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Unusual presentation of lupus erythematous profundus in a young female treated by intralesional trimacinolone and oral methotrexate: A case report

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ABSTRACT

Systemic lupus erythematosus is a chronic autoimmune condition that can attack almost any organ. In fact, its clinical manifestations and serological abnormalities are the most diverse one among other autoimmune diseases. An uncommon distinctive entity of this disease is lupus erythematosus profundus. Herein, we report a case of 14 years old female complained of a progressive lesion over her right arm that was ulcerated later. Lupus erythematosus profundus diagnosis was confirmed through multiple histopathological examination. Patient was started on hydroxychloroquine 200 mg twice daily, but she could not tolerate it. Accordingly, she was switched to methotrexate in addition to trimacinolone injection, as she showed more tolerance and improvement. The patient was seen for almost two years.

Keywords: Systemic lupus erythematosus, lupus erythematosus profundus, trimacinolone injection, oral Methotrexate.

1. INTRODUCTION

Autoimmune diseases are complex and unpredictable as they can involve several organs and systems (Park et al., 2012). Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that can affect almost any organ (Wallace & Hahn, 2012). In fact, its clinical manifestations and serological abnormalities are the most diverse one among other autoimmune diseases (Font et al., 2004). A mixture of constitutional complaints with, musculoskeletal, cutaneous, mild hematologic, and serologic involvement is the most common pattern. Cutaneous lupus erythematosus (CLE) is subclassified into three subsets includes acute cutaneous lupus erythematosus, subacute cutaneous lupus erythematosus, and chronic cutaneous lupus erythematosus (CCLE). CCLE encompasses discoid lupus

erythematosus, lupus erythematosus tumidus, lupus profundus (also known as lupus panniculitis), chilblain lupus erythematosus, and lichenoid cutaneous lupus erythematosus-lichen planus overlap syndrome (Biazar et al., 2013). CLE can occur independently from SLE, or as a part of the clinical picture of SLE. An uncommon distinctive entity of CLE is lupus erythematosus profundus (LEP).

LEP is characterized by a painful indurated plaques or subcutaneous nodules that occur on the scalp, face, upper arms, flank, lower back, chest, buttocks, or upper thighs (Crowson & Magro, 2001). Uncommonly, the site of the nodule become ulcerated or calcified and can result in depressed areas of lipoatrophy upon resolution (Martens et al., 1999). The most common age group at the time of presentation is between the third and the sixth decades, with a strong female predominance accounts for 90% of the cases (Park et al., 2010). The modality of choice in diagnosing LEP is histopathologic examination. Here, we report a case of 14 years old female complained of a progressive lesion over her right arm that was ulcerated later. LEP diagnosis was confirmed through histopathological examination. The patient was seen for almost two years.

2. CASE PRESENTATION

A 12 years old Saudi female, who is medically free, came to our dermatology clinic complaining oftwo months history of single lesion over the right arm, started spontaneously and became progressing in size and started to ulcerate. This lesion was not associated with pain and not preceded by any trauma or insect bite. There were no systemic symptoms.Family and drug histories were unremarkable. On examination, there was a solitary, indurated, deep, firm, and mobile nodule over the right arm with overlying erythema, loss of hair, and minimal scaling (Figure 1). The differential diagnoses at first were lipoma, dermoid cyst, and dermatofibroma. Two biopsies were taken, one was incisional biopsy for the lesion, and a second biopsy was taken from adjacent normal skin. In Figure 2A, the incisional biopsy of ulcerated skin lesions shows infiltration of the deep dermis and subcutaneous fat lobules by intense chronic inflammation associated with fat necrosis, and mild fibrosis (arrow). The inflammatory component consists almost exclusively of small-size lymphocytes, plasma cells, and occasional macrophages, small-size blood vessels show thickening and hyalinization, hematoxylin and eosin (H&E) x400. In figure 2B for non-ulcerated skin, the epidermis shows moderate superficial and deep perivascular chronic inflammation comprises lymphocytes and plasma cells (asterisk). The inflammatory infiltrate shows periadnexal/perifollicular involvement. No perceivable vacuolar or inflammatory interface (dermal-epidermal junction) changes are present. The epidermis shows mild acanthosis and basket-weave orthokeratosis, H&E, x200. In addition, Figure 2C the dermis demonstrates deposition of granular blue mucinous material between collagen bundles identified by Alcian blue stain, pH 2.5, x400.

