GOAP Dermatology

Distinguishing Cutaneous Manifestations of Lupus from Panniculitis-like T-Cell Lymphoma

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Received date: September 20, 2020; Accepted date: Nov 20, 2020; Published date: Nov 25, 2020

Abstract

Distinguishing cutaneous manifestations of lupus, such as lupus profundus and discoid lupus, from a variant of T-cell lymphoma known as subcutaneous panniculitis-like T-cell lymphoma. These disease states are characterized by tender and hard nodules that are localized on the face, arms, shoulders, breast, and buttocks. In this case we describe a 69 year-old female with a pre-vious history of systemic lupus erythematosus and discoid lupus who presented with worsening alopecia and frontal scarring despite being previously well controlled on Hydroxychloroquine alone. She was evaluated by dermatology and was receiving intralesional steroid injections, but ultimately did not have any improvement. She had a skin biopsy to evaluate further that was consistent with subcutaneous panniculitis-like T-cell lymphoma and she was started on Methotrextate and Prednisone 1 mg/kg, resulting in improvement of her symptoms. As demonstrated by Arps & Patel and the above case presentation, the importance of a biopsy and inflammatory markers for distinguishing between a cutaneous manifestation of lupus (i.e. discoid lupus vs panniculitis vs discoid lupus with features of panniculitis) from panniculitis-like T-cell lymphoma becomes essential and should be a consideration when a patient is having worsening lesions despite already being on appropriate medical therapy.

Keywords: Discoid lupus, Lupus profundus, Subcutaneous panniculitis-like T-cell lymphoma

Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease with a variety of clinical manifestations that can range from milder skin rashes to life threatening complications [1]. Skin involvement occurs in 70-85% of SLE patients and is classified based on histopathologic find-ings and can be subcategorized into specific cutaneous systemic lupus erythematosus (CLE) and non-specific cutaneous systemic lupus erythematosus [2]. Specific lesions are further subcategorized into chronic, subacute, and acute. Of the chronic categorization, discoid lupus (DLE) is the most common form of chronic CLE and may be the initial presentation of SLE in up to 10% of cases [3]. Of important note, 10% of patients that have DLE develop lupus panniculitis, which is characterized by tender and hard nodules localized on the face, arms, shoulders, breast and buttocks [4]. This presentation is difficult to distinguish from a rare variant of T-cell lymphoma known as subcutaneous panniculitis-like T-cell lymphoma as this will also present with tender and hard nodules or plaques that are typically the result T-cell infiltration of the subcutaneous fat tissues [5]. There is an association with autoimmune disorders, with approximately 20% of patients having an associated autoimmune disease, most commonly SLE [6]. Although these two disease processes are distinct from a histopathologic standpoint, distinguishing these two processes remains to be difficult. In this case, we present a patient whose presentation illustrated the difficulty of characterizing the difference between discoid lupus with concomitant panniculitis vs subcutaneous panniculitis-like T-cell lymphoma, which
was able to be illustrated via biopsy of a progressive lesion on her scalp and improvement with more appropriate medical therapy.

**Case Report**

A 69-year-old African American female with a past medical history of SLE characterized by joint pain and discoid lupus and hypertension presented with concerns of worsening parietal and frontal scarring alopecia, and fatigue.

**Figure 1:** Patient's presenting alopecia and frontal scarring.

She was diagnosed with discoid lupus many years ago with characterized by circular plaques with central scarring on her buttocks, bilateral shoulders, and bilateral arms. She had positive ANA titers 1:320, an anti-Smith/RNP antibody level of 7.1 AI (normal 0.0-0.9 AI), an anti-chromatin antibody level of 1.1 AI (normal 0.0-0.9 AI), and an anti-SS-A antibody of 1.3 AI (normal 0.0-0.9 AI), normal double stranded DNA (normal 0.0-0.9 AI), and normal complements (normal C3 80-160 mg/dL, normal C4 15-45 mg/dL). Her skin biopsy was consistent with discoid lupus. She was initiated on Hydroxychloroquine 200 mg daily with stabilization of her symptoms. She was compliant with this regimen and routinely followed with Rheumatology and Dermatology for monitoring of her SLE.

Given worsening scarring alopecia, it was assumed that her discoid lupus was flaring. Her Dermatologist performed an intralesional triamcinolone injection into the enlarging lesion on her scalp however this did not result in any improvement. Based on the lack of improvement, this lesion was biopsied and demonstrated an atypical dermal and subcutaneous lymphoid infiltrate around the adipose tissue, consistent with subcutaneous panniculitis-like T-cell lymphoma. To rule out systemic T-cell lymphoma, serum TCR-beta and TCR-gamma sequencing was performed and was negative, however immunofluorescence from the biopsied lesion was positive for TCR-gamma, further confirming the presence panniculitis-like T-cell lymphoma. A PET scan was performed rule out metastasis as well, given the propensity of developing aggressive lymphoma. She was initiated on Methotrexate 20 mg weekly, along with folic acid and prednisone at a dose of 1 mg/kg. On this regimen, her scalp lesions began to improve, with subsequent decrease in size of her lesion and regrowth of her hair. Her oral prednisone was tapered off with complete resolution of her symptoms.

