Panniculitis; A Rare Cutaneous Manifestation of Sjögren’s Syndrome

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ABSTRACT

Objective: Panniculitis, an inflammatory disorder primarily affecting subcutaneous adipose tissue, is frequently associated with inflammatory rheumatic diseases. Panniculitis is an uncommon cutaneous manifestation of Sjögren’s syndrome (SS), an autoimmune disease primarily known for causing dry eyes and mouth.

Case: We report the case of a 56-year-old female initially diagnosed with rheumatoid arthritis, who later met the 2016 ACR-EULAR criteria for primary Sjögren’s syndrome. Following several treatments, she developed nodular lesions in the interscapular area, which were later diagnosed as septal panniculitis associated with SS. Panniculitis can sometimes precede the formal diagnosis of systemic diseases, such as SS. Therefore, it is essential to consider conditions like Sjögren syndrome in the differential diagnosis of unexplained panniculitis.

Conclusion: This case underscores the potential link between Sjögren's syndrome and panniculitis. Panniculitis may manifest before the formal diagnosis of Sjögren's syndrome. It is imperative to include connective tissue diseases, including not only the more common lupus erythematosus and dermatomyositis but also Sjögren's syndrome, in the differential diagnosis when confronted with unexplained cases of panniculitis.

Keywords: Sjögren's syndrome, Panniculitis, rheumatoid arthritis.

INTRODUCTION

Panniculitis is an inflammatory condition characterized by inflammation of the subcutaneous adipose tissue. Rheumatologists, in particular, may be involved in assessing and managing panniculitis, often as part of inflammatory rheumatic diseases. Sjögren’s syndrome (SS), an autoimmune disease primarily characterized by dryness of the eyes and mouth, can also present with various cutaneous manifestations. Cutaneous manifestations associated with SS include purpura and urticaria, usually with small-vessel vasculitis. Panniculitis associated with SS is an unusual cutaneous manifestation. In this case report, we describe the case of a patient with SS and septal panniculitis.

CASE

A 56-year-old female patient was referred to our outpatient rheumatology clinic with complaints of lassitude, fatigue, widespread joint pain, myalgia, low-grade fever, and shortness of breath for two months. The patient had been diagnosed with rheumatoid arthritis two years ago and had used DMARDs such as methotrexate and leflunomide. However, she did not experience any therapeutic benefit and discontinued medication for a year. The patient had no comorbidities.

The rheumatological evaluation revealed widespread arthralgia and morning stiffness for more than 30 minutes. She also complained of severe dryness of her mouth and eyes. There were no signs of arthritis, back pain, Raynaud’s phenomenon, photosensitivity, diarrhea, oral ulcers or genital ulcers. She had no history of uveitis, thrombosis or abortions.
The findings of laboratory tests showed anti-nuclear antibody (ANA) 4 +, anti-Ro/SSA 3+, anti-La/SSB 2+, RF 94 IU/ml, elevated erythrocyte sedimentation rate (102 mm/h) and decreased haemoglobin (9.8 g/dL). Her other results for liver and renal function tests, urine analysis, anti-dsDNA, anti-sm, anti-CCP and complements were within normal limits. Viral panels for hepatitis B/C and HIV were negative, and serum protein electrophoresis results were normal. Schimmer’s test was 3 mm for both eyes. Salivary gland biopsy was performed, and results showed lymphocytic sialadenitis in more than three focuses. The patient fulfilled the 2016 ACR-EULAR classification criteria for primary Sjogren’s syndrome.

In the chest CT scan, prominent perivascular markings and increased density were observed in the regions of both lower lung lobes. Additionally, there was a 1.5 cm pericardial effusion, although it did not affect cardiac function. Based on these findings, the patient was initiated on treatment with methylprednisolone at a dose of 16 mg/day, hydroxychloroquine, and azathioprine at a dose of 100 mg/day. However, the patient developed intolerance to azathioprine, and it was subsequently replaced with mycophenolate mofetil.