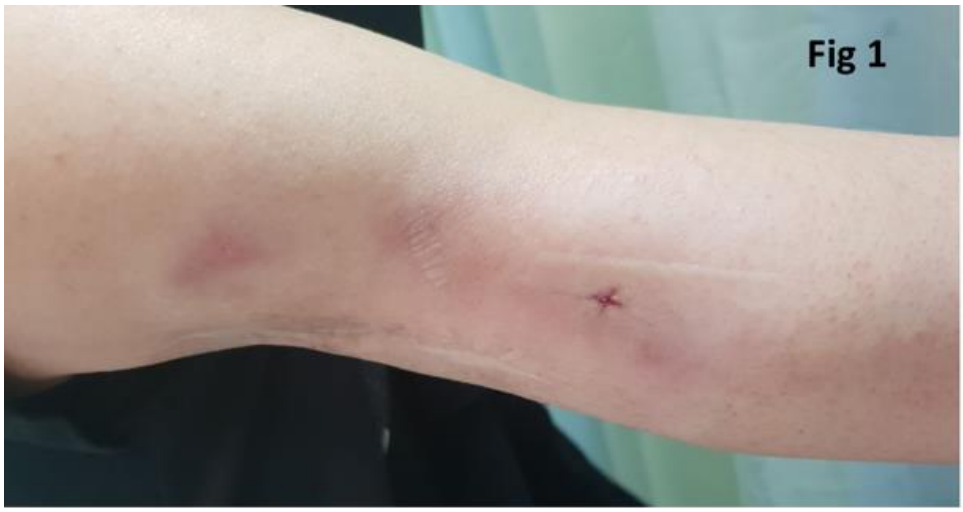

Figure 1 a solitary, indurated, deep, firm, and mobile nodule over the right arm with overlying erythema, loss of hair, and minimal scaling is noted

The histological findings revealed subcutaneous lobular lymphocytic panniculitis with fat necrosis (figure 2). So the patient was diagnosed with lupus profundus. Laboratory tests showed weak positivity for double strand DNA (dsDNA) and antinuclear antibody (ANA). The patient was referred to rheumatology to exclude systemic lupus involvement. Patient then was given hydroxychloroquine 200 mg twice a day. Patient improved initially, but later could not tolerate it as she stated that it affected her memory and her school performance. Accordingly, she was switched to methotrexate in addition to trimacinolone injection. Patient

showed more improvement and response to the new combination. Both were tolerated well and the patient did not complain of any side effects. On the following visits, the patient was improving with no new lesions with on and off ulceration of the primary lesion.