**Discussion**

The diagnosis of panniculitis-like T-cell lymphoma from discoid lupus/panniculitis is a distinction that has been proven to remain challenging. Arps & Patel detail a similar clinical scenario but in a 25-year-old female who presented with tender nodules on her bilateral arms and flanks. She had a punch biopsy that revealed lobular panniculitis that expansion into extended inter-lobular septa [6]. She had multiple aggregates of CD3-positive T cells however her biopsy results and her serum markers were negative for TCR alpha, beta, and gamma. As a result, she was diagnosed with discoid lupus complicated by panniculitis and improved with topical 1% Clobetasol [6]. However, prior to the biopsy results, the authors noted that distinguishing this patient's presentation from a subcutaneous panniculitis-like T-cell lymphoma was very difficult, which is why they immediately pursued biopsy [6].

As demonstrated by Arps & Patel and the above case presentation, the importance of a biopsy and inflammatory markers for distinguishing between a cutaneous manifestation of lupus (i.e. discoid lupus vs panniculitis vs discoid lupus with features of panniculitis) from panniculitis-like T-cell lymphoma becomes essential. Magro et al. note that the lesions that appear as a result of discoid lupus complicated by panniculitis is cutaneous lymphoid dyscrasias [7,8]. Multiple authors have classified this even further, referring to these processes as lymphocytic lobular panniculitis [7-9]. In order to dis-cuss properly distinguishing these disease entities, it is important to review how these looks from a histopathologic standpoint. Discoid lupus will demonstrate epidermal atrophy, interface changes, a
thickened basement membrane, superficial and deep perivascular and periadnexal lymphocytic inflammation, and increased dermal mucin [6]. In the remainder of cases, findings consist of a predominantly lobular lymphocytic panniculitis in the absence of typical epidermal and dermal changes of lupus [6]. Lupus panniculitis will demonstrate lymphoplasmacytic inflammation, thickened septa, lymphocytic vasculitis, mucinous change, and the presence of occasional eosinophils [6]. Because of some of the overlapping characteristics noted between the two, it is common that these processes are concomitant [6,7,9].

Subcutaneous panniculitis-like T-cell lymphoma will have a similar histopathologic appearance as lupus panniculitis, however immunofluorescence and plasma expression of important markers will become critical for distinguishing further, as noted with the case described above and the case by Arps & Patel. The profile of these markers and immunofluorescence patterns tend to be as follows: CD3+, CD8+, CD4−, TCR-alpha, TCR-beta, TCR-gamma, with expression of cytotoxic markers such as TIA1, perforin, and granzyme B [6,7]. Although our patient did not have the cytotoxic markers expressed, she was CD3+, CD8+, and TCR-gamma positive. It was because she had the TCR-gamma expression that she had a PET scan, as this subset has a higher proclivity to transforming to aggressive T-cell lymphoma and is generally associated with poorer prognosis when it does [4-6]. Fortunately, our patient also did not express these markers in her serum and were strictly limited to cutaneous involvement and she improved after initiating on higher dose Prednisone and Methotrexate.

**Conclusion**

The importance in distinguishing cutaneous manifestations of lupus from panniculitis-like T-cell lymphoma is illustrated in the above case, as once the diagnosis was successfully made and she received appropriate treatment with higher dose Prednisone and Methotrexate with improvement in her scalp lesion. It is important to pursue punch biopsy for prompt diagnosis with understanding of the major histopathologic changes that can be seen with discoid lupus, lupus panniculitis, and panniculitis-like T-cell lymphoma. To further clarify, it is important to perform both immunofluorescence and cytotoxic markers/serum anti-inflammatory markers to help make the precision of diagnosis as precise as possible. Most importantly, as also noted with the above case, when patient is having recurrent lesions that appear especially while on appropriate medication, it is important to monitor for the occurrence of panniculitis-like T-cell lymphoma with a diagnosis of an autoimmune disease (especially SLE) given its association.

**Acknowledgements and Declarations**

Ethics and consent were obtained for participation. Patient consent was obtained for both write up and publication as well. Data and material were obtained via electronic medical record access. No conflicts of interest are noted with the information presented in this manuscript. No funding was required for this study. We would like to thank the ChristianaCare Health System and the University of Pennsylvania for allowing the opportunity to evaluate and treat this patient as we did in the case depicted above.

**References**


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