Overlap of Ankylosing Spondylitis and Systemic Lupus Erythematosus: A case report

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ABSTRACT

Ankylosing spondylitis is a typical, very heritable incendiary joint inflammation, influencing principally the spine and pelvis. Inflammatory arthritis in ankylosing spondylitis causes pain and stiffness and progressively leads to new bone formation and ankylosis (fusion) of affected joints. Systemic lupus erythematosus (SLE, lupus) is a highly complex and heterogeneous autoimmune disease that most often afflicts women in their child-bearing years. It is characterized by circulating self-reactive antibodies that deposit in tissues, including skin, kidneys, and brain, and the ensuing inflammatory response can lead to irreparable tissue damage. There are few reports of coexistence of Ankylosing spondylitis and Systemic lupus erythematosus which firmly emphasis on an overlap phenomenon between these two disorders. A 30 year old woman was admitted to our hospital due to signs of butterfly-shaped rash on her cheeks, which became prominent after exposure to sunlight and severe inflammatory low-back pain. About ten year earlier, AS had been diagnosed and treatment started with non-steroidal anti-inflammatory drug (NSAID). To the best of our knowledge, the present case is one of 10 reported cases of coexistence of these two disorders in English literature. The coexistence of these two diseases with different genetic backgrounds and clinical symptoms may implicate the importance of shared environmental factors.

Keywords: Ankylosing spondylitis (AS), Systemic lupus erythematosus (SLE), Overlap syndrome, Connective tissue disease (CTD).

INTRODUCTION

The spondyloarthropathies (SpA), now better known as spondyloarthritides (SpAs), are a diverse range of interrelated inflammatory arthritides that share multiple clinical features and common genetic predisposing factors. This group includes not only the prototypical disease, ankylosing spondylitis (AS), but also reactive arthritis (ReA), psoriatic arthritis (PsA), Crohn's disease, undifferentiated SpA, and juvenile-onset spondyloarthritis\cite{1}.

The clinical features of AS includes inflammatory back pain, asymmetrical peripheral oligoarthritis, enthesitis, and specific organ involvement, such as anterior uveitis, psoriasis, and chronic inflammatory bowel disease \cite{2}. Its major clinical features include sacroiliitis, loss of spinal mobility, and spinal inflammation. Chronic inflammation leads to fibrosis and ossification, where bridging spurs of bone known as syndesmophytes form, especially at the edges of the inter-vertebral discs, producing the ankylosing\cite{3}.
AS is more prevalent in men in comparison with women, at a ratio of 2:1 [4]. The prevalence of the disease is between 0.1% and 1.4% of general populations [2]. Studies conducted in different countries have shown that the incidence of AS varies from 0.5 to 14 per 100,000 people per year [2]. Diagnoses of AS are based more on clinical features than on laboratory tests; currently, diagnoses are made in accordance with the modified New York criteria [5].

Systemic lupus erythematosus (SLE or lupus) on the other hand, is a complex and severe rheumatic disease with exceedingly diverse clinical manifestations. The most characteristic symptoms that allow recognition of the disease includes skin lesions (malar rash, discoid rash), sores in mouth or nose, musculoskeletal manifestations as arthritis, arthralgia or myositis, bone fragility fractures and secondary pain amplification [6]. Currently the diagnosis is based on American College of Rheumatology (ACR) criteria. Simultaneous presence of four (or more) out of eleven criteria allows the identification of the disease.

SLE is classified among systemic autoimmune disorders because of the presence of autoantibodies such as: antibodies directed against double-stranded DNA (anti-dsDNA), anti-small nuclear RNA-binding proteins, anti-phospholipid antibodies (aPL) or anti-Smith (anti-Sm) nuclear antigens in abnormal titer [7].

Overall improvements in medical care including the availability of antibiotics, antihypertensives, and renal replacement therapy coupled with the judicious use of glucocorticoid, antimalarial, and immunosuppressive drugs have led to improved survival of SLE patients in the past 50 years [8]. Despite the improvements in care, patients often suffer long-term morbidity that can adversely affect their quality of life and their ability to work, resulting in substantial direct and indirect costs.

Ethnicity, a broader construct than is implied by the term “race,” encompasses genetic, geographic, cultural, social, and other characteristics shared within a population [9-11]. Not surprisingly, the phenotypic expression of lupus varies between individuals of different ethnic groups. A growing body of research has sought to characterize the influence of socioeconomic factors and ethnicity on the incidence, activity, and progression of the disease[12]. Less attention has been paid to the influence of psychosocial factors on disease progression. Unlike AS, SLE affects only about 10% of male population. SLE is often described as a disease that most often strikes reproductive-age women[13].

Figure 1. Anteroposterior view of the pelvis showing sclerosis, narrowing, and erosions of both sacroiliac joints.

These two autoimmune rheumatologic diseases, which have a different aetiopathogenesis as well as diverse clinical and genetic characteristics, are rarely seen together. To the best of our knowledge, there are only 9 reported cases of the coexistence of SLE and AS in the English literature. Here, we report another case with the coexistence of these two diseases. The present patient is a 30 year-old female who was being followed-up due to the diagnosis of AS, and later received the additional diagnosis of SLE.
Case Presentation
A 30 year old woman was admitted to our hospital due to signs of butterfly-shaped rash on her cheeks, which became prominent after exposure to sunlight and severe inflammatory low-back pain. About ten year earlier, AS had been diagnosed and treatment started with non-steroidal anti-inflammatory drug (NSAID). The diagnosis of AS was made based upon the presence of inflammatory arthritis, bilateral sacroiliitis, limitation in back movements and positive HLA B27. 3 months before admission signs of malar rash and photosensitivity began and patient started to develop oral ulcers, alopecia, together with pleuritic chest pain. These symptoms along with anti-dsDNA and antinuclear antibody (ANA) positivity and low serum complement levels were resulted in diagnosis of SLE. Liver and renal function tests, serum protein and creatinine phosphokinase levels, platelet count, urine analysis and thyroid function tests were within the normal limits. The chest x-ray was normal. Her family history was not positive for SLE or AS. An anteroposterior view (fig 1) of the pelvis showed bilateral sacroiliitis.

DISCUSSION
Although connective tissue diseases (CTDs) can generally be clinically and serologically defined as distinct and separate entities, many patients diagnosed with autoimmune rheumatic disease cannot be categorized easily into one of the established conditions. Several diseases share similar genetic backgrounds, as reflected by study of loci within the major histocompatibility complex and some genetic defects can predispose patients to more than one autoimmune disease[14]. The existence of patients with signs, symptoms and certain laboratory test results suggestive of a systemic autoimmune disease but fulfilling more than one classification criteria for well-defined CTDs is a more and more common experience in clinical practice. As opposed to some early stages of CTDs that might be undefined, unclassifiable or perhaps incomplete, with clinical elements and laboratory results suggestive of a systemic disease but not fulfilling criteria for well-defined CTDs, overlap syndromes define patients exhibiting enough features to meet the diagnosis of several CTDs at the same time. Thus, they “overlap” two or more diseases. Any CTD can be a partner in an overlap disorder[15]. For example, patients can have a combination of RA and SLE («rhupus»), or SSc and PM.

Generally, any CTD can appear in conjunction with features of another connective tissue diseases and thus, it does no longer fit in the traditional classification. The problem is even more complex by the tendency for one disease to merge with another, resulting in a continuous spectrum, with the traditionally accepted entities such as SLE or SSc occupying only part of the continuum with the overlap syndromes lying between. It is not only a co-association, but also a confluence and union of autoimmune disorders.

Therefore, to identify an overlap syndrome it is essentially necessary to identify a constellation of distinctive features that constitute a true syndrome. Mixed connective tissue disease (MCTD) is the prototype of an overlap syndrome. Since its original description by Sharp and colleagues in 1972[16], as an apparently unique syndrome combining clinical elements of SSc, SLE and PM, associated with antibodies to RNA se sensitive extractable nuclear antigen, many clinical, serologic, and genetic studies have analyzed the different aspects of this entity. The relevance of defining MCTD as a separate disease entity has been challenged, some authors considering it just a subset of SLE.

Including the present case, most of the reported cases of SLE and AS coexistence are females and generally SLE precedes the occurrence of AS. The coexistence of these two diseases with different genetic backgrounds and clinical symptoms may implicate the importance of shared environmental factors.

REFERENCES