

Jaccoud's arthropathy in primary biliary cirrhosis.

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Abstract

Jaccoud's arthropathy (JA) is a deforming arthropathy, nowadays present mostly in systemic lupus erythematosus (SLE). Primary biliary cirrhosis (PBC), which predominantly affects middle-aged women, is histologically characterized by chronic non-suppurative destructive cholangitis. The chief target of PBC is the liver. We describe a rare case of PBC with hand deformities typical of JA.

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Introduction

Primary biliary cirrhosis (PBC) is a chronic progressive cholestatic liver disease with fibrosis leading to cirrhosis [1-3] and liver failure [4,5]. PBC is often thought of as an organ-specific autoimmune disease, which mainly targets the liver; however, other tissue also can be a site for autoimmune involvement of PBC. Jaccoud's arthropathy (JA) was a non-erosive deforming arthropathy initially described in patients with rheumatic fever (RF). Presently, the majority of the cases are seen in systemic lupus erythematosus (SLE) [6-8]. Whereas, this complication is associated with other clinical conditions.

Case Report

We report this case to discuss the relationship between PBC and JA.

A 57-year-old man was referred to us for assessment of a 2-year history of ascites. During past two years, he had experienced repeated abdominal effusion and jaundice, and an abdominal ultrasound scan showed liver cirrhosis. Because of severe abdominal distension, he was hospitalized at our institution. The patient presented that his hands and feet deformed since nine years ago. At the age of 30 he had acute painless joint swelling of both hands which lasted for one week. Then he felt episodic stiffness of hands but did not notice the deformities until nine years ago. During the course, there were no recurrent fever and pain. Having received a diagnosis of rheumatoid arthritis, the patient once was treated with non-steroid anti-inflammatory drugs (NSAIDs) and prednisone several months but not receive improvement of the deformities. He reported no consumption of alcohol. On physical examination, the patient was without fever and had normal blood pressure. We did not identify abnormalities during the cardiorespiratory evaluation. On abdominal examination, we identified shifting dullness but no hepatosplenomegaly. The patient had swan-neck deformity of the fingers, slightly ulnar deviation of metacarpal bones and comparatively obvious ulnar deviation of right phalanxes. (Figure 1). The function of his hands was limited and the deformities were partly reversible. The X-ray of the hands showed the deformities of the metacarpophalangeal (MCP) and interphalangeal joints, normal joint space and no bone erosion (Figure 2). Meanwhile, there were degenerative changes of the joints. Laboratory tests results indicated haemoglobin of 10.5

g/dL, white blood cell count 2700 cells/mm³; the erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) were 77 mm/h and 127 mg/L; IgG (36.2 g/L), IgA (7.2 g/L) and IgM (8.9 g/L) increased while serum complements (C3 0.304 g/L, C4 0.081 g/L) decreased. Antimitochondrial-M2 antibodies (AMA-M2) (62 normal<1) and antinuclear antibodies (ANA) (1:1000) were high-positive; rheumatoid factors, anti-dsDNA, and anti-CCP were negative. In the serum markers of hepatitis virus, HBsAb was positive; HBsAg, HBeAg, HBeAb, HBcAb, Anti-HAV, Anti-HCV, Anti-HDV and Anti-HEV were negative. The liver function tests revealed following: aspartate aminotransferase (AST), 68 U/L; alanine aminotransferase (ALT), 21U/L, gamma-glutamyltransferase (GGT), 411U/L; alkaline phosphatase (AKP), 642 U/L; total bilirubin (T-BIL), 41.9 umol/L; albumin, 34 g/L. On the basis of these results, we made the diagnosis of PBC and JA. The patient was treated with ursodeoxycholic acid (UDCA) (750 mg/day), diuretics and drawing ascites. The patient's clinical findings and biological data showed improvement and the patient was discharged 20 days after admission.

Discussion and Conclusion

PBC is an important cause of liver cirrhosis and endstage liver disease [2]. It has 4 histologic stages: (1) portal inflammation with or without florid bile duct lesions; (2) increase in size of periportal lesions with interface hepatitis; (3) distortion of hepatic architecture with numerous fibrous septa; and (4) cirrhosis [9]. Our patient belonged to stage 4. Both genetic and environmental triggers have been considered important for the induction of PBC, as well as other autoimmune diseases [10-12]. Many reports indicate that PBC, as a kind of autoimmune liver diseases (AILD), is related to other autoimmune diseases, such as Sjogren's syndrome (SS), systemic lupus erythematosus (SLE), systemic sclerosis (SSc), rheumatoid arthritis (RA), dermatomyositis and so on [13]. However, there was no described case of JA associated with PBC. Jaccoud's arthropathy is a syndrome characterized by progressive, painless and nonerosive deformity mainly affecting the joints of hands and feet. But a limitation on the study of JA is the lack of definite diagnostic or classification criteria. The previous studies demonstrate that the classical JA include the presence of "reversible deformities" and



Figure 1. Swan-neck deformity of the fingers and ulnar deviation.



Figure 2. Swan-neck deformity of the fingers and ulnar deviation.

absence of erosions on X-rays and rheumatoid factor negativity [14] or as “any deviation of the metacarpus finger axes assessed by a goniometer” [15]. Basing on these clinical features, Spronk et al. [16] developed a diagnostic “index” which allowed for the presence of different deformities and attributing JA a score of over five points. Our patient presented the features of JA. The mechanisms responsible for development of JA are not yet well defined. Persistent synovitis and fibrotic retraction of the joint capsule are the mechanisms involved in the development of JA [17]. It was unknown if PBC led to JA in our patient. Because the pathogenesis of two diseases maybe involves autoimmune and environmental factors [3,6,7], PBC and JA in our case possibly were caused by the common factors.

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