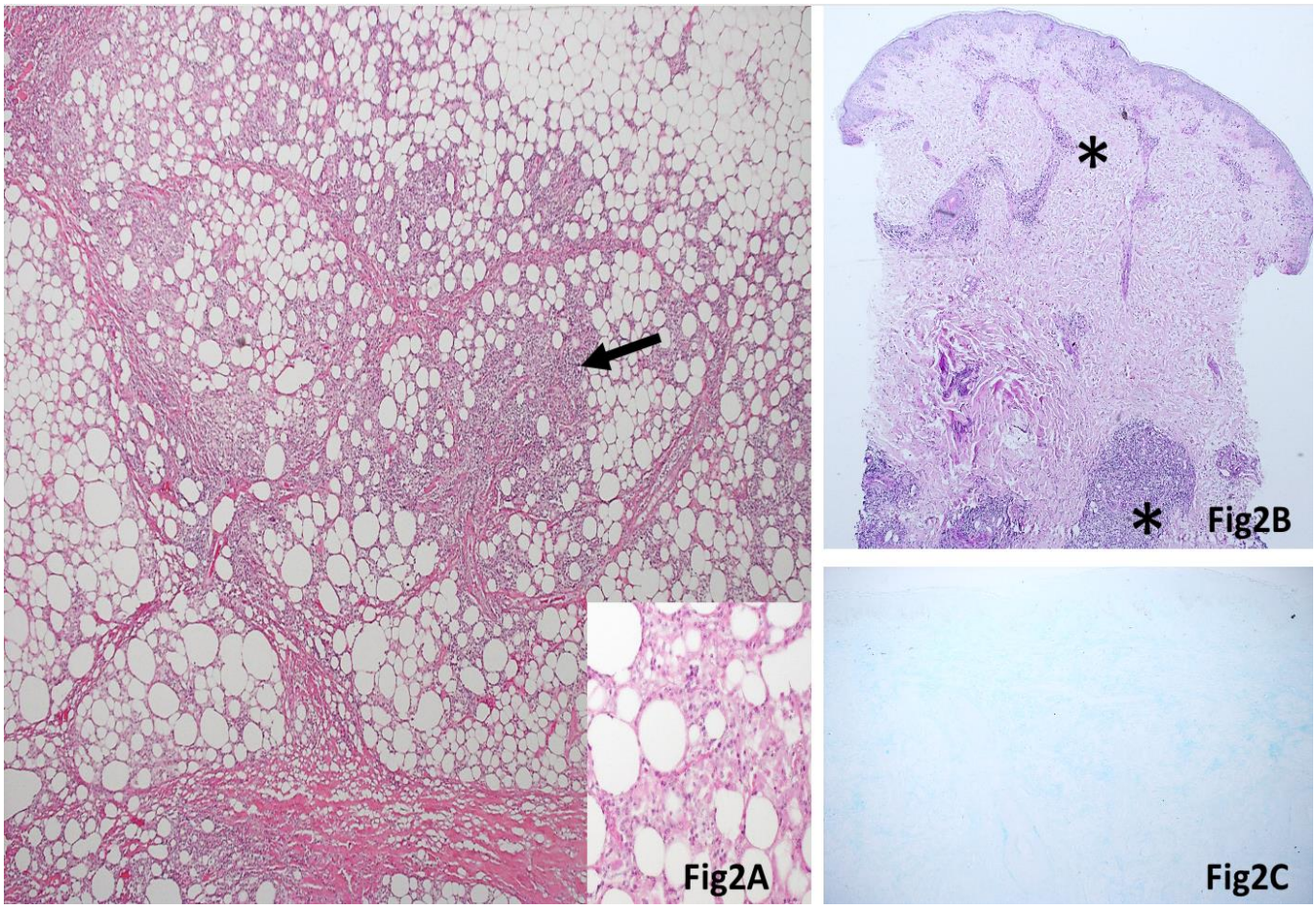

Figure 2A the incisional biopsy shows infiltration of the deep dermis and subcutaneous fat lobules by intense chronic inflammation associated with fat necrosis, and mild fibrosis (arrow). **2B** the epidermis shows moderate superficial and deep perivascular chronic inflammation comprises lymphocytes and plasma cells (asterisk). **2C** the dermis demonstrates deposition of granular blue mucinous material between collagen bundles identified by Alcian blue stain

3. DISCUSSION

Regarding the definition of Lupus erythamoutous profundus, some authors use the term of lupus profundus interchangeable with lupus panniculitis, while others define it as lupus panniculitis with discoid changes (Bolognia et al., 2018). It is a rare and less common variant of CLE. It can occur as a sepearte entity, which is more common than its occurrence with DLE or SLE. Addtionally, only 10% of LEP patients diagnosed with SLE, and one-third of the patients have a coexistent discoid lupus erythematosus (Callen et al., 2006). The pathogenesis of LEP is thought to be similar to the autoimmune dysregulation found in SLE, with T cells and macrophage playing a crucial role in the development of LEP. In some patients, C4 complement deficincy thought to cause this disease, especially in childhood or generalized disease (Bolognia et al., 2018).

Case reports addressing LEP are scarce. Through a medical literature search, not many cases were found over the globe, and much less locally. Despite the fact that LEP usually affects adults, two cases documented that the presentation was at earlier age before the twenties, which is similar to our patient (Zhao et al., 2016 ; Zhang et al., 2018). However, Bednarek et al., (2018) reported a case of LEP which presented at mid-forties of age. Unsurprisingly, all three above mentioned cases were female which reflects the female high proportion. In two cases of them, the lesions were extremely painful and occurred in patients' limbs (Zhang et al., 2018; Bednarek et al., 2018). On the other hand, non-acral involvement was reported by Zaho et al., (2016), in one patient complained of non-painful lesion that was involving the face, back, and limbs. In contrast to our case, which had a sudden onset of the lesion, LEP can occur at site of previous trauma, as shown in one case reported by Castrillón et al., (2018), in which the lesion developed two months after a blunt contusion at the same site.

The diagnosis of LEP requires carfeul history and physical examination, with the histopathologic study is the mainstay in the confirming the diagnosis of LEP. It will reveal perivascular infiltrates of mononuclear cells and panniculitis, manifested as hyaline fat necrosis with mononuclear cell infiltration and lymphocytosis. Direct immunofluorescence, additionally, will show immune deposits in the dermal-epidermal junction (Crowson & Magro, 2001). In addition, laboratory and radologic investigations are needed to exclude systmic involvement and other autoimmune disorders (Bolognia et al., 2018). Diffrential diagnosis of such cases include subcutaneous panniculitis-like T-cell lymphoma, panniculitis associated with DM or morphea, pancreatic panniculitis and erythema nodosum (Bolognia et al., 2018).

Management of such cases is usually challenging due to the nature and the chorniciy of this disease (Bolognia et al., 2018). While antimalarial therapies are considered the first line in treatment, systemic corticosteroids can be used in the initial phases of lesions. Other immunosuppressive therapies such as methotrexate, cyclosporin, dapsone, thalidomide, or Intravenous Immunoglobulins (IVIG) can be used (Bolognia et al., 2018). Intralesional steroid injections can be use cautiously to avoid further skin atrophy. Locally, only two cases found during literature review (Mustafa et al., 2012; Al Kaltham et al., 2012). Unexpectedly, one case reported an eight years old boy patient was complaining of swelling and lesions in his nose and the right side of his cheek (Mustafa et al., 2012).

4. CONCLUSION

lupus erythematosus profundus is a rare variant of cutanoeus lupus. Carfeul history, examination, and histopathologic examination are needed to diagnose such cases. However, there is a deficiency of reports addressing LEP, especially locally, which reflects the need for further reports and studies in order to establish a guideline that will make the physician more confident when encounter such cases.

Authors Contribution

All Authors had contributed equally in literature review, writing the manuscript, and reading the pathology slides.

Informed consent

A concent was taken from the patient.

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Conflict of Interest

There are no conflicts of interest.

Data and materials availability

All data associated with this study are present in the paper.

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