago, with consulted for a severe Raynaud's syndrome, digital necrosis in the last 3 years and two episodes of digital amputations in an orthopaedic department. Past medical history revealed treated and cured declared pulmonary tuberculosis. There was no evidence for addiction or blood transfusion. In addition, there was a notion of familial polyarthritis. Physical examination showed necrosis of the fourth and fifth finger of the right hand, amputation of the second and third homo lateral fingers (Figure 1) and stellar scars on the pulp of the fingers of the left hand (Figure 2).

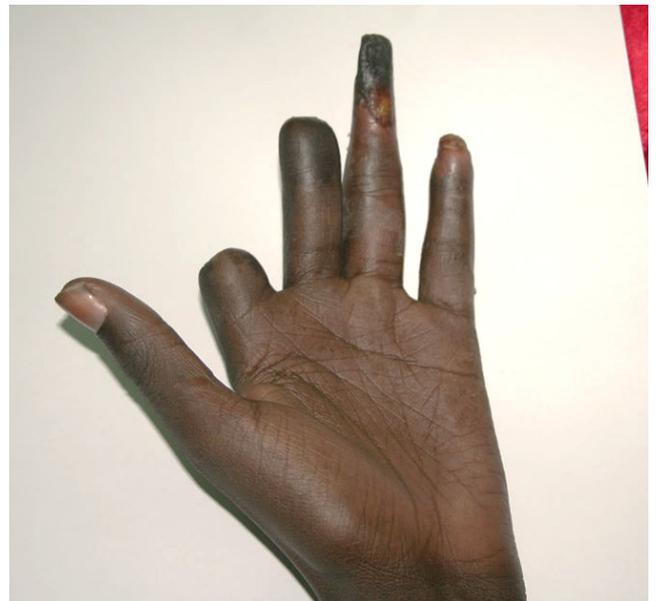


Figure 1: Finger necrosis and a previous amputation.



Figure 2: Finger scarring in a SLE patient with Raynaud phenomenon.

The left radial and ulnar pulses were weakened and cardiac hyperretism. The rest of the physical examination was normal. The blood count was normal, the C-Reactive Protein was 24 mg/l, and the first hour ESR 50 mm, BUN and serum creatinine and 24-H proteinuria as well as U/A were normal. HIV serology and AgHbs were negative. The anti-U1RNP, anti SmRNP and anti Sm antibodies were all greater than 8 (according to laboratory standards, these values are positive if they are greater than 1.2). The p-ANCAs were positive, with an anti-MPO and anti-PR3 negative. Anti-scl-70, anti-centromere, anti-Jo-1, anti-SSA/Ro, SSB/lupus anticoagulant and anti-phospholipids antibodies (anticardiolipin, lupus anticoagulant and anti- β 2-glycoprotein I) were negative. The radiograph of the hands showed a bone demineralization (Figure 3).



Figure 3: Demineralization of the hand bones of the in a SLE patient with digital necrosis.

The electrocardiogram showed sinus tachycardia, an incomplete right branch block and left ventricular hypertrophy. Doppler echocardiography and upper echocardiography were normal. Thoracic CT showed superior left lobar parenchymal lesions with no other abnormalities. Based on these clinical and immunological signs, the diagnosis of lupus vasculitis was retained. Disarticulation of the necrotic phalanges was performed. The patient treated with prednisone, at 1 mg/kg per 24 h, hydroxychloroquine at 400 mg per 24 h, combined with an oral anticoagulant (Acenocoumarol). Dildiazem was stored as a symptomatic treatment of Raynaud's phenomenon. The evolution was marked by a complete regression of necrosis (Figure 4), we noticed no recurrence and after a 3-years follow-up.

