Coexistence of Primary Biliary Cirrhosis and Rheumatoid Arthritis: Case Report

**Abstract**

Primary biliary cirrhosis (PBC) and rheumatoid arthritis (RA) are both well defined autoimmune diseases. Associations between PBC and other autoimmune diseases have been reported, but their true extent and pattern is unknown. The most frequent coexisting disorder with PBC is Sjögren’s syndrome. The coexistence of PBC and RA in the same patient is an unusual condition and it’s not noted in the textbooks. We report a case that met the disease-defining criteria for both PBC and RA and intend to attract the attention to the possibility of this association.

**Key Words:** Arthritis, rheumatoid; liver cirrhosis, biliary

**Case Report**

A 58 year-old female was admitted to the hospital with complaints of fatigue, arthralgia and...
pruritus 4 years ago. Laboratory examination had revealed elevations in liver enzymes and AMA positivity. She was diagnosed as PBC. She has used ursodeoxycholic acid (UDCA) since then and her complaints recovered with UDCA therapy. Last year, she began to suffer from pain, swelling and stiffness of the hand joints. She experienced morning stiffness of greater than 2 hours duration and deformities of the hands. She also complained of dry mouth but did not have dry eyes. Physical examination revealed symmetric enlargement of wrist joints, swelling and tenderness of metacarpophalangeal (MCP) and proximal interphalangeal (PIF) joints and atrophy of interosseus muscles (Figure 1). The Schirmer test was negative for dry eyes. Laboratory findings were as follows: Hemoglobin 12.2 gr/dL, hematocrit 36.2%, leukocyte count 5800/mm$^3$, platelet count 225000/mm$^3$, Westergreen erythrocyte sedimentation rate (ESR) 91 mm/h, C-reactive protein (CRP) 16.8 mg/dL and serum rheumatoid factor (RF) 508 IU/mL (25-fold of the upper limit). Posteroanterior hand x-ray revealed narrowing of MCP and PIF joints. The patient met the revised criteria for the classification of RA and was diagnosed as RA. Extraarticular manifestations were searched by high resolution lung CT (HRCT) and echocardiography. Physical and laboratory examinations revealed no sign of heart disease or interstitial lung disease. In addition to UDCA for PBS, prednisolone 100 mg/day iv for three days and a maintenance dose of 8 mg/d po thereafter was administered for RA. Sulphasalazine 2 mg/day was used as a disease modifying antirheumatic drug (DMARD). Bone mineral density measurement by DXA indicated severe osteoporosis and biphosphonate therapy was initiated. With this treatment her arthralgia improved, swelling and tenderness in several joints diminished.

**Discussion**

The diagnosis of PBS is currently based on three criteria: The presence of AMA in serum, elevation of liver enzymes (most commonly ALP) for more than six months, and a liver biopsy finding compatible with the disease. The value of biopsy findings for the diagnosis is questionable and some hepatologists believe that a liver biopsy is not required for diagnosis but only for the staging of the disease. Our patient presented with complaints of fatigue and pruritus, the most common presenting symptoms of PBC and with the detection of elevated liver enzymes and AMA seropositivity she received PBC diagnosis.

The diagnosis of RA depends on well defined criteria. The American Rheumatism Association 1987 revised criteria are:

1) Morning stiffness in and around joints lasting for at least 1 hour before maximal improvement;

2) Soft tissue swelling (arthritis) of 3 or more joint areas observed by a physician;

3) Swelling (arthritis) of the PIF, MCP, or wrist joints;

4) Symmetric swelling (arthritis);

5) Rheumatoid nodules;

6) The presence of RF; and

7) Radiographic erosions and/or periarticular osteopenia of hand and/or wrist joints.

Criteria 1 through 4 should be present for at least 6 weeks. RA is defined by the presence of at least 4 criteria. Our patient had symmetrical arthritis of the wrist, MCP and PIF joints that was defined by the clinician on physical examination.
morning stiffness in and around the joints lasting for more than 1 hour before maximal improvement, serum rheumatoid factor positivity and radiographic changes on posteroanterior hand x-ray. With the presence of six out of seven criteria, RA diagnosis is definite.

Liver function tests may be abnormal in up to 6% of patients with RA and mainly involve increases of ALP and GGT levels. Additionally NSAIDs, sulfasalazine and methotrexate, which are among the most common medications for RA treatment are known to affect liver function. Clinicians tend to consider a drug effect when they meet an abnormal liver test in a RA patient.

Arthritis is not an infrequent finding in PBC. A nonrheumatoid inflammatory polyarthritis may be present in 9% of patients. Although some researchers point out to an association between RA and PBC, current data do not confirm the coexistence of RA and PBC.

Joanna and colleagues reviewed all patients with both RA and PBC, recorded at the Mayo Clinic between 1976-1999 and identified twenty-five patients meeting strict disease-defining criteria for both of these conditions. Their findings suggested that patients with both conditions did not have more severe disease than those with only one condition and most of the patients had received RA diagnosis many years before PBC. This unique cohort should prompt us to consider the association of PBC and RA in a RA patient with elevated ALP levels. This is particularly important with regard to treatment since methotrexate, which is a first-line DMARD was shown to increase mortality in PBC.

Although an unusual condition, the possibility of PBC and RA coexistence should be kept in mind by clinicians. Correct diagnosis of RA in a PBC patient provides aggressive therapy with DMARDs and prevents the development of deformities. Similarly, to consider PBC in a RA patient with elevated liver enzymes provides correct treatment of PBC, slowing the progression to liver failure that may result with liver transplantation or mortality.

REFERENCES