After four weeks of treatment, the patient showed significant improvement in clinical symptoms, and the steroid dose was gradually tapered. Six months after the initial diagnosis, the patient presented to the clinic with complaints of a tender, indurated nodular plaque on the interscapular area (Fig. 1). A biopsy was performed with a preliminary diagnosis of sarcoidosis, lupus panniculitis, erythema nodosum, and cutaneous T-cell lymphoma.

Figure 1. The Indurated Nodular Plaque Lesion of the 56-year-old Female Patient on the Interscapular Area at 6 Months Post-Diagnosis

The biopsy revealed superficial perivascular mild inflammatory cell infiltration in the dermis. There was also thickening of collagen bundles in the deep dermis with fibrosis in the subcutaneous fatty tissue. Moreover, a mixed inflammatory cell infiltration rich in eosinophils was occasionally seen extending into lobules (Fig. 2.a-b). Based on these results, the patient was diagnosed with Sjogren's syndrome-associated septal panniculitis.

Colchicine and methylprednisolone were added to her treatment regimen, and the patient's lesions resolved within six weeks. She continued to receive hydroxychloroquine and mycophenolate mofetil for the management of her underlying Sjogren's syndrome.

DISCUSSION

Panniculitis, an inflammatory disorder that primarily affects the subcutaneous fat tissue, is characterized by erythema, swelling, and tenderness. It can manifest in various forms, including septal panniculitis and lobular panniculitis, and is often associated with systemic conditions such as connective tissue diseases, autoimmune disorders, and vasculitis (1,2).
In connective tissue diseases, panniculitis may manifest either as an isolated presentation of the disease or in conjunction with other clinical features of the underlying pathological process (3). The most well-documented variations of panniculitis occurring in the setting of connective tissue diseases encompass lupus erythematosus panniculitis (LEP), lupus profundus, panniculitis linked with dermatomyositis, as well as panniculitis associated with morphea and scleroderma.

The pathogenesis of panniculitis is complex and involves immune-mediated processes, as well as vasculopathic changes. Histopathologically, these disorders are characterized by liponecrobiosis, the existence of granulocytic infiltrates in both lobules (lobular panniculitis) and septae (septal panniculitis), while vasculitic lesions may or may not occur (4). Diagnosis relies on clinical evaluation, histopathological examination, and identification of the underlying systemic diseases.

Sjögren's syndrome patients usually present with various cutaneous manifestations such as dry skin (xeroderma), purpura, Raynaud's phenomenon, vasculitis, and erythema (5). Panniculitis linked to Sjögren's syndrome represents an uncommon cutaneous presentation. The clinical and pathological observations in our patient were consistent with panniculitis associated with Sjögren's syndrome. The mechanisms responsible for these occurrences in Sjögren's syndrome patients remain unclear, likely attributed to the scarcity of documented cases. Nevertheless, immune complexes have been postulated as potential mediators.

For panniculitis treatment, numerous therapeutic approaches have been used, encompassing anti-malarials, dapsone, colchicine, glucocorticosteroids, methotrexate, azathioprine, and IL-1 receptor antagonist inhibitors (7-8). However, there is currently a lack of studies that definitively advocate for the most suitable treatment regimen. Typically, the choice of treatment is determined based on the clinician’s expertise and experience.

CONCLUSION

We have presented this case to emphasize the connection between Sjögren's syndrome and panniculitis. Panniculitis may manifest before the formal diagnosis of Sjögren's syndrome. It is crucial to consider connective tissue diseases, including not only the more prevalent lupus erythematosus and dermatomyositis but also Sjögren's syndrome, in the differential diagnosis when confronted with unexplained cases of panniculitis.

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Ethical approval: The present study was conducted in strict accordance with the principles outlined in the Declaration of Helsinki. Ethical approval for the study was obtained from the appropriate ethics committee, and all participants provided informed consent before participating in the study.

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