

Lupus Profundus as an Initial Manifestation of a Late Indolent SLE

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Abstract

After 11 years from the onset of developing lupus profundus the patient described here began to intermittently express hematological and serological features of SLE yet failed to reach the minimally required four criteria for the classification of the systemic disease. Lupus profundus in this case was the initial manifestation of a late but indolent SLE. Regardless whether the patient eventually will develop the complete form of lupus or not, the case stresses on the importance of the constant and regular follow up of patients with isolated skin lesions as they may turn into a wider form of lupus even after lengthy period of time.

Keywords: Lupus profundus; Systemic lupus erythematosus (SLE); DNA antibodies; Panniculitis

Introduction

Lupus profundus (LP) or panniculitis is a variant of cutaneous lupus erythematosus. It affects the subcutaneous fat and characterized by one or more nodules and or/plaques or ulcers, and frequently heals with atrophy or scars. Several sites of the body can be affected including the scalp [1]. It occurs in 2%-3% of patients with SLE. Conversely, between 10%-15% of patients with lupus profundus will have or eventually develop systemic disease [2,3]. Lupus profundus was seen in 6/228 (2.6%) and 4/86 (4.6%) in two series of discoid lupus (DLE) patients [3]. Despite all the above lupus profundus is not considered as a criterion for the classification of SLE [4]. The lesion was also described in a few cases of dermatomyositis and morphea/scleroderma [5,6]. In this report we describe a case that after 11 years from the development of lupus profundus the patient evolved only into an indolent form of lupus.

Case Report

In 2000, a 25 year old Arab female initially developed nodular lesion in the inner part of the left thigh that became large ulcerative and non-healing overtime (Figure 1). There was no other systemic manifestation or rash elsewhere. Her past medical history was unremarkable. She was treated with systemic antibiotics and local care of ulcer. With the possibility of pyoderma gangrenosum, morphea profundus and lupus profundus in mind, a biopsy was taken from the edge of the lesion which showed atrophic epidermis and focal basal vacuolation. The dermis was oedematous with erythrocytic infiltration leading to fibrosis. Necrotic fat lobules containing hyalinized interstitial fibrin deposits and lymphocytic infiltration were seen in the subcutaneous tissue. These were consistent with clinical diagnosis of lupus profundus. Routine investigations including hematology, biochemistry, urine analysis, VDRL, RF, ANA, ds DNA and ENAs were within normal limits and remained as such during the follow up. As the lesion became scarring and disfiguring (Figure 2), she underwent plastic surgery in 2002 (Figure 3). In the subsequent 9 years she remained asymptomatic and the lupus profundus lesion did not worsen or flare up. She had two successful pregnancies and deliveries since.



Figure 1: Early ulcerative lesion of the lupus profundus.

In early 2011, her CBC showed Hb of 12.6 g/dl, mild leukopenia (WCC of 3400 cc³; borderline reticulocytosis of 2.6% (N<2%) and positive direct Coomb's test. The ANA index (ELISA) tested positive of 2 (N<1.2), but with negative ds DNA Abs (<20 IU), ENAs, VDRL and RF. She was given short course of corticosteroids with good response and normalization of the blood picture.

In late 2011, the ds DNA Abs raised to 35.2 IU, CRP of 10.3 mg/dl (N<5) and ESR of 49 mm/h. By 2013 the ds DNA Abs reached 75.2 IU. No other symptoms were reported and the skin lesion remained inactive. She became mother for the third time. In 2014, the patient was noticed to have Hb of 9.9 g/dl, mild leukopenia of 3050 mm³, normal platelets count, ESR of 88 mm/h, ANA index: 1.5 (positive) and ds DNA Abs of 30 IU. Both ACL Abs and ENAs were negative yet the C3 complement was marginally low, 78 mg/l (N 83-193 mg/l). Serum urea, creatinine and urine analysis were within the normal range. The patient was commenced on hydroxychloroquine and oral corticosteroids and lately been maintained on prednisolone 5 mg and hydroxychloroquine 200 mg daily. Her latest results (1 month ago) showed Hb: 12.8 g/dl, WCC: 4490 mm³ with normal differential count, platelets: 235,000 and ESR of 41 mm/h. The direct Coomb's test was still testing positive but the reticulocytic count remained below 2%, CRP: negative, ANA screen: positive, ds DNA: 33 (N<5), ENAs:

negative, urine routine: normal outcome, C3: 79 mg/l (N 90-180 mg/l) and C4: 14 mg/l (N 10-40). Her plain X-ray of the thigh showed dystrophic calcinosis (Figure 4). However, she remained asymptomatic otherwise.



Figure 2: Post ulceration scarring and disfigurement.



Figure 3: Post plastic surgery healing.



Figure 4: Subcutaneous dystrophic calcinosis.

Discussion

Reviewing the pertinent literature of LP suggests a degree of diversity in the clinical presentation, association and prognosis of the disease. It may arise in patients with discoid or systemic lupus or as isolated phenomenon. Consistent with SLE, LP is more common in females than males and most often presents clinically with single or multiple nodules or plaques involving the arms, shoulders, buttocks or face. Ulceration, depressing scars, lipoatrophy and disfigurement may also develop [1-3,5,7-10]. Some cases of LP have been associated with an antecedent trauma to the site and exacerbation of LP has been reported following excision of lesions [11]. Although not all cases progress into the systemic form of SLE, LP patients tend to have a mild clinical form of the disease [2,3,10-12]. The lack of renal complications in some series was in keeping with the overall benign tendency of LP [10-12]. The time lapse for the evolution is variable though and may take some years. Lupus profundus is a chronic relapsing condition. The treatment options include intralesional and systemic corticosteroids, antimalarial drugs, mycophenolate mofetil, dapsone, cyclosporine and thalidomide. It appears that the early treatment of the nodular or plaques has good prognosis [8,11]. Successful treatment with Rituximab has also been reported recently [13].

The main characteristics of the patient reported here that she remained having inactive LP for 11 years then suddenly began to intermittently, depict features of systemic disease without reactivation of the initial skin lesion. However, the patient never fulfilled the criteria of the SLE at any point in time. The disease remained within the indolent form of lupus. However, one can't discard the role of treatment with hydroxychloroquine and corticosteroids here as it could have possibly ameliorated the evolution of the condition following the control of the mild hemolytic anemia and leukopenia. Nonetheless, the constant follow up of the case has proven very effective in monitoring the disease progress. The exceptionally lengthy time lapse between the major events in this case implies that even with an apparently benign form of chronic cutaneous lupus a vigilant and regular follow up is warranted. Dystrophic calcinosis which is known to occur as sequelae of tissue damage due to inflammatory process is not unusual in patients with LP [2].

Finally, given the fluctuations of the lengthy course of the disease and the failure to develop the full blown picture of systemic disease suggests that lupus in some patients may behave as unusually indolent disease. The case may also demonstrate the incapacity of modern approach in medicine to define some systemic diseases unless a certain number of criteria become available to the clinicians.

Conflict of Interest

The author(s) declare(s) that there is no conflict of interest regarding publication of this article.

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