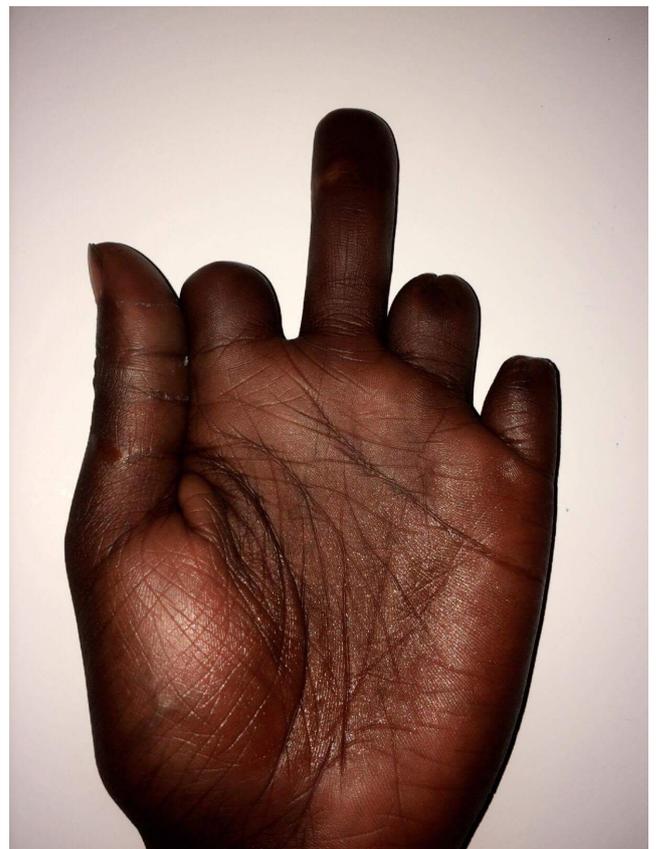


Figure 4: Amputation stumps six months after treatment in a SLE patient with digital necrosis.

Discussion

Our patient had lupus vasculitis. In addition of SLE, the other possible diagnoses were systemic scleroderma, Buerger's disease and cryoglobulinemia. The diagnosis of systemic scleroderma could not be established because of the lack of the required criteria (ACR/EULAR criteria 2013) [5]. Buerger's disease is an elimination diagnosis. For cryoglobulinemia, the HbsAg was negative and our patient did not have any risk factors for hepatitis C exposure. More over our patient showed SLE specific anti-Sm antibodies.

Indeed, it is obviously demonstrated that the presence of anti-Sm antibodies is very specific for systemic lupus [6]. In addition, the

presence of both anti-Sm and anti-U1RNP antibodies is more common in SLE patients with darker phototype, as demonstrated in African-Americans [7]. In dark phototype patients with SLE as in our patient, when both anti-Sm and anti-U1RNP antibodies are associated, the visceral and hematological manifestations are less frequent [8].

Severe Raynaud phenomenon and finger necrosis as the only manifestation of SLE in a young African patient with dark phototype is very rare, what makes the distinctiveness of our observation exceptional. Raynaud phenomenon is described only in 10 to 45% of patient with SLE [9] and it seems to be related to age and the presence of other skin manifestation [9,10]. Our patient had no other skin manifestation. As Immunologic point of view, studies have shown that the Raynaud phenomenon is linked to the presence of antibodies against U1RNP [11,12]. During SLE what is the case in our patient. Finger necrosis is rarely reported during SLE. They usually occur several years after the appearance of other manifestations [13]. Liu et al. [13] showed that long lasted course, presence of a Raynaud phenomenon and a positive CRP are the major predictors of digital necrosis in a patient with SLE [13].

Our patient had only the two latest risk factors. However, his Job as a cook could worsen for Raynaud phenomenon due to a frequent cold exposure. The causes of Finger necrosis in SLE are mainly vasculitis or vascular thrombosis, especially when SLE is associated with antiphospholipid syndrome (APS), hypercoagulability, hyperhomocysteinemia or early onset atherosclerosis [13,14]. Indeed, digital necrosis is more frequent in APS associated SLE, with a frequency between 3.3 and 7.5% [15,16]. In our patient, the arterial sonography of the upper limbs was normal, that is in favour of a vasculitis. This vasculitis with isolated digital necrosis, during the SLE is exceptional in the African dark phototype, especially in men. To our knowledge, only two female cases were reported by a Nigerian team in 2012 [17] and one of them showed positive β 2 glycoproteins.

The other uniqueness of our observation is the p-ANCA positivity. These antibodies are linked to lupus nephropathy, during SLE, and not to digital necrosis [18]. Laboratory finding of renal disease in our patient were negative.

As treatment, the authors suggest a daily dose of ≥ 1 mg prednisone associated with cyclophosphamide and an anticoagulant or platelet aggregation inhibitors [13]. They had demonstrated that the introduction of this corticosteroid therapy, within the first 3 weeks, reduces the risk of amputation. In our patient, the lack of knowledge about this form of SLE was the origin of the diagnostic delay, what conducted to two episodes of amputation.

Conclusion

Digital necrosis has a wide range of causes and requires an exhaustive clinical investigation. Given this clinical picture, a close collaboration between surgeon and dermatologist is necessary for a proper management these patients to avoid diagnostic mistakes and repeated amputations.

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