Simultaneous Presentation of Ocular Sarcoidosis and Early Axial Spondyloarthritis in a Young Woman

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Axial spondyloarthritis and sarcoidosis are both inflammatory multi-system diseases. Having different pathophysiology, they develop different typical lesions. The co-occurrence of both diseases is rare and nature of the association between the entities is unknown.

Key Words. Axial spondyloarthritis, Sarcoidosis

Introduction

Axial spondyloarthritis (SpA) is an inflammatory disease that predominantly affects vertebral and sacroiliac joint. Sarcoidosis is an inflammatory multi-system disease associated with the formation of non-caseating granulomas within involved systems (1). Both diseases can involve ocular manifestations of which anterior uveitis are common feature. Articular involvement has been reported in about 5% of cases with sarcoidosis (2). Therefore, it is sometimes difficult to distinguish one disease from the other. Sacroiliac joint involvement, in which the hallmark of axial SpA (3,4), is not a common feature of sarcoidosis. Here, we present a case with simultaneous onset of early axial SpA with ocular sarcoidosis.

Case Report

A 27-year-old woman was admitted to our ophthalmologic clinic with complaints of conjunctival injection and a pricking sense in both eyes for 2 months. Her medical history and familial history were unremarkable without constitutional symptoms, fever, weight loss, fatigue, arthralgia, myalgia and night sweating.

On the funduscopic examination, anterior chamber cell and perivascular sheathing were observed in both eyes. Furthermore, it showed peripheral vascular and disc leakage on fundus fluorescent angiography. It is consistent with bilateral anterior uveitis with retinal vasculitis. She was referred to a rheumatology clinic for an evaluation about the underlying disease causing uveitis. A physical examination revealed cervical lymphadenopathy. In a laboratory assessment, rheumatoid factor, anti-nuclear antibody, C3/C4, erythrocyte sedimentation rate, C-reactive protein, and other blood chemistry values were <2.0 IU/mL (0∼20), negative, 121/22 mg/dL (76∼139/12∼37), 23 mm/hr (0∼20), 0.13 mg/dL (0.01∼0.47), respectively. HLA-B27 was positive. Tuberculin skin test and interferon gamma release assay were negative.

Chest computed tomography (CT) scan revealed enlarged lymph nodes in the mediastinum and hilum (Figure 1). A biopsy of the left cervical lymph node showed non-caseating granuloma, consistent with sarcoidosis (Figure 2). Angiotensin converting enzyme (ACE) level was elevated up to 131 more than 2-fold over the upper normal limit (18∼55). Plain film of the sacroiliac joint did not show a definite lesion. However, sacroiliac joint magnetic resonance image (MRI) was performed because she complained low back pain in the morning and HLA-B27 was positive. Subchondral erosion and bone marrow edema were observed on MRI Short Tau Inversion Recovery (STIR) sequence (Figure 3). These findings were in favor of axial SpA diagnosis.
Figure 1. Chest computed tomography shows lymphadenopathy of mediastinum (left) and hilum (right). The white arrows indicate enlarged lymph nodes in mediastinum (left) and hilum (right).

Figure 2. Multiple non-caseating granulomas on cervical lymph node biopsy and macrophage infiltration (×400).

Figure 3. MRI STIR sequence of sacroiliac joint shows bone marrow edema (white arrow) and subchondral erosion (white arrow head).

The case was diagnosed as the co-existence of ocular sarcoidosis and early axial SpA. Oral prednisolone, celecoxib, methotrexate were administered 30 mg/day, 200 mg/day, 10 mg/week, respectively. She had a regular check-up in ophthalmology clinic because ocular symptoms improved. After tapering of the prednisolone to 10 mg/day through 11 weeks, patient’s low back stiffness and pain deteriorated. At that time, Bath Ankylosing Spondylitis Disease Activity Index (BASDAI) score was 6.65. So infliximab was started in February 2012. The low back stiffness and pain improved after infliximab treatment. A CT scan and ACE levels were examined for sarcoidosis follow-up. The CT scan showed interval regression of mediastinal and hilar lymphadenopathy, and the ACE level was 50.2, which was within normal limit.

Discussion
Co-existence of axial SpA and sarcoidosis has been reported in only a small numbers of cases (5-7). Many of the cases were newly developed sarcoidosis in patients with advanced state of axial SpA. The difference with our case was a time of sarcoidosis development in which both diseases developed simultaneously in present case. Importantly, we had to differentiate two manifestations for proper diagnosis.

First, uveitis can develop in both diseases. It is critical which disease causes ocular symptoms that the patient complained. When the two diseases are present at the same time, it is not easy to clarify whether one disease is the cause. Although up to a third can be asymptomatic, acute anterior uveitis is the most common presentation in sarcoidosis. Involvement is usu-
ally bilateral, which distinguishes it from the unilateral uveitis seen in HLA-B27 associated spondyloarthropathies. Sarcoidosis can be presented by posterior uveitis including retinal vasculitis (8). But, bilaterality and posterior uveitis are rare in axial SpA (9,10). Therefore, the ocular symptom in our patient may be caused by sarcoidosis.

Second, sacroiliitis may be presented with not only axial SpA but sarcoidosis, due to sarcoid deposition (5). In a cross-sectional study, four of the sixty-one sarcoidosis patients (6.6%) had radiological erosive sacroiliac joint (3). Relatively high sacroiliitis incidence in sarcoidosis group rather than general population may be caused by sarcoid deposition in sacroiliac joint. Only sacroiliac joint biopsy can reveal noncaseating granuloma consistent with sarcoid deposition. To our knowledge, MRI findings in patients with sarcoidosis of sacroiliac joint, not spinal involvement have not been reported. Therefore, it is difficult to confirm what the cause of sacroiliitis is. In present case, low back pain did not improve despite pulmonary hilar lymphadenopathy and ocular sarcoidosis ameliorated after steroid treatment. And low back pain showed good response after infliximab therapy. So we considered that sacroiliitis is caused by axial SpA.

As above, axial SpA and sarcoidosis have similar clinical appearance, which may be due to common pathophysiology such as increased CD4+ T lymphocyte. CD4+ and CD8+ T lymphocytes are induced in patients with axial SpA, which is important role for alloreactivity (11). CD4+ T lymphocytes are involved in the formation of non-caseous granulomas in patients with sarcoidosis.

Although anti-tumor necrosis factor (TNF) therapy is helpful to axial SpA, it is controversial about effect to sarcoidosis. Several studies have reported that de novo sarcoidosis can occur when treating patients with anti-TNF therapy (12,13). All three anti-TNF agents, etanercept, adalimumab and infliximab, have been observed to cause sarcoidosis-like granuloma formation, suggesting a ‘class effect’ rather than being drug specific (13). On the other hand, it has been reported that anti-TNF therapy is effective for refractory sarcoidosis (14,15).

Summary

It has been reported that anti-TNF therapy may deteriorate disease course of sarcoidosis. If sarcoidosis develops in a patient treating anti-TNF therapy, cessation of the anti-TNF therapy and administration of steroid can induce a good response. Until now, it is not worsen not only axial SpA but also sarcoidosis using anti-TNF antagonist. However, she is carefully supervised about both diseases